Referral and Management Guidelines for Skin Cancers within North Trent

August 2012

Date for guideline review: August 2013

Produced by the
North Trent Cancer Network Skin Cancer Group
# North Trent Skin Cancer Guidelines

August 2012

## REFERRAL AND MANAGEMENT GUIDELINES FOR SKIN CANCERS WITHIN NORTH TRENT

## CONTENTS

<table>
<thead>
<tr>
<th>Section</th>
<th>Content</th>
<th>Page No</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.0</td>
<td>Network Configuration of Teams</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>1.1 Network Configuration for Community Skin Cancer Services</td>
<td>4</td>
</tr>
<tr>
<td>2.0</td>
<td>Primary Care Referral Guidelines</td>
<td>5</td>
</tr>
<tr>
<td>3.0</td>
<td>Clinical Guidelines:</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3.1 Basal Cell Carcinoma</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>Squamous Cell Carcinoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Malignant Melanoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3.2 Dermatofibromasarcoma Protuberans</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>3.3 Atypical Fibroxanthoma</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>3.4 Kaposi's Sarcoma</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>3.5 Merkel Cell Carcinoma</td>
<td>16</td>
</tr>
<tr>
<td></td>
<td>3.6 Skin Cancer at Particular Anatomical Sites</td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>3.7 Cutaneous Lymphoma and Supra Network MDT</td>
<td>23</td>
</tr>
<tr>
<td></td>
<td>3.8 Angiosarcoma</td>
<td>26</td>
</tr>
<tr>
<td>4.0</td>
<td>Imaging</td>
<td>28</td>
</tr>
<tr>
<td>5.0</td>
<td>Pathology</td>
<td>30</td>
</tr>
<tr>
<td></td>
<td>5.1 North Trent Skin Pathology</td>
<td>30</td>
</tr>
<tr>
<td></td>
<td>5.2 Minimum Data Set Reporting</td>
<td>31</td>
</tr>
<tr>
<td></td>
<td>5.3 Double Reporting</td>
<td>36</td>
</tr>
<tr>
<td></td>
<td>5.4 LSMDT Pathology</td>
<td>38</td>
</tr>
<tr>
<td></td>
<td>5.5 SSMDT Pathology</td>
<td>40</td>
</tr>
<tr>
<td></td>
<td>5.6 Operational Management</td>
<td>46</td>
</tr>
<tr>
<td></td>
<td>5.7 Laboratory investigations</td>
<td>47</td>
</tr>
<tr>
<td></td>
<td>5.8 Audit</td>
<td>54</td>
</tr>
<tr>
<td></td>
<td>5.9 External Quality Assurance (EQA)</td>
<td>55</td>
</tr>
<tr>
<td>6.0</td>
<td>Palliative Care</td>
<td>57</td>
</tr>
<tr>
<td>7.0</td>
<td>Teenager and Young Adults</td>
<td>59</td>
</tr>
</tbody>
</table>
# 1.0 Network Configuration of Teams

<table>
<thead>
<tr>
<th>Name of MDT</th>
<th>Type of MDT Level of Care provided</th>
<th>Host Organisation</th>
<th>Hospital Contact Point</th>
<th>Referring PCT</th>
<th>Catchment Population</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barnsley Skin MDT</td>
<td>Local Level 1,2,3,4</td>
<td>Held at Barnsley Hospital NHS Foundation Trust</td>
<td>01226 432000</td>
<td>Barnsley Primary Care Trust</td>
<td>231,551</td>
</tr>
<tr>
<td>Chesterfield Skin MDT</td>
<td>Local Level 1,2,3,4</td>
<td>Held at Chesterfield Hospital NHS Foundation Trust</td>
<td>01246 516126</td>
<td>Derbyshire County Primary Care Trust (excludes High Peak &amp; Dales)</td>
<td>363,056</td>
</tr>
<tr>
<td>Doncaster &amp; Bassetlaw Skin MDT</td>
<td>Local Level 1,2,3,4</td>
<td>Held at Doncaster &amp; Bassetlaw NHS Foundation Trust</td>
<td>01302 381495, or 01302 647207</td>
<td>Doncaster Primary Care Trust Bassetlaw Primary Care Trust</td>
<td>293,143</td>
</tr>
<tr>
<td>Rotherham Skin MDT</td>
<td>Local Level 1,2,3,4</td>
<td>Held at The Rotherham NHS Foundation Trust</td>
<td>01709 307162</td>
<td>Rotherham Primary Care Trust</td>
<td>243,889</td>
</tr>
<tr>
<td>Sheffield Skin MDT</td>
<td>Local Level 1,2,3,4,5, Specialist</td>
<td>Held at Sheffield Teaching Hospital Foundation Trust</td>
<td>0114 226 1394</td>
<td>Sheffield Primary Care Trust All above</td>
<td>534,251</td>
</tr>
</tbody>
</table>

**Total:** 1,772,483
1.1 Network Configuration of Skin cancer in the Community (April 2009) (08-1A-205j)

The Network Board has agreed with the PCT’s, the NSSG and the cancer lead clinicians of each trust in the North Trent Cancer network, the Networks configuration for community skin cancer services which specifies which of the three models of service provision each PCT will commission and the names of the relevant MDT’s, GPwSIs and location of any community facilities.

Within NTCN this process of providing skin cancer services in the community is still been developed and the table showing the configuration includes practitioners who will be accredited according to the NSSG accreditation process and are not practising as GPwSI according the peer review measures at present.

<table>
<thead>
<tr>
<th>MDT location</th>
<th>Proposed Service Model</th>
<th>Location community facilities</th>
<th>Current Service Model</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barnsley NHS FT Hospital (LSMDT)</td>
<td>1</td>
<td>Dr J Peace</td>
<td>No</td>
</tr>
<tr>
<td>Chesterfield Royal Hospital (LSMDT)</td>
<td>1</td>
<td>Dr L Moss</td>
<td>No</td>
</tr>
<tr>
<td>Doncaster and Bassetlaw Hospitals NHS Trust (LSMDT)</td>
<td>0</td>
<td>No</td>
<td>N/A</td>
</tr>
<tr>
<td>Rotherham NHS FT Hospital (LSMDT)</td>
<td>0</td>
<td>No</td>
<td>N/A</td>
</tr>
<tr>
<td>Sheffield Teaching Hospitals NHS FT (SSMDT)</td>
<td>0</td>
<td>No</td>
<td>N/A</td>
</tr>
</tbody>
</table>

*To be accredited by the NSSG accreditation process.*
2.0 Primary Care Referral Guidelines

Primary Care Practitioners should refer patients defined as ‘urgent, suspicious of cancer’ using the criteria of the NICE Referral Guidelines on the agreed network forms to the relevant contact point of a single named local skin MDT as defined below (the network is currently moving towards electronic choose and book referral for all patients). (08-1C-113j)
See Appendix 1 for NTCN 2 week wait referral form for skin cancer (hard copy)

Patients should be referred in accordance to defined National standards. The NSSG agrees that patients should be referred in accordance to these levels of care.

The levels of care as specified in the Manual for Cancer Services (Skin Measures) 2008 are given in the Terms of Reference document (figure 2) and these identify where care should be provided.

**Level 1 care**  Actinic keratoses and precancerous lesions may be treated by any GP.

**Level 2 care**  GPs should refer suspected cases of skin cancer requiring treatment including BCC to the local contact point according to the appropriateness of the lesion and the availability of local services. Level 2a BCC’s can be referred to community practitioners approved to work to the ‘DES/LES’ model and level 2b BCC’s to ‘model 1’ approved community practitioners, see NICE guidance on cancer services: skin tumours including melanoma (update 2011) and Constitution for the skin NSSG may 2012.

**Level 3 Care**  Requires mandatory treatment care under a hospital doctor who is a core member of the local MDT. This includes both high risk BCC and SCC.

### Clinical features of High risk BCC’s (any of these)

<table>
<thead>
<tr>
<th>Site</th>
<th>Face (specifically nose and lips including nasofacial sulci and nasolabial folds or around eyes) scalp, around ears</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size</td>
<td>1 cm or more if above clavicle 2 cms or more if below clavicle</td>
</tr>
</tbody>
</table>
| Circumstances | Immunocompromised patient  
Genetically predisposed(Gorlins)  
Recurrent or persistent previously treated lesion  
Flat lesion, hard thickened skin (morphoeic), infiltrative or baso-squamous appearance  
Under 24 years |
Lesions biopsied in Primary Care that prove to be Malignant should be referred urgently under the 2 Week Wait rule.
A copy of the Histology report should be either faxed with the referral or accompany the patient to the clinic appointment. If a photograph of the pre biopsy lesion was taken this should also accompany the patient. These patients will be discussed at the local MDT and future treatment plans communicated back to the GP after discussion of treatment options with the patient.

**Level 4**
It is agreed that Level 4 care will be provided by the Local Skin MDT and that the LSMDT which will provide mandatory individual case review (the SSMDT and its core members acting as LSMDT) for the following cases:

- High risk BCC
- SCC
- Malignant Melanoma- new, single primary, adult, non-metastatic, not for approved trial entry, up to and including stage IIa (must fulfil all these criteria)
- Radiotherapy if attendance by Clinical Oncologist at LSMDT
- Lesion where diagnosis uncertain but maybe malignant
- Incompatible clinical and histological findings

**Level 5**
It is agreed that all LSMDTs (Barnsley, Chesterfield, Doncaster and Bassetlaw and Rotherham) within NTCN refer all cases of the types of skin cancer needing care level 5 as specified by the skin cancer measures to the SSMDT based in Sheffield for discussion and management. This will include new cases of:

- Selected BCCs and SCCs needing plastic/reconstructive surgery by SSMDT core member
- Radiotherapy (if not discussed by LSMDT clinical oncology core member)
- Metastatic SCC on presentation or newly metastatic
- Malignant Melanoma stage IIb or more, or < 19 years or metastatic on presentation or newly metastatic or recurrent or for approved trial entry.
- Any cases for approved trial entry
- Any cases for adjuvant therapy
- Histology opinion from SSMDT core pathology member
- Mohs Surgery
- Skin cancer in immunocompromised patients including organ transplant recipients
- Skin Cancer in genetically predisposed patients including Gorlins Syndrome
- Cutaneous lymphoma
- Kaposi's sarcoma
- Cutaneous sarcoma above superficial fascia
- Other rare skin cancers
3.0 Clinical Guidelines

3.1 Basal Cell Carcinoma/Squamous Cell Carcinoma/Malignant Melanoma

The network follows the national guidance given by the BAD on BCC, SCC, Melanoma and Cutaneous T Cell Lymphoma.

References

Appendix 2 – (hard copy)
Guidelines for the management of basal cell carcinoma.

Appendix 3 – (hard copy)
Multiprofessional guidelines for the management of the patient with primary cutaneous squamous cell carcinoma.
R. Motley, P. Kersey, C. Lawrence.

Appendix 4 – (hard copy)
U.K. guidelines for the management of cutaneous melanoma 2010
D.L.L.Roberts, A.V.Anstey, R. J.Barlow, N.H.Cox, J.A.Newton Bishop,
P.G.Corrin, J .Evans, M.E.Gore, P.N.Hall, N.Kirkham.
3.2 Dermatofibromasarcoma Protuberans

3.2.1 Clinical behaviour
DFSP is a low grade sarcoma probably of fibroblast origin. Thought to arise in the dermis and invade deeply. Population incidence is approx 1 case per 4 million per annum. Complex, irregular 3D tumour structure explains high local recurrence rates. The risk of regional spread is approx 1% and metastatic spread 5%. DFSP is slow growing. Diagnosis is often delayed. It may start as a papule gradually enlarging into a nodule or plaque. Colour varies from pink to red to violaceous. Plaques may ulcerate. DFSP most commonly occurs on the trunk, followed by the proximal extremities. It rarely occurs above the neck.

3.2.2 Diagnosis and investigation
Suspicious lesions should be referred to the SSMDT. Initial core or incision biopsy is recommended. Equivocal histology may require further biopsy. Imaging is not recommended as the risk of metastasis is low.

3.2.4 Level of care
Suspected or biopsy proven DFSP above superficial fascia should be referred to the SSMDT.

3.2.5 Treatment
Resection is the first line treatment, margins can be controversial. Most authorities recommend a 3 cm clearance, down to and including fascia, if possible.

Five year recurrence free survival rates increased with increasing margins of resection:
• 59 % less than 1 centimetre clearance.
• 66 % greater than or equal to 1 centimetre clearance.
• 70 % greater than or equal to 2 centimetre clearance.
• 80 % greater than or equal to 3 centimetre clearance.

A 4cm macroscopic margin is said to give clearance of 95% for DFSP.

Mohs’ surgery may further improve outcome (local recurrence rate 6.6%) vs. Wide excision (local recurrence rate 10%).

If there is doubt about resection margins, split skin graft will allow monitoring of the bed.
Additional treatments.

Radiotherapy
Should be considered when complete excision is not obtainable or when resection of metastatic disease is not feasible.

Systemic treatment
Imatinib is approved in the UK for “treatment of adult patients with unresectable dermatofibrosarcoma protuberans (DFSP) and adult patients with recurrent and/or metastatic DFSP who are not eligible for surgery.” Patients being considered for
systemic therapy should be discussed with the sarcoma MDT, as this is where experience with Imatinib lies.

Follow-up:  
Year 1 – 3 monthly review  
Year 2 – 3 monthly review  
Year 3 – 3 monthly review  
Year 4 – 6 monthly review  
Year 5 – 6 monthly review  
Consider annual review thereafter

A history & examination for local recurrence and regional lymphadenopathy is appropriate but further investigation is not warranted unless metastatic disease (lung is the commonest) is suspected.

Because of the indolent nature of DFSP the patient should be coached regarding lifelong surveillance.

References

www.nccn.org/professionals/physician_gls/PDF/dfsp.pdf
3.3 Atypical Fibroxanthoma

3.3.1 Clinical
Most authorities would hold that atypical fibroxanthoma (AFX) is a dermal lesion with a favourable prognosis. It most commonly occurs on the face, nose, ears & scalp of elderly individuals and may be linked to ultraviolet radiation. It is more frequent in males (70% men; 30% women), average age 70. AFX is a nodular or plaque like erythematous or tan coloured dermal lesion which typically shows rapid growth. Lesions are usually less than 2 cm in diameter and half the cases are ulcerated at presentation. It is also reported in younger individuals in non exposed sites of trunk and limbs and also immune suppressed transplant patients. Currently there is debate as to whether it is a benign or malignant tumour. It has been reported to metastasise but there is argument that these are cases of pleomorphic malignant fibrous histiocytoma (MFH).

3.3.2 Pathology
Most AFX cases (80%) are restricted to the reticular dermis. The lesion may extend into the upper one-third of the subcutaneous adipose tissue. Tumours that extensively involve subcutis, penetrates fascia, or displays necrosis, perineural or lymphovascular invasion should be diagnosed as MFH. Cell types include spindle, epithelioid, multinucleate and bizarre cells, in various proportions, arranged in haphazard, vaguely fascicular or storiform patterns with hyperchromatic and multilobulated nuclei.

3.3.3 Treatment
There are no widely agreed recommendations for the treatment of AFX. In the absence of clear guidelines a 10mm clearance in the first instance would be suggested. If this does not obtain clearance wider excision would be recommended, Moh’s surgery should be considered.

Levels of Care

Cases of AFX should be treated as for high risk SCC patients as per the peer review measures and should be referred to the SSMDT

3.3.4 Follow-up
There are no widely agreed recommendations for follow-up. In the absence of clear guidelines:

Year 1 – 3 monthly review
Year 2 – 3 monthly review
Year 3 – 3 monthly review
Year 4 – 6 monthly review
Year 5 – 6 monthly review

References

Atypical Fibroxanthoma A.SAKAMOTO.Clinical Medicine..Oncology 2008; 2: 1192129
3.4 Kaposi’s Sarcoma

3.4.1 Introduction
Kaposi’s sarcoma (KS) is a tumour caused by Human Herpes Virus 8 (HHV8). Although there are different clinical subtypes, HHV-8 is common aetiological cause. Not considered a true sarcoma it arises from malignant transformation in lymphatic endothelium.

3.4.2 Classic Kaposi’s Sarcoma
Typically affects elderly men of Mediterranean or Eastern European descent where HHV-8 infection is relatively high.

3.4.3 Endemic Kaposi’s Sarcoma
Typically affects young sub-Saharan Africans. Unrelated to HIV it is more aggressive than classic KS. The most common presentation is with extensive infiltration of skin, especially lower limbs.

3.4.4 Transplant Related Kaposi’s Sarcoma
Occurs in relation to suppressed T-cell function when an HHV 8-infected organ is transplanted or when the recipient already has pre-existing HHV 8 infection.

3.4.5 Epidemic Kaposi’s Sarcoma
Is AIDS related in patients with suppressed T-cell function where HHV 8 is usually sexually transmitted.

3.4.6 Level of care
Optimal care can only be achieved by the close co-operation of oncologists, haematologists and HIV physicians. Professor Woll at Weston Park Hospital has a particular interest in Kaposi Sarcoma.

3.4.7 Clinical features
Kaposi’s Sarcoma lesions are nodular or plaques that may be red, purple, brown, or black. Tumours are vascular and extravasation into surrounding tissues produces its discolouration. Inflammation produces pain and swelling. Lesions are most commonly found on the skin but may occur in gastrointestinal or respiratory mucosa. Rate of progression and mortality varies widely.

3.4.8 Diagnosis
Clinical features and index of suspicion must be confirmed by biopsy. Histological features and the detection of the KSHV protein LANA-1 (latency-associated nuclear antigen) in tumour cells confirms the diagnosis.

3.4.9 Staging
The AIDS Clinical Trials Group (ACTG) 1988 proposed a staging classification for AIDS-related Kaposi sarcoma (KS) that considers 3 criteria:
1. the extent of the tumour (T)
2. the status of the immune system (I), as measured by the CD4 count
3. the extent of involvement within the body or systemic illness (S)

Under each of these major headings, there are 2 subgroups identified by either a zero (0) or a one (1). A zero (0) means good risk, while a one (1) means poor risk.
**T (tumour) status**

**T0 (good risk):** Localized tumour
KS is confined to the skin and/or the lymph nodes. If there are lesions in the mouth, they can only be on the palate (roof of the mouth), and those lesions are flat (not raised or rounded).

**T1 (poor risk):** Disseminated (widespread) tumour. One or more of the following is present:
- oedema due to the tumour
- lesions are ulcerated
- extensive oral KS (nodular lesions and/or lesions not limited to the palate)
- KS is affecting the stomach or intestines
- KS is in the lungs or other internal organs.

**I (immune system) status**

**I0 (good risk):** CD4 cell count is at least 200 cells/ microliter. (Normal range 600-1,500). Some experts use a lower cut-off for good risk such as 150 or 100.

**I1 (poor risk)**
CD4 count is lower than 200 (again some have used lower cut-offs such as 150 or 100).

**S (systemic illness) status**

- **S0 (good risk):** No systemic illness present and all of the following are true:
  - No history of opportunistic infections or thrush (a fungal infection in the mouth)
  - No B symptoms are present. B symptoms are:
    - unexplained fever
    - night sweats
    - weight loss of more than 10% (without trying to lose weight)
    - diarrhoea for more than 2 weeks
    - The patient is doing well - up and about most of the time and he can take care of himself. Karnofsky performance status needs to be at least 70 to be in the good risk category.

- **S1 (poor risk)**
  - Systemic illness is present; and one or more of the following is true:
    - History of opportunistic infections or thrush
    - One or more B symptoms is present
    - Performance status score under 70
    - Other HIV-related illness is present, such as neurological (nervous system) disease or lymphoma.

**Survival**

T and S factors seemed to be the most important.

Ninety percent of KS patients who are at good risk in these categories (T0S0) survive for at least 3 years after diagnosis.

Only fifty percent of the KS patients who are at poor risk in both of these categories (T1S1) are still alive 3 years after diagnosis.

The immune system status when the patient is first diagnosed seems to be less important if HAART is given.

**3.4.10 Treatment**

Kaposi's sarcoma is not currently curable. Treatment is palliative. In KS associated with immunodeficiency or immunosuppression, treating the immune dysfunction can slow or halt progression of KS. In 40% or more of patients with AIDS-associated Kaposi's sarcoma, the Kaposi lesions will shrink upon first starting highly active antiretroviral therapy (HAART). However, in a certain percentage of such patients,
Kaposi's sarcoma may again grow after a number of years on HAART, especially if HIV is not completely suppressed. Patients with a few local lesions can often be treated with local measures such as radiation therapy or cryosurgery. Surgery is generally not recommended as Kaposi's sarcoma can appear in wound edges.

British HIV Association guidelines for HIV-associated malignancies 2008 make the following recommendations

**Early-stage KS (T0 stage)**
HAART.
Consider local radiotherapy or liposomal anthracycline for rapidly progressing or cosmetically disfiguring disease.

**Advanced-stage KS (T1 stage)**
1) HAART and liposomal anthracycline (either DaunoXome 40 mg/m2 every 14 days or Caelyx 20 mg/m2 every 21 days).
2) Anthracycline-refractory KS
3) HAART and paclitaxel (100 mg/m2 every 14 days) (individual case basis).

The South Yorkshire HIV Network
Referral Pathway for Kaposi's Sarcoma in HIV infected patients is:

1. All HIV positive patients presenting with KS like lesions should have the diagnosis confirmed by biopsy. This should also exclude conditions like Bacillary angiomatosis which can may misdiagnosed as KS and which require specific treatment.
2. All KS biopsies should be reviewed by the Network sarcoma MDT lead pathologist (Dr David Hughes, RHH; david.hughes@sth.nhs.uk; 0114 271 3008) and discussed at the Network sarcoma MDT (Thursdays, 1330h, MDT room, J floor, RHH). To list cases for discussion, please contact the MDT co-ordinator (sht-tr.cancer-sarcoma@nhs.net; 0114 271 2513)
3. We are setting up a register and database of HIV positive patients who develop malignancies to facilitate pathway developments across the HIV network, audit and possible research (contact Dr Christine Bowman for details Christine.Bowman@sth.nhs.uk) We therefore recommend registration of all new cases of KS (regardless of whether immediate referral to oncology is indicated). See attached HIV patient Cancer Registration form and Database consent form.
4. The HIV Network Lead Oncologist for KS is Professor Penella Woll who can be contacted by email p.j.woll@sheffield.ac.uk (0114 226 5235). She is happy to discuss the management of any cases of KS.
6. A full assessment of the extent of the KS, the patient’s immune status and general health are required to inform management decisions. This should include careful examination of the entire skin surface and mucosal surfaces of the genitals and mouth for KS lesions and any associated oedema, examination for lymphadenopathy and hepatosplenomegaly, chest X-ray to look for pulmonary involvement (CT scan and bronchoscopy if abnormal or respiratory symptoms), gastrointestinal endoscopy if any features of GI blood loss or significant iron deficient anaemia.
7. All patients with KS should be started on HAART regardless of CD4 count. Choice of ARVs should be informed by viral resistance, standard guidelines and patient choice. There is no clinical data to support Protease inhibitors being superior to NNRTIs in KS patients.

8. Patients with limited cutaneous disease, slow progression of lesion development, lack of pulmonary, GI or other visceral involvement and CD4 counts >150 may respond adequately to HAART alone.

9. If these patients with limited, slowly progressive KS lesions have local complications eg. leg oedema they may benefit from radiotherapy. The sarcoma MDT radiotherapist is Dr Martin Robinson (WPH Sheffield). Prof Woll is happy to discuss the appropriateness of this option and refer onto radiotherapy if indicated.

10. Patients with extensive cutaneous disease, rapid progression of lesions, evidence of pulmonary, GI or other visceral involvement and/or low CD4 counts (<150) may have a rapidly progressive condition that requires urgent referral to Prof Woll and the sarcoma MDT.

11. Chemotherapy, if indicated, will be managed under the care of Professor Woll at Weston Park Hospital, Sheffield. First line chemotherapy will be Liposomal doxorubicin. If this fails to adequately control the KS the patient may be offered Paclitaxel.

12. See algorithm on page 15
**SOUTH YORKSHIRE HIV CLINICAL NETWORK**

**ALGORITHM FOR MANAGING KAPOSI’S SARCOMA**

Lesions suspicious of KS

↓

Biopsy

↓

KS biopsies should be reviewed by the Network sarcoma MDT lead pathologist david.hughes@sth.nhs.uk and discussed at the Network sarcoma MDT (coordinator sht-tr.cancer-sarcoma@nhs.net)

↓

Register KS cases with SYHCN Cancer Register and Database Contact Christine.bowman@sth.nhs.uk

↓

Assess patient for extent of muco-cutaneous KS, associated local oedema or ulceration, symptoms and signs of pulmonary or gastrointestinal involvement. Determine general health status and degree of immunocompromise

↓

If symptoms or signs of visceral involvement proceed to CT scan / bronchoscopy/ GI endoscopy as appropriate

↓

Start patient on HAART

↓

Discuss case with Prof Penella Woll p.j.woll@sheffield.ac.uk (0114 226 5235).

---

**EARLY STAGE KS**

- Cutaneous lesions
- Minimal oral involvement
- Lymph nodes may be involved
- CD4 count >150
- HAART alone
- Consider radiotherapy or chemotherapy if associated oedema or cosmetically disfiguring

**LATE STAGE KS**

- Extensive or rapidly progressing mucocutaneous lesions
- Pulmonary KS
- Extensive gastrointestinal involvement
- Low CD4<150
- HAART plus early referral for chemotherapy:
  - Liposomal doxorubicin
  - Paclitaxel if 1st line fails to control KS

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**3.4.11 Clinical Trials**

Professor Woll’s team at Weston Park Hospital are leading a national trial of a novel treatment (selumetinib) for HIV-associated KS – SCART

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**References**


South Yorkshire HIV Network guidelines for Kaposi’s sarcoma. Dr Christine Bowman & Professor P Woll, 2009
3.5 Merkel Cell Carcinoma

3.5.1 Key Points

- Merkel cell carcinoma (MCC) is a rare and aggressive form of skin cancer that can grow rapidly and metastasise (spread) to regional lymph nodes and/or other distant sites.
- MCC often presents as a smooth, painless, firm lump located in, or just beneath, the skin. It can appear pink, blue or the same colour as the surrounding normal skin, and most commonly occurs in sun-exposed skin.
- Any patient who presents with a lesion that has features suspicious for MCC should be referred urgently for assessment by a core member of the Specialist Skin Multi-Disciplinary Team (SSMDT).
- Any patient found to have a histological diagnosis of MCC following biopsy of a lesion should be referred urgently for assessment by a core member (dermatologist or plastic surgeon) of the SSMDT.

INTRODUCTION

MCC is a rare form of skin cancer that is thought to arise within neuroendocrine cells in the skin. Hence, it is sometimes known as primary cutaneous neuroendocrine carcinoma. MCC mainly affects adults over the age of 50 years (95% of cases) and is more common in white males than in other groups. It usually arises in sun-exposed skin, presenting as a painless, smooth lump that is firm and appears within or just beneath the skin. The appearance can vary in colour from pink to blue to normal skin colour. It is an aggressive cancer that can grow rapidly, recur locally following excision and metastasise to regional lymph nodes and/or distant sites, such as the lungs, liver and brain. It is because of these features that the management of all patients with MCC should be carried under the guidance of a SSMDT.

LEVEL OF CARE

Any patient who presents with a lesion that has the appearance of MCC should be referred urgently to a core member of the SSMDT who is able to confirm the diagnosis and initiate appropriate treatment (dermatologist or plastic surgeon).

Any patient who has undergone biopsy of a lesion that is subsequently found to be an MCC should be referred urgently to a core member of the SSMDT who is able to perform a wide local excision (plastic surgeon) or Moh’s surgery (dermatologist).

Any patient who presents with metastatic MCC should be referred for discussion at the SSMDT meeting and assessment by a core member of the SSMDT (oncologist, plastic surgeon, and dermatologist).
MANAGEMENT

There are currently no UK national guidelines for the management of MCC. Guidelines have been produced for MCC in the USA by the National Comprehensive Cancer Network, as part of the NCCN Clinical Practice Guidelines in Oncology (www.nccn.org). In general, the evidence base for these guidelines consists of lower level studies rather than randomised controlled trials and meta-analyses. The recommendations contained within the present guidelines arose from interpretation of such existing guidelines within the context of current regional practices and available resources. The guidelines are intended to illustrate the standard practice for the management of MCC within Trent region. However, it is acknowledged that there will be occasions when it is appropriate for the management of a patient to follow a course that differs from that outlined in these guidelines. This should generally only occur following consideration of the case by the SSMDT.

Investigation

Primary disease:

Patients presenting with primary lesion suspicious for MCC or following diagnostic biopsy of primary lesion.

- Perform diagnostic biopsy (preferably excision biopsy) if not already performed to obtain histological diagnosis. Histology slides from biopsies performed outside the remit of the SSMDT should be forwarded for review by a consultant histopathologist who is a core member of the SSMDT.
- In addition to routine haematoxylin and eosin (H&E) staining, histological analysis should involve immunohistochemistry with an immunopanel preferably containing cytokeratin-20 (CK-20) and thyroid transcription factor –1 (TTF-1), to differentiate MCC from metastatic small cell lung carcinoma, the histologic appearance of which can be very similar.
- Patients should undergo clinical examination of the entire skin to look for concurrent lesions. Clinical examination of regional and distant lymph node basins should be performed to detect nodal metastases and abdominal examination for masses and/or hepatosplenomegaly should be carried out.
- Liver function tests should be performed as a screening test for liver metastases and if significantly deranged, radiological imaging of the liver should be performed.
- Further imaging modalities such as plain chest radiography, computed tomography, ultrasound and positron emission topography should only be performed as clinically indicated.
- If the CK-20 is negative and/or there is clinical suspicion that the lesion may not be a primary tumour, investigations should be considered to evaluate for a non-cutaneous primary carcinoma, such as small cell lung carcinoma.
- If the lesion is believed to be a primary MCC and there is no evidence of metastases treatment should follow the guidelines for Stage I disease.
Local recurrence and intransit metastases:
Patients presenting with possible or confirmed metastatic nodal disease.

- Obtain, confirm or review diagnosis of metastatic nodal MCC. Patients with a previous history of MCC who present with a possible recurrence should be referred to the SSMDT or to an appropriate core member who is capable of confirming the diagnosis. Histology slides from biopsies performed outside the remit of the SSMDT should be forwarded for review by a consultant histopathologist who is a core member of the SSMDT.
- A clinical examination should be carried out as for patients presenting with a primary lesion (see above). If there is no previous history of MCC, particular attention should be directed towards detecting a primary lesion.
- FNA (fine needle aspiration) with or without ultrasound imaging/guidance would normally be performed on the clinically suspicious lymph node.
- Histological analysis of the FNA should include an immunopanel containing CK-20 and pancytokeratins.
- If FNA is negative an open excision biopsy of the lymph node should be performed.
- If FNA is positive then further staging investigations may be appropriate to assess the extent of nodal involvement and/or detect distant metastases.
- Once the diagnosis has been confirmed, distant metastasis has been excluded by staging investigations as clinically indicated, and the case has been referred for discussion at the SSMDT meeting, treatment should follow the guidelines for Stage II disease.

Metastatic disease:
Patients presenting with possible or confirmed distant metastatic disease.

- Obtain, confirm or review diagnosis of metastatic MCC. Patients with a previous history of MCC who present with a possible distant metastasis should be referred to the SSMDT or to an appropriate core member who is capable of confirming the diagnosis. Histology slides from biopsies performed outside the remit of the SSMDT should be forwarded for review by a consultant histopathologist who is a core member of the SSMDT.
- A clinical examination should be carried out as for patients presenting with a primary lesion (see above). Particular attention should be directed towards other possible sites of distant metastasis, as guided by the clinical history.
- Appropriate radiological studies should be performed as indicated to aid diagnosis and plan treatment.
- Histological confirmation of the diagnosis may be possible but in some cases this may not be appropriate.

Treatment

MCC can be staged according to the American Joint Committee on Cancer (AJCC) TNM staging classification for merkel cell cancer (7th Ed., 2010)
Local Disease

**Excision of primary tumour**

- Treatment aims to achieve complete excision of the primary tumour demonstrated histologically by clear surgical margins.
- A wide local excision of the primary tumour with 1-2cm margins down to the investing fascia of the muscle or to the pericranium is recommended where possible.
- Excision with margins ≥0.5cm has been shown to be associated with a very high recurrence rate (100%). The local recurrence rate with margins ≥2.5cm appears to be reduced but still remains substantial (49%).
- Moh’s micrographic surgery, modified Moh’s (with permanent section assessment of the final margin) and excision with complete circumferential and peripheral deep-margin margin assessment are all accepted techniques for excision of MCC.
- The extent of the role for Moh’s micrographic surgery for primary MCC excision remains unclear. One small study (12 patients) found the rate of recurrence to be similar to that of wide local excision (50%). However, in cases where the tumour is in an anatomically sensitive location Moh’s surgery should be considered as an alternative to wide local excision.

**Reconstruction of defect following excision of primary tumour**

- The choice of reconstruction for the defect resulting from wide local excision will be influenced by a variety of factors including the level of certainty that excision is complete, the general health of the patient and cosmetic concerns.
- Primary closure will usually be performed if possible.
- When primary closure is not possible a split skin graft should be considered as an option which provides thin skin cover and thus facilitates monitoring for local recurrence.
- If reconstruction using local or distant flaps is to be performed it is recommended that the procedure be delayed until histological confirmation of clear surgical margins has be received.

**Sentinel node biopsy**

- Sentinel node biopsy should be considered for all patients who present with a primary MCC without clinical evidence of metastasis, and should be performed at the time of wide local excision.

**Radiotherapy to the primary tumour site**

- For small tumours that are completely excised, and in the absence of other adverse risk factors, observation only should be considered.
Radiotherapy to the primary tumour site should be considered for large tumours, tumours with microscopically involved margins following wide local excision or macroscopically involved margins where complete excision is not possible. Radiotherapy to in-transit lymphatic and draining nodal basins should be considered for patients with head and neck primary tumour sites.

Chemotherapy
- There are currently no indications for chemotherapy for local disease.

Regional disease
- The treatment options for patients with histologically-confirmed metastatic MCC in regional lymph nodes include lymph node dissection, radiotherapy or both and the decision should be tailored to the individual patient following discussion at the SSMDT.

Radiotherapy to in-transit lymphatics and/or draining nodal basins
- Radiotherapy should be considered for patients at high risk for regional recurrence and for patients with confirmed nodal metastases who are not undergoing lymph node dissection.

Following lymph node dissection, post-operative radiotherapy is indicated for patients with multiple involved nodes or extra capsular spread. Chemotherapy
- Chemotherapy has not been shown to improve disease-free or overall survival for patients with regional disease and is therefore not recommended outside clinical trial situations.

Metastatic Disease
- Treatment options should be discussed at SSMDT should be tailored for the individual patient and may include best supportive care with or without surgery, radiotherapy and chemotherapy alone or in combination.

Follow-up

The recommended follow-up schedule is the same for all patients with MCC irrespective of the stage of their disease.

Year 1 and 2: Review every 3-6 months
Year 3 onwards: Review every 6-12 months.

References

www.nccn.org/professional/physician_gls/PDF/mcc.pdf
3.6 Skin cancer at particular anatomical sites

3.6.0.1 Management of patients with skin cancer may require knowledge and technical expertise which is best provided by practitioners who are not members of the skin MDT. In these cases, patients will be referred to the appropriate MDT or individual(s) explicitly for advice or practical input. Advice should be fed back promptly to the SSMDT and the SSMDT should be informed promptly about a patient’s progress and be provided with histopathology results so that the SSMDT can monitor the patient’s progress, provide necessary advice & input and maintain records.

3.6.1 Perianal Skin Cancer [08-1A-213]
Patients with invasive perianal skin cancers should be referred to the anal cancer/colorectal MDT for advice and if necessary, management. Management of perianal skin cancer patients requiring reconstructive surgery should normally involve Specialist Skin cancer MDT (SSMDT) core members. Histology and operative findings should be reviewed by the SSMDT.

SSMDT review of management is recommended for all patients with perianal melanoma.

3.6.2 Head & Neck Skin Cancer [08-1A-212]
Patients with head & neck skin cancer should be managed by the Local Skin cancer MDT (LSMDT) or Specialist Skin cancer MDT (SSMDT) in accordance with levels of care defined in the skin peer review measures 2008 except when there is involvement of cervical lymph nodes or the parotid gland or extensive uncontrolled disease.

Patients with skin cancer involving cervical lymph nodes or the parotid gland should be discussed at the SSMDT and Head & Neck MDT and subsequent surgery should be carried out by a surgeon with suitable current experience and currency, at the Royal Hallamshire Hospital. The Specialist Head and Neck MDT is based in Sheffield.

Patients with peri-ocular skin cancer should be referred to a surgeon who is a core member of a skin MDT.

Patients with known or suspected ocular mucosal melanoma should be referred to the ophthalmology oncology MDT and the SSMDT.

Management of all patients with cutaneous melanoma of the head & neck should be reviewed by the LSMDT or SSMDT in keeping with levels of care defined in the Skin Peer Review measures 2008. Cases of cutaneous eyelid melanoma managed by the ophthalmology oncology MDT are not excluded from this requirement.

3.6.3 Urogenital Skin Cancer [08-1A-215]
Patients with invasive male genital skin cancers should be referred to the urology MDT which will refer appropriate cases on to the Supra-network penile cancer MDT in Leeds. SSMDT review of management is recommended for penile melanoma cases in addition to the Supra-network penile cancer MDT.

3.6.4 Vulval Skin Cancer [08-1A-214]
Patients with invasive vulval skin cancer should be referred to the Gynaecology MDT for discussion. Management of vulval skin cancer patients requiring reconstructive
surgery by Specialist Skin Cancer MDT (SSMDT) core members should be reviewed by the SSMDT. SSMDT review of management is recommended for all patients with vulval melanoma.

3.6.5 Haemato-oncology [08-1A217j]
1. Patients presenting with skin lesions associated with known or suspected systemic haemato-oncological malignancy should be reviewed by the haemato-oncology MDT before starting definitive treatments such as radiotherapy, chemotherapy or photophoresis.

2. Patients presenting in any speciality with known or suspected primary cutaneous lymphoma should be reviewed by the Specialist Skin Cancer MDT (SSMDT) before starting definitive treatments.

3. Patients discussed at the haemato-oncology MDT who transpire to have primary cutaneous lymphoma should be referred to the SSMDT before starting definitive treatments.

3.6.6 Sarcoma [08-1A-217j]
After SSMDT review, patients with cutaneous sarcomas that involve or penetrate the superficial fascia or cutaneous sarcomas potentially requiring radiotherapy or chemotherapy should be referred to the sarcoma MDT based in Sheffield. See the relevant subsection of this guidance for specific types of sarcoma.
3.7 Cutaneous Lymphoma

3.7.1 Local Skin Cancer MDT

All new cases of cutaneous lymphoma to be referred from LSMDT (care level 4) to SSMDT (care level 5)

Referral Pathways:

i). Clinical referral (Management referral of special groups)

Referral pathways of these patients into the SSMDT will be via the agreed pro-forma. It is helpful to include sufficient clinical information and possible digital photographs. Photographs should normally be sent at the time of referral unless there are medical reasons why this is not possible.

And/or

ii). Pathology referral:

Dr David Slater is the present named lead for cutaneous lymphoma for the Network and this work is carried out under his leadership. The other SSMDT Pathologist, however, will also participate in this work for the following reasons:
   a) for purposes of leave cover;
   b) to ensure continuous exposure and sharing of expertise amongst the group of Pathologists involved;
   c) for succession planning and continuity of service in the future.

3.7.2 Specialist Skin Cancer MDT [08-1A-207j]

Sheffield Teaching Hospitals Foundation Trust SSMDT is the only named MDT to manage cutaneous lymphoma within North Trent Cancer Network.

All LSMDTs refer their patients for level 5 care to the Specialist Skin MDT in Sheffield.

The IOG requires that all cases should be reviewed by a dermatopathologist designated by the Network to have an interest and lead responsibility for cutaneous lymphoma reporting (within the requirements of 3.7.1 ii)

When updated national clinical guidelines are available, the content will be incorporated within the guidelines for North Trent Cancer Network.

Pathology guidelines for diagnosis and assessment (SSMDT Level)

All lymphoma patients should undergo diagnostic biopsies for histology, immunophenotyping and molecular studies and this should be correlated with clinical presentation for accurate diagnosis and prognosis.

It is recommended that the WHO primary cutaneous lymphoma classification should be used to classify cases.
Management

The IOG states (page 119) that all patients should be seen by and managed by the SSMDT. It should be understood and expected that any case referred from the LSMDT to the SSMDT for discussion may be taken on for treatment by the SSMDT without further permission from the referrers.

All cases to be referred to SSMDT for histopathology review, clinical review and management advice. There will be flexibility on whether cases are referred back to LSMDT for management, managed by clinicians within the SSMDT or referred to the Supranetwork SSMDT for TSEB or photopheresis.

LSMDTs should be involved once the diagnosis and staging has been confirmed by the SSMDT. Patients with lymphomatoid papulosis and stage 1a mycosis fungoides could be managed locally by the LSMDT after diagnosis is confirmed.

Initial staging imaging is required on all patients with the exception of stage 1 mycosis fungoides and lymphomatoid papulosis.

All patients with Stage IIB-Iv MF/SS and rare CTCL variants to be reviewed by the Supranetwork SSMDT for diagnostic confirmation/management plan to be implemented by Network SSMDT unless specific therapies/trials are only available in the supranetwork centre

Patients with early stage MF (IB-IIA) who are resistant to skin-directed therapies should be reviewed by the Supranetwork SSMDT as at risk of disease progression.

All cases of possible primary cutaneous lymphoma with a lack of diagnostic consensus at Network SSMDT should be reviewed by the Supranetwork SSMDT

3.7.3 Haemato-oncology Links

Patients presenting in any speciality with skin lesions associated with known or suspected systemic haematopoietic malignancy should be reviewed by the Network Haemato-oncology MDT before starting definitive treatments such as radiotherapy, chemotherapy or photopheresis.

Patients discussed at the Network Haemato-oncology MDT that transpire to be primary cutaneous lymphoma should be referred to the Network SSMDT before starting definitive treatments.
Referral Pathway

Patients regarded by the Network SSMDT to require staging will be referred to the North Trent Haemato-Oncology Network MDM meeting via the patient’s clinical consultant or the MDM co-ordinator. When necessary this could involve SSMDT core membership attendance (in particular dermatologist/pathologist)

Referral to the Supranetwork SSMDT will be by the NetworkSSMDT.

Supranetwork SSMDT

Incorporated into this framework, when necessary, is

- Case discussion between SSMDT and Haemato-oncology diagnostic service (HODS) pathologists and when necessary attendance at the North Trent Haemato-oncology Network MDM

3.7.4 Supranetwork MDT for Total Skin Electron Beam Therapy [08-1A-209]

A Supranetwork SSMDT has now been established to review the diagnosis and management of patients with skin lymphoma from and Hull and Yorkshire Coast Cancer Network (HYCCN), North Trent Caner Network (NTCN) and Yorkshire Cancer Network (YCN). This allows pooling of expertise in this relatively rare cancer and is compliant with the Improving Outcomes Guidance for Skin Cancer, being based in Leeds where the Total Skin Electron Beam Therapy service is provided. Further details on the Supranetwork MDT and it pathway can be found in Appendix 8 (hard copy).

3.7.5 Photopheresis [08-1A-210]

Cases of erythrodermic cutaneous T-cell lymphoma, stages 3 and 4, having both skin involvement and circulating T-cell clones should be discussed with the clinician in charge of the named photopheresis facility for potential referral and treatment by photopheresis.

The Skin Cancer NSSG agrees that the consultant responsible for photopheresis should discuss all cutaneous lymphoma cases referred for potential photopheresis at an MDT meeting prior to treatment.

Dr Peter Taylor, Consultant Haematologist, Rotherham Hospitals NHS Foundation Trust is the named Consultant within North Trent Cancer Network for Photopheresis treatment.

Potential cases will be discussed at the Network SSMDT and then referred to the Haemato-oncology Network MDT meeting.
3.8 Angiosarcoma

3.8.1 Angiosarcoma

Patients with Angiosarcoma should be referred to the sarcoma MDT, which is leading a national trial of axitinib treatment in angiosarcoma – Axi-STS. A laboratory project associated with the sarcoma MDT is also in progress.
4.0 Imaging Guidelines

Introduction:

The majority of skin cancers do not require imaging. There are no national guidelines available, and in their absence, this document aims to optimise imaging of skin cancer in the North Trent Cancer Network. The use of imaging is in line with Revised UK management guidelines for the management of cutaneous melanoma 2010.

Skin cancer imaging is usually used to determine either the possible extent of local invasion and/or detect distant metastases.

Local disease:

When there is clinical concern with regards to the possible extent of local invasion, the choice of imaging will depend on the site of the lesion. This aspect of imaging may vary depending on local radiological expertise.

MRI for local extent of skin cancer

When local disease is extensive or when it is close to vital structures, MRI scanning can be very useful. It has superior soft tissue contrast especially in the head and neck region. In the head and neck region, a neck node screen should be part of this examination.

CT for local extent of skin cancer

MDCT can be use as an alternative to MRI scans in some patients who have difficulty with MRI. CT is better at detecting cortical bone destruction.

For patients who are being considered for a nodal block dissection, a CT scan of the chest, pelvis and abdomen should be considered before the procedure.

Ultrasound for local extent of skin cancer

Ultrasound can provide useful imaging information with regards to the local extent of disease. The information obtained often saves the need for performing further complex imaging.

A high-resolution ultrasound scanner with a linear 7 – 12 MHz probe is often used for this examination.

Metastatic skin cancer

This group of skin cancers includes high-grade squamous cell carcinoma and melanoma.
MDCT

MDCT scan is commonly used in most centres for the evaluation of metastatic skin cancer. The areas scanned will depend on the site of the primary lesion and local lymph nodes.

PET / PETCT

PET / PETCT can be used to exclude metastatic melanoma in cases where there is equivocal diagnostic imaging and in cases where surgical options are affected by the possible presence of metastatic disease.

Ultrasound

High-resolution ultrasound has very good soft tissue contrast and when combined with FNAC can have high accuracy in detecting metastatic lymphadenopathy.

Cutaneous Lymphoma

All patients newly diagnosed with cutaneous lymphoma except cases of stage I mycosis fungoides should undergo staging CT scans of thorax, abdomen and pelvis (Whittaker et al, 2003) The neck should also be included in the CT scan, particularly if the lesions are on or above the thorax.

References

S.J.WHITTAKER, J.R.MARSDEN,* M.SPITTLE AND R.RUSSELL JONES
5.0 Pathology Guidelines

5.1 North Trent Skin Pathology

The NSSG should agree network-wide pathology guidelines for the diagnosis and assessment of skin cancer.

It is agreed that there should be equity of access to that all tissue samples are reviewed in high-quality histopathology services. Accurate diagnosis in dermatopathology depends on clinicopathological correlation, involving input from both clinician and pathologist. Although this can be achieved in difficult cases by inter-specialist discussion or seeing the patient records, in some instances (such as cutaneous lymphoma) it may be essential for the patient to be seen jointly. Accordingly, for good clinical governance, it is recommended that the histopathology reporting of any specimens likely to be considered by a skin cancer MDT should be undertaken in a laboratory having easy access to relevant clinicians, patient records and the attending patient.

Histopathology services for skin cancer should be part of managed pathology network or equivalent model. Histopathology reporting should be provided by histopathologists who participate in EQA. This may be a general histopathology EQA scheme that includes skin or a more specialist skin EQA scheme. When appropriate the EQA scheme should cover lymphoma. Given the overlap with head and neck cancer services, it should be noted that the head and neck histopathology EQA scheme includes skin cancer.

All histopathology reports relating to skin cancers should conform to the Royal College of Pathologists minimum datasets on cancer in order to provide adequate and appropriate information on prognosis, planning individual patient treatment, supporting epidemiology and research and to evaluate clinical services and support clinical governance.

Pathology reports should include all the information required by the current Royal College of Pathologists minimum dataset for the relevant cancer. The histopathologists engaged in skin cancer diagnosis should participate in an appropriate external quality assessment (EQA) scheme and demonstrate evidence of continuing professional development (CPD) relevant to skin cancer. The lead histopathologist should attend over 50% of MDT meetings. Other histopathologists reporting skin cancer should be able to demonstrate some MDT activity.

Histopathology services for skin cancer should be part of a managed pathology network or equivalent model.

The Network agrees to implement the following:-

1. All skin cancers to be reported according to the RCPath minimum datasets.
2. Any histopathologist engaged in skin cancer reporting should show evidence of participation in EQA’s, MDT attendance and demonstrate CPD evidence relevant to skin cancer.
3. All severely dysplastic naevi and malignant melanomas are to be double reported.

4. Specialist MDT to review skin cancer cases of greater risk or rarity. These cases usually represent advanced stages of a disease or those that are difficult or complex. Mandatory referral of such cases is indicated from the LSMDT to the SSMDT.

5. General EQA participation which includes skin for any pathologist involved in local MDT (LSMDT).

6. National Specialist Dermatopathology EQA participation for any pathologist involved in reviewing cases for the specialist MDT (SSMDT).

7. All newly diagnosed cutaneous lymphomas should be seen and managed by the SSMDT which should include a dermatopathologist with expertise in cutaneous lymphoma. There will be a named histopathologist for the Network who will act as the lead in cutaneous lymphoma. Cases of cutaneous lymphoma will be dealt with by specialist dermatopathologists/histopathologists working at the SSMDT level, with a low threshold for discussing cutaneous lymphoma cases with the named lead.

8. All skin cancers are staged as per AJCC 7 (effective from Jan. 2012)

5.2 Minimum Data set Reporting

All skin cancers are reported based on the guidelines issued by the Royal College of Pathologists dataset for the histological reporting of common skin cancers, issued in February 2002. It would be preferable for all common skin cancer reporting to be pro-forma based. Gross sampling of skin cancers should be in accordance to those advocated in the dataset in the chapter of handling of gross skin specimens. The dataset has been locally modified to take into consideration recommendations from the skin IOG and peer review measures. Template pro-forma reporting (which can be done by dictation or by completing the paper version) is encouraged for all common skin cancers so that the following can be achieved.

a) All dataset items as agreed by the network is recorded for each case

b) In order to provide adequate and appropriate information on prognosis, planning individual patient treatment, supporting epidemiology and research, to evaluate clinical services and support clinical governance

c) To ensure clarity of communication with service users

Template pro-forma reporting is recommended as a standard practice for reporting common melanoma and non-melanoma skin cancers. The pro-forma used depends on the one accepted by individual laboratories but the network wide agreed datasets relevant to each cancer should be included.
The pro-forma should include relevant macroscopic and microscopic items. The use of the pro-forma is maximised in this way with the aim to condense all relevant information necessary for clinical management of the patient.

Nearest excision margins measured to the nearest mm is recommended, however if this is less than 1 mm, measurement to the nearest tenth of a mm is advisable.

The gross maximum tumour diameter is provided (usually this is the overall maximum tumour diameter). However, rarely the microscopic tumour diameter is greater, and in this case, it is more useful to record this measurement.

The network agreed to adopt the new AJCC 7 staging system effective from 01 Jan. 2012.

Three different staging systems are used for skin cancers in the new AJCC 7. These are:

1. Non-melanoma, non-merkel cel carcinomas including adnexal carcinomas, Therefore the proforma should be the same for all these tumours.

2. Malignant Melanoma

3. Merkel cell carcinoma

Within the network, a separate proforma for BCC with less parameters have been agreed upon for ease of use.

a) For BCC, the following items should be recorded in the pro-forma.

1. Diagnosis
2. Type of biopsy
3. Maximum gross tumour diameter
4. Growth patterns – only the common patterns are outlined. Any other pattern is indicated as other and specified in brackets e.g. other (fibroepithelioma of Pinkus)
5. Absence of presence of perineural invasion/lymphovascular invasion
6. Nearest peripheral and deep excision margins
7. pTNM stage as per AJCC 7

Staging
e.g. used in STH

Histology shows a primary invasive basal cell carcinoma

Type of biopsy:

Maximum diameter of tumour (clinical / macroscopic):

Growth pattern:

Perineural / lymphovascular invasion:

Nearest surgical margins: Peripheral - …… mm / involved, deep - …… mm / involved

AJCC7 Staging

pTNM stage:

For MDT: No / Yes

b) Proforma for Squamous cell carcinoma and non-melanoma, non-merkel skin cancers including basosquamous ans adnexal

The following items should be recorded in the pro-forma.

1. Diagnosis
2. Type of biopsy
3. Maximum gross tumour diameter
4. Nature of lesion
5. Subtype – only common subtypes are outlined. Any other types are indicated as other and specified in brackets e.g. other (verrucous carcinoma)
6. Differentiation grade
7. Maximum tumour depth
8. Extent of invasion
9. Absence or presence of lymphovascular invasion
10. Absence of presence of perineural invasion
11. Nearest peripheral and deep excision margins
12. Staging

e.g. used in STH
Histology shows a primary invasive squamous cell carcinoma / basosquamous carcinoma / adnexal carcinoma (specify type) / other

Type of biopsy:

Maximum diameter of tumour (clinical / macroscopic): ≤ 2 cm / > 2 cm / unknown

Maximum tumour depth: ≤ 2 mm / ……. mm / Uncertain / Not applicable

Growth pattern (for basosquamous carcinoma only):

Subtype:

Differentiation grade:

Clark’s level: < 4 / ≥ 4 (reticular dermis / subcutis / fascia / muscle / cartilage / bone) / Uncertain / Not assessable

Perineural / lymphovascular invasion:

High risk staging features:

Nearest surgical margins: Peripheral - ……. mm / involved, deep - ……. mm / involved

AJCC7 Staging

pTNM stage:

For MDT: No / Yes

2. Malignant Melanoma

The following items should be recorded in the pro-forma.

Diagnosis
Type of biopsy
Relevant gross findings including :-
Maximum gross tumour diameter
Presence or absence of nodule and measurements if applicable
Comments on pigmentation
Nature of lesion
Subtype – only common subtypes are outlined. Any other type indicated as other and specified in brackets e.g. other (balloon cell melanoma)
Growth phase
Breslow thickness – measured to the tenth of a mm.
Mitotic count per sq. mm
Clark level
Absence or presence of ulceration
Absence or presence of lymphovascular invasion
Absence of presence of perineural invasion
Absence or presence of regression
Absence or presence of microsatellites
Absence or presence of co-existing naevus and if present, benign or dysplastic
Type of tumour infiltrating lymphocytic response
Nearest peripheral and deep excision margins
Staging

e.g. used in STH

Histology shows a primary malignant melanoma

Type of biopsy:

Specimen measurement:

Subtype:

Growth phase:

Breslow thickness:

Mitotic rate per sq mm:

Ulceration:

Clark’s level:

Perineural/lymphovascular invasion:

Regression:

Microsatellites:

Tumour infiltrating lymphocytes:

Co-existing naevus:

Nearest surgical margins:-
For in-situ: Peripheral - …… mm / Involved
For invasive: Peripheral - …… mm / involved, deep - …… mm / involved

AJCC7 staging

pTNM stage:

Minimum stage group:

For MDT
3. Merkel cell carcinoma

Histology shows a primary Merkel cell carcinoma (cutaneous neuroendocrine carcinoma)

Type of biopsy:

Maximum tumour dimension (clinical / macroscopic):

Maximum tumour thickness:

Clark’s level:

Lymphovascular/perineural invasion:

Nearest surgical margins: Peripheral - …… mm / involved, deep - …… mm / involved

AJCC7 Staging

pTNM stage:

For MDT
5.3 Double Reporting

All malignant melanomas and severely atypical naevi should be double reported if resources allow the report to be generated within 2 weeks. In the long-term, it is desirable that eventually all skin cancers are double reported to achieve consistency and accuracy in diagnosis.

There is evidence from observational studies that there are discrepancies between general pathologists and specialist dermatopathologists and specialists in pigmented lesion pathology in the reporting of melanomas and other pigmented skin lesions.

Observational studies provide evidence that histopathological examination has high sensitivity and specificity for melanoma. These studies also report difficulty with diagnostic accuracy and consistency for melanocytic lesions that are considered borderline for malignancy. Other lesions that are reported as presenting difficulty for pathological diagnosis include childhood melanoma, atypical naevi and Spitz naevi.

One retrospective study compared pathology reports from routine pathology services with a pathology review at a multidisciplinary pigmented lesion clinic. Diagnoses were revised by the multidisciplinary clinic in 11% of all reports, and review of surgical margins led to change in surgical margin status for 12% of patients. The study concluded that review by experienced pathologists within MDT’s provides internally consistent diagnoses and valuable second opinions.

With respect to double reporting, the Network agrees with the following:-

All severely dysplastic naevi and malignant melanomas will be double reported as agreed by the NSSG. Funding has been approved by the network for this purpose. Initial funding is based on estimated melanoma figures obtained from all the localities and adjusted to include estimation of severely dysplastic naevi. Subsequently it is expected that this will be based on more accurate figures obtained in the future. The amount of funding approved is sufficient for complete/full double reporting.

The exact mechanism of double reporting is entirely dependant on the individual laboratory and it will be a matter for local decision as to how the second opinion is recorded and communicated in the final report, to be prepared by the first pathologist.

1. Although the level of funding approved is sufficient for complete double reporting, the network has agreed that the double reporting will be done to confirm the diagnosis and the TNM stage; it is necessary to agree on the individual staging parameters only to the extent that there is agreement on the stage of the tumour. It is not necessary for all dataset parameters to be double reported The case should however be made available in its full extent to the second pathologist who may or may not wish to look at all the slides and other dataset items.

2. The first pathologist should issue a report that has been double reported, to avoid the risk of a revised diagnosis being issued after double reporting. The network has agreed that only one report will be issued following the double reporting process.

3. The histopathology report will contain both names of the pathologists who have been involved in this process. The report may contain a comment indicating
4. If there is a discrepancy that arises in the opinions between the two pathologists, it is the responsibility of the first pathologist to seek mechanisms to arrive at a consensus opinion. If no agreement can be reached, the differences in opinions should be indicated and the case sent to the SSMDT for a third opinion.

5. Although both pathologists will have responsibility over the case, the responsibility of making the case available for double reporting, ensuring that the second opinion is recorded and steps taken for reconciliation of differences in opinion will all be the responsibility of the first pathologist.

6. If a case required an external opinion to reach a diagnosis of melanoma or severely dysplastic naevus, then it will be deemed to be double reported.

It may be useful to record discrepancies between the first and second pathologists and any discrepancies between the general pathologists and specialist dermatopathologists/histopathologists. This will enable an audit to be performed in the future to evaluate the use of double reporting and the stage at which this is useful.

Please also refer to notes prepared by Dr John Goepel, Chair of the Network Pathology Group, as per his presentation to Skin NSSG on 12 February 2009. (Appendix 5 – hard copy)

5.4 LSMDT Pathology

Histopathologists should take a lead role in skin cancer. Pathology reports should include all the information required by the current Royal College of Pathologists minimum dataset for the relevant cancer. The histopathologists engaged in skin cancer diagnosis should participate in an appropriate external quality of assessment (EQA) scheme and demonstrate evidence of continuing professional development (CPD) relevant to skin cancer. The lead histopathologist should attend over 50% of MDT meetings. Other histopathologists reporting skin cancer should be able to demonstrate some MDT activity.

Refer cases requiring a second histological opinion to the lead histopathologist in the SSMDT.

Patients to be referred for LSMDT review

Care level 4:

High risk BCC – Recurrent or with positive excision margin
SCC – Recurrent or with positive excision margin
MM – New, single primary, adult, non-metastatic, not approved for trial entry, up to and including stage IIa.
Radiotherapy if attendance by clinical oncologist at LSMDT
Lesion where diagnosis is uncertain but may be malignant
Incompatible clinical and histological findings

The network agrees to the following with respect to LSMDT's

All LSMDT cases will be reviewed and discussed
Each LSMDT will:
- Refer cases requiring specialist management to the SSMDT
- Refer cases requiring a second histological opinion to the SSMDT
- Collect data for the network-wide audit
- All pathologists participating at the LSMDT will show evidence of involvement in a general EQA which includes skin
- The core histopathology member(s) will show evidence of over 50% MDT attendance
- Cover arrangement for the core histopathology members should be in place

The Sheffield LSMDT work will be incorporated within the Sheffield SSMDT
5.5 **SSMDT Pathology**

All cases referred to the SSMDT should receive formal diagnostic histopathology review.

All cases requiring a tertiary histopathology opinion should be supported by the SSMDT on a commissioning basis.

The LSMDT should provide rapid referral service to the SSMDT for patients who require specialist management.

- Refer cases requiring a second histological opinion to the lead histopathologist in the SSMDT.
- Cases to be discussed as LSMDT are summarised in Table 2. LSMDT’s will concurrently refer certain patients on to the SSMDT (see Table 4), IOG pg. 58.
- Any patients who are recognised (clinically or histologically) to have skin cancers with the characteristics listed in Table 4 should be referred directly to the SSMDT.

Ideally there should be at least two specialist dermatopathologists or histopathologists with a special interest in dermatopathology. This is to provide flexibility and adequate cover during leave periods.

There should be a designated lead in the area and ideally a deputy lead. The lead and deputy lead engaged in reviewing and reporting SSMDT skin cancer cases should each attend over 50% of SSMDT’s. Other histopathologists reviewing and reporting SSMDT work should be able to demonstrate some MDT activity.

All specialist histopathologists reviewing and reporting common and rare skin cancers should be able to demonstrate experience, competency and skills sufficient to fulfil the task, or undertake appropriate training to acquire the skills. The level of competence and skills for this activity is broadly that of the RCPath Diploma in Dermatopathology and American Board Certification in Dermatopathology. These qualifications are not however regarded as mandatory. All specialist histopathologists engaged in this work should participate in some CPD relevant to common and rare skin cancers and participate in an appropriate EQA scheme. Ideally this should be a national specialist EQA scheme in dermatopathology, when available. Those reporting primary cutaneous lymphoma must participate in an EQA scheme, including this group of diseases. It is also desirable that the CPD is facilitated by membership of appropriate national societies (such as the British Society for Dermatopathology and/or the UK Cutaneous Lymphoma Group). Each cancer or pathology network could hold a panel of histopathologists suitable for SSMDT participation based on these criteria.

Histopathologists restricting their activity in the SSMDT centre to work at the LSMDT level should be able to demonstrate the same activity as defined previously.
Patients to be referred for SSMDT review

Care level 5
Selected BCC’s and SCC’s needing plastic/reconstructive surgery by SSMDT core member (as per network agreed clinical guidelines)
Radiotherapy (as per network clinical guidelines). If not discussed and treated by LSMDT clinical oncology core team member
Metastatic SCC on preservation or newly metastatic
Malignant melanoma – Stage IIb or more, or less than 19 years or metastatic on presentation or newly metastatic or recurrent or for approved trial entry.
Any cases for approved trial entry
Any case for adjuvant therapy (as per network clinical guidelines)
Histology opinion from SSMDT core pathology team member
Mohs Surgery
Skin cancer in immunocompromised patients including organ transplant recipients
Skin cancer in genetically predisposed patients including Gorlin’s syndrome
Cutaneous lymphoma
Kaposi’s sarcoma
Cutaneous sarcoma above superficial fascia (Breslow fascia refer to sarcoma MDT)
Other rare skin cancers (see below)
All possible primary cutaneous lymphomas with a lack of diagnostic consensus at SSMDT should be reviewed by the Supra-Network

List of Rare Skin Tumours:
Epidermal and Appendage Tumours
Apocrine carcinoma
Hidradenocarcinoma
Eccrine porocarcinoma
Sebaceous carcinoma
Tumours associated with Muir-Torre syndrome
Eccrine epithelioma (syringoid carcinoma)
Microcystic adnexal carcinoma
Primary adenoid cystic carcinoma
Primary mucoepithelioid carcinoma
Primary mucinous carcinoma
Digital papillary adenocarcinoma
Malignant cylindroma
Malignant spiradenoma (spiradenocarcinoma)
Malignant pilar tumour
Malignant pilomatrixoma
Neuroendocrine carcinoma (Merkel cell tumour/trabecular carcinoma)
Dermal and Subcutaneous Tumours
Atypical fibroxanthoma (AFX)
Dermatofibromasarcoma protruberans (DFSP)
Leiomyosarcoma
Angiosarcoma
Kaposi’s sarcoma
Haemangioendothelioma
Epithelioid sarcoma
Primary cutaneous rhabdomyosarcoma
Cutaneous malignant nerve sheath tumours (including cutaneous neurofibrosarcoma and malignant schwannoma)
As the SSMDT serves as the LSMDT for the local catchment population the pathologists participating in this MDT can include pathologists who participate in the general EQA and NSDEQA. At the present time all 4 SSMDT pathologists participate in the NSDEQA (Rokiah Ali, David Slater, Nick Tiffin and Chris Warren)

The level of participation in the EQA will indicate the care level at which the pathologists are involved. All pathologists who participate in the general EQA will be working up to care level 4 cases. All pathologists who participate in the NSDEQA will be working up to care level 5 cases

Cases referred from the network to the SSMDT will be reviewed only by pathologists who participate in the NSDEQA. This includes Rokiah Ali, David Slater, Nick Tiffin and Chris Warren. The lead skin pathologist for STH is Dr. Nick Tiffin.

The core histopathology members will have to demonstrate over 50% of MDT attendance, while the other pathologists have to show evidence of some MDT attendance

Leave cover will be arranged amongst the participating SSMDT pathologists. for the level of care appropriate

All cases will receive a formal diagnostic histopathology review. For most cases this formal review report represents the second opinion. For cases requiring double reporting before issuing a first report, this formal review represents the third opinion

Cases requiring SSMDT will be initiated by the clinician in charge at the locality of origin who will send an SSMDT referral summary sheet to the SSMDT Co-ordinator. The clinician will simultaneously also request the relevant laboratory to send the slides and copy reports to the SSMDT. It is essential that a copy of the SSMDT summary sheet is included for pathologists reviewing the case, so that when reviewing the case, important clinical questions can be addressed.

The respective laboratory (where the case is originating from) will arrange for all slides, blocks, copy of the histopathology report and a copy of the SSMDT referral summary sheet to be sent to the SSMDT.

This should be addressed to:
Skin Secretaries,
Department of Histopathology
E Floor (till end of 2012) & F Floor (from 2013 onwards)
Royal Hallamshire Hospital
Glossop Road
Sheffield Teaching Hospitals NHS Foundation Trust
S10 2JF

The slides will be addressed to the SSMDT. The cases are either booked in by the skin secretaries depending on the material received. If only slides are received, this is booked in by the skin secretaries and if there are also blocks included, then booking in is done by the laboratory staff.
The case(s) will then be allocated and delivered to the pathologists on rota for the day.

If necessary further investigations will be carried out.

Cases of cutaneous (newly diagnosed or otherwise) lymphoma will also be reviewed by the SSMDT pathologists. There will be a low threshold for these cases to be discussed with the lymphoma lead (Dr David Slater) in case of uncertainty.

On completion, the SSMDT case(s) reviewed will be listed for the SSMDT and discussed at the next earliest SSMDT.

SSMDT case(s) may be presented by another pathologist other than the one who initially reviewed the case depending on whoever is covering the SSMDT.

Once management decision is made, the original slides and blocks are returned to the locality the case originated from, except for cases of cutaneous lymphoma, when the case will be first circulated to the other SSMDT pathologists who have not seen the case (to ensure continuous exposure and sharing of expertise of these less common group of disorders).

Any immunos carried out in Sheffield will be retained in file. For educational purposes, the index pathologist may wish to request for any of the slides from Sheffield and subsequently return them for filing purposes.

A clear record is maintained as to where the slides and blocks are filed, to enable easy access to the material if required in the future.

**Cutaneous Lymphomas**

All patients should be seen and managed by the SSMDT which should include a dermatopathologist with expertise in cutaneous lymphoma (NICE guidance in Improving Outcomes in Haematological Cancers). Close liaison should be maintained with a haemato-oncopathologist, as appropriate. Cases of possible systemic haematological malignancy involving the skin should be referred to the appropriate haematological malignancy MDT.

LSMDT’s should be involved once the diagnosis and staging has been confirmed by the SSMDT.

All lymphoma patients should undergo diagnostic biopsies for histology immunophenotyping and molecular studies, and this should be correlated with clinical presentation for accurate diagnosis and prognosis.

The SSMDT should have access to specialist laboratory testing of tumour tissue and blood for immunophenotyping, molecular analysis and blood viral serology.

The SSMDT should have access to bone marrow aspirate and trephine biopsies for complete staging of all patients with B and NK-cell lymphomas and for patients with CTCL variants and late stages of mycosis fungoides (stage IIb or above).
The World Health Organisation (WHO) – EORTC primary cutaneous lymphoma classification should be used to classify primary cutaneous lymphomas.

The Network agrees with the following practices in regards to cutaneous lymphoma

1. Diagnostic biopsies will be carried out for suspected cases of lymphoma. A detailed clinical history and relevant findings on examination should be provided in the histology request form for close clinicopathological correlation and accurate diagnosis.

2. Although Dr David Slater will be the named lead for cutaneous lymphoma for the network and this work carried out under his leadership, the other SSMDT pathologists will also participate in this work for the following reasons:
   a) For purposes of leave cover
   b) To ensure continuous exposure and sharing of expertise amongst the group of pathologists involved
   c) For succession planning and continuity of service in the future

3. It is recommended that only a basic panel of immunohistochemistry is carried out at the localities mainly to confirm or distinguish between a lymphoproliferative disorder and other causes. This is to avoid any unnecessary immunohistochemistry and also to avoid repetition of immunohistochemistry at the SSMDT level.

4. After referral to the SSMDT, only relevant investigations pertinent to the diagnosis (as opposed to educational purposes) should be performed to keep costs to a minimum. However, there should be no restrictions in the extent of immunohistochemistry if needed to establish a diagnosis. This also covers necessary genotypic, cytogenetic studies, and viral serological tests which will be covered by cross charging.

5. If the case then falls under the category of LSMDT, the case is then returned for LSMDT discussion at the locality. If the case falls under the category of SSMDT, the case can be retained and returned after SSMDT discussion.

6. There should be a low threshold for sharing cases with the haematopathologist. Any cutaneous lesions of systemic haematological origin should be discussed with the haematopathologists and referred to the HODS MDT (see measures 08-1A-216j).

7. Cases needing specialist advice from supranetwork centres, should be referred on.
Skin Sarcomas

Skin cancer MDT’s should liaise with sarcoma MDT’s in the management of patients with cutaneous sarcomas. As stated in the section on SSMDT’s, it is essential for all cutaneous sarcomas to receive specialist histopathology review.

It is essential and there is close liaison between the SSMDT and sarcoma MDT’s. This is particularly important for patients whose sarcomas are large or penetrate the superficial fascia or are of a histological type requiring chemotherapy (e.g. rhabdomyosarcoma, Ewing’s sarcoma).

The Network agrees with the following practices in regards to cutaneous sarcoma

1. All sarcomas will be referred to the SSMDT.

2. Investigations (immunohistochemistry, FISH/chromosomal analysis) deemed appropriate for establishing a diagnosis will be supported. This will be covered by cross charging.

3. A formal histopathology review will be issued by the SSMDT pathologist.

4. There should be a low threshold for sharing cases with the sarcoma pathologists.

5. Cases that fall under the category of SSMDT will be discussed at the SSMDT.

6. Those needing sarcoma MDT discussion will be referred on, either directly or via referral from the skin SSMDT.
5.6 **Operational Management**

The following arrangements outline the details of the SSMDT WorkHandled in STH:-

1. SSMDT cases from the localities in the network will be identified by the clinician in charge, who will complete the SSMDT referral summary sheet and send it to the SSMDT Co-ordinator. The clinician will also inform the respective laboratory simultaneously with a copy of the SSMDT referral summary sheet. These are classed as SSMDT review cases.

1. Slides will be sent in from the localities for SSMDT usually accompanied by copies of the SSMDT referral summary sheet and histopathology report. Sometimes cases may be sent in (without a referral summary sheet) for SSMDT histopathology opinion. These are classed as SSMDT consultation cases.

The slides and blocks with copies of SSMDT referral summary sheet and histopathology report will be placed in an envelope and addressed to:
Skin Secretaries,
Department of Histopathology
E Floor (till end of 2012) & F Floor (from 2013 onwards)
Royal Hallamshire Hospital
Glossop Road
Sheffield Teaching Hospitals NHS Foundation Trust
S10 2JF

2. These cases will be handled by the skin secretaries. The case should be date and time stamped on receipt.

3. The cases will be booked in either by the secretaries themselves or by a laboratory staff depending on the type of material received.

4. The cases are tagged with automatic review code (SSMDT) to enable future identification, audit and charging purposes.

5. The number of blocks and slides received are entered onto the system. The pathologist’s name to whom the case is allocated to is entered into the SSMDT database to enable easy tracing of cases.

6. The cases will then be distributed to the SSMDT pathologist(s) (R. Ali, D N Slater, Nick Tiffin and C W Warren) according to the skin team rota. The case will be date and time stamped prior to despatch.

7. Cases will alternate between Skins 1 and 2, as per the rota.

8. The cases will be placed in the tray or given directly to relevant pathologist(s) to whom the case is allocated indicating SSMDT.

9. All cases including cutaneous lymphoma will be dealt with in this way.
10. 4 pm is the cut off point for allocating cases to the pathologists on rota that day and after 4, the cases will be allocated to the pathologists on rota the next day.

11. If the pathologist on rota is away e.g. sick leave, the pathologist’s secretary will ensure that the case is passed on to the covering pathologist(s) for the day.

12. Any investigations including spare H&E, immunohistochemistry, genotypic or cytogenetic studies will be requested after the pathologist has seen the original sides and read the report.

13. For cutaneous lymphoma cases, if there is any uncertainty with regards to the diagnosis the case will be shown to Dr David Slater (lead pathologist for cutaneous lymphoma) for his opinion. There should be a low threshold for this interaction.

14. When the case is completed, a formal histopathology review is issued, addressed to the original pathologist with copy requests for the clinician in charge and the SSMDT Co-ordinator.

15. A clear record indicating the whereabouts of slides and blocks is entered in the SSMDT database system and in the notes on Apex.

16. MDT presentation of SSMDT cases is mostly done by the reporting pathologists or their cover.

5.7 Laboratory Investigations and their Indications [08-1C-112]

The NSSG should agree network-wide pathology guidelines for the diagnosis and assessment of skin cancer. The guidelines should address:-

Laboratory investigations
Their specific indications

A document regarding Quality assurance in histopathology reporting practice has been published by the Royal College of Pathologists in February 2009. It is recommended that all pathologists reporting skin cancers practice in accordance with guidelines issued in this document for quality assurance purposes.

Skin tumours should be classified in accordance to the World Health Organisation Classification of tumours (WHO classification – Skin Tumours published by IARC Press, International Agency for Research on Cancer). Please refer to this book and other standard textbooks for detail laboratory investigations for each and every specific skin cancer including lymphomas. However, recommended laboratory investigations have been outlined for the more common skin cancers, a few of the rarer skin cancers and for many of the cutaneous lymphomas. The recommendations here are only the minimum and it may be that more immunohistochemistry is needed depending on individual cases.
If it is known that the case will be referred to the SSMDT it would be acceptable if the index pathologist chooses not to perform any further investigations for confirmation and refers on directly. If there is uncertainty with the diagnosis, the case can be sent to the SSMDT pathologist for histological opinion without any prior immunohistochemistry.

As for haemato-oncology cases anything other than minimal local investigations is strongly discouraged.

Recommended laboratory investigations for non-melanoma skin cancers:-

**Basal cell carcinoma (BCC)**

BerEp4 positive (diffuse)
BCL2 positive (diffuse)

**Squamous cell carcinoma (SCC)**

EMA positive
High molecular weight cytokeratins (MNF116) positive
BerEp4 may be positive in basaloid SCC
Ki67 usually higher in SCC than BCC
P63 positive

**Basosquamous carcinoma**

BCC component positive for BerEp4 and negative for EMA
Squamous component positive for EMA. Basaloid SCC may be positive for BerEp4

**BCC vs Desmoplastic Trichoepithelioma (DTE)**

BerEp4 – Diffuse positivity in BCC and focal positivity in DTE
BCL2 – Diffuse positivity in BCC and focal positivity in DTE
CD34 – Negative in BCC and stromal cells positive in DTE

**Microcystic Adnexal Carcinomas**

High and low molecular weight cytokeratins positive
EMA and CEA highlight the ducts

**Sebaceous Carcinoma**

EMA – Positive (may enhance the cytoplasmic ‘bubbliness’ of the tumour cells)
CK7 positive, BerEp4 positive
MNF116 – Positive for the basal layer but negative in the sebocytes

* The dimorphic staining pattern of basal cells and mature sebocytes with cytokeratins and EMA is quite useful in the diagnosis

**Merkel Cell Carcinoma**

CK20 – ‘Dot’ positivity (sensitive and specific marker)
Cam 5.2 – Positive
Pancytokeratin – Positive
EMA – Positive
BerEp4 – Positive
Chromogranin – Positive
Synaptophysin – Positive
CD99 – Positive (¼ cases)
TTF-1 – Negative (to differentiate from metastatic small cell carcinoma lung, which
are TTF-1 positive and <10% of these tumours show positivity for CK20)
LCA/CD45 – Negative
S100 – Negative

Mammary Paget’s Disease

Low molecular weight or pancytokeratin positive e.g. CK7 +, Cam 5.2 +, AE1/AE3 +,
EMA +
CEA +
GCDFP15 + (50% positive)
ER and PR positive (5% cases)
LCA/CD45 – Negative

Extramammary Paget's Disease

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<th>Secondary</th>
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<tr>
<td>CK7 +</td>
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<tr>
<td>GCDFP15 +</td>
<td>GCDFP15 -</td>
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<tr>
<td>CK20 -</td>
<td>CK20 +</td>
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<tr>
<td>Cam 5.2 +</td>
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Kaposi’s Sarcoma

CD34 + and CD31 +
HHV8 +

Leiomyosarcoma

SMA + Desmin +

* The above immunos are usually positive in leiomyosarcoma although none of them
is absolutely specific for smooth muscle or muscle in general. It is advisable to
demonstrate positivity for both markers rather than positivity for only one of the
markers before a diagnosis of leiomyosarcoma is made.

Atypical Fibroxanthoma

P63 negative (more useful than cytokeratins)
Broad cytokeratins e.g. CK5/6 or AE1/AE3 negative
Cam 5.2 -, MNF116 -
CD10 +
CD99 +
SMA, EMA, CD68 – Can show focal positivity
S100 – Can show focal positivity for background dendritic cells
(Procollagen – 1 is more specific and is preferable if available)

**Dermatofibromasarcoma Protruberans**

CD34 +
F13A – Negative

**Recommended laboratory investigations for melanocytic skin lesions:**

With difficult melanocytic lesions, immunohistochemistry can be useful in the diagnosis. Depending on the case, it is recommended to choose from the following maximum panel of immunohistochemistry for melanocytic lesions:

S100, Melan A, HMB45, Cyclin D1, Ki67 and p16

S100 – Most sensitive melanocytic marker (“gold standard” stain for melanocytic lesions). It is advisable to include this in the panel for any lesion suspicious of melanoma
Mel A – Less sensitive but more specific
HMB45 – Aberrant expression (diffuse vs. zonal pattern) is useful in the diagnosis of malignancy e.g. in naevoid melanoma. However this has to be interpreted cautiously as aberrant expression can also be seen in blue naevus, deep penetrating naevus and combined naevus
Ki67 – An increased proliferation fraction of > 5% is usually indicative of malignancy. However this has to be interpreted with caution if the lesion has associated lymphocytic response which can express Ki67 suggesting an increased nuclear turnover
Cyclin D1 – Overexpression (diffuse vs. zonal pattern) is usually in malignancy.
p16 – Loss of expression correlates with invasive and metastatic melanoma. Strong expression is more common in benign melanocytic lesions
BRAF for those patients who are being considered for systemic treatment of metastatic melanoma
Especially in lentigo maligna or in also other types of melanomas, where there is a suspicion of invasion, immunohistochemistry can be useful to determine presence or absence of an invasive component. In this context, Melan A and S100 are recommended.

**Desmoplastic Malignant Melanoma (MM)**

Is usually strongly positive for S100 but negative for HMB45

**Recommended laboratory Investigations for Cutaneous Lymphoma:**

All new presumed cases of cutaneous lymphomas should be referred for a second histopathology opinion. This being the case, cutaneous lymphomas need not be worked up in detail by the index pathologist referring the case. The extent of work-up of lymphoma prior to referral is entirely up to the index pathologist. However, pathologists are discouraged to perform anything more than the minimal necessary. A limited basic immuno panel may include T and B cell markers, CD30 and CD56, depending on the provisional diagnosis.
However, this guideline covers the panel of immunohistochemistry recommended for specific types of lymphoma.

**Mycosis Fungoides**

CD2, CD3, CD4, CD5, CD7, CD8, CD30, CD45RO, CD20, CD79a, Ki67, TIA-1 and Granzyme B

**Sezary Syndrome**

CD2, CD3, CD4, CD5, CD45RO, CD7, CD8, CD26, CD45

**Lymphomatoid Papulosis**

CD30, CD45, CD3, CD4, CD8, CD2, CD5, CD7, CD56, TIA-1, Granzyme B, CD25, CD15, MUM-1

**Anaplastic Large Cell Lymphoma**

CD30, CD2, CD3, CD4, CD45RO, CD25, CD30, TIA-1, Granzyme B

In addition, ALK and EMA to distinguish between nodal ALCL with secondary in the skin or primary cutaneous ALCL (both negative in primary cutaneous ALCL).

**Subcutaneous Panniculitis like T Cell Lymphoma (Alpha-Beta)**

CD8, TIA-1, Granzyme B, CD3, CD4, CD56, CD30, Ki67 and Beta F1 (if available)

**Cutaneous Gamma-Delta Positive T Cell Lymphoma**

EBV, CD3, CD2, CD7, CD5, CD4, CD8, TIA-1, Granzyme B, CD56

**Primary Cutaneous Aggressive Epidemotropic CD8+ Cytotoxic T Cell Lymphoma**

CD3, CD8, Granzyme B, TIA-1, CD2, CD4, CD5, CD7, EBV, and Beta F1 (if available)

**Primary Cutaneous CD4+ Small/Medium Pleomorphic Lymphoma**

CD3, CD4, CD8, CD30, CD2, CD5, CD7, Beta F1 (if available)

**Primary Cutaneous Peripheral T Cell Lymphoma – Unspecified**

CD2, CD3, CD5, CD7, CD4, CD8, CD30

**Adult T Cell Leukaemia/Lymphoma**

CD3, CD4, CD25, CD45RO, CD7, CD8, CD20, CD30

**Extranodal NK-T Cell (nasal type)**

EBV (EBER ISH preferred over LMP), CD56, CD2, TIA-1, Granzyme B, CD43, CD3, CD4, CD8
Cutaneous Marginal Zone B Cell Lymphoma

CD20, CD79a, CD5, CD43, BCL6, BCL2, CD21, CD23, CD138, CD10, CD3, CD45RO

Cutaneous Follicle Centre Lymphoma

CD79a, CD20, BCL6, CD10, BCL2, CD21, CD23, CD35, CD5, CD43, Ki67, CD3, CD45RO

(Secondary cutaneous follicle lymphoma – CD10 + BCL2 +)

Cutaneous Diffuse Large B Cell Lymphoma – leg type

CD20, CD79, BCL2, MUM-1, CD10, CD138, BCL6

Cutaneous Diffuse B Cell Lymphoma – Other

Diagnosis of exclusion – Not B cell leg type and not follicle centre lymphoma
BCL2 and BCL 6

Lymphomatoid Granulomatosis

CD79a, CD20, CD3, CD4, EBV

Mantle Cell Lymphoma

CD5, CD10, CD23, Cyclin D1, BCL6, CD20 and CD45

All cancer networks should have easy access to appropriate immunophenotypic, molecular biological and cytogenetic facilities. Some of the latter are very specialised pathology services and may not be provided by pathology laboratories within the LSMDT or SSMDT.

The SSMDT histopathologists will be allowed relevant investigations if deemed appropriate to the case dealt with, if this is pertinent to the diagnostic process. This may be in addition to any investigations done at the locality of origin. It may or may not be necessary to repeat immunos depending on the case. The costs for additional immunohistochemistry will be covered by cross charging. This has to be kept to the minimum necessary to achieve accurate diagnosis as agreed by the network.

The entire extent of investigation necessary for diagnostic purposes needs to be supported by the Network.

In some cases this may include other ancillary investigations such as:-

1. Genotypic analysis (TCR and IgH gene rearrangement) – for cutaneous lymphoma
2. FISH/Chromosomal analysis – for soft tissue or cutaneous lymphomas
3. Electron Microscopy – To determine tumour cell lineage in some difficult cases where light microscopy and/or immunophenotyping cannot confirm the diagnosis
5.8 Audit

The Skin Cancer NSSG agrees that each locality will have a policy whereby the lead clinician of the MDT should make recommendations to the cancer clinician lead(s) of referring PCT’s when any of their clinicians who are not accredited GPwSIs are found to be excising skin cancers by the alert system of the pathology laboratory.

It is proposed that this may include an audit of the management of all patients with excised BCC’s and SCC’s not discussed at MDT meetings. This audit should be presented to the MDT on a quarterly basis.
5.9 External Quality Assurance (EQA) [08-2J-113]

The core histopathology members of the LSMDT should be taking part in a general histopathology EQA that includes skin pathology. The SSMDT pathologists should be participating in the National Specialist Dermatopathology EQA (NSDEQA)

Note: They may be taking part in the national specialist dermatopathology EQA, in which case, this confers compliance with the above requirement.

The columns below highlight the pathologist against the relevant EQA involved by the pathologist

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<tr>
<th>Sheffield Teaching Hospitals</th>
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<tr>
<td>Rokiah Ali</td>
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References:

Royal College of Pathologists – Quality Assurance in histopathology reporting practice GO82

http://www.rcpath.org.resources/pdf/go82draftqahistoreportingct08.pdf log in required.

National Institute for Clinical Excellence – Improving outcomes for people with skin tumours including melanoma.

www.nice.org/guidance/cancer


Royal College of Pathologists – Standards and Minimum Datasets for Reporting Cancers. Minimum dataset for the histopathological reporting of common skin cancers.


Pathology Subgroup Members – North Trent Skin NSSG

Dr Rokiah Ali – Author for pathology guidelines & Lead for Skin NSSG (STH)
Prof. John Lee (Skin Lead for Rotherham)
Dr Mahariz Muzaffar (Skin Lead for Doncaster)
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Dr. Mini Varghese (Skin Lead for Barnsley)
6.0 Supportive and Palliative Care Guidelines

Supportive and palliative care is a cross-cutting issue that affects all cancer patients, at all stages from pre-diagnosis to survivorship, or death. The NICE guidance on supportive and palliative care for adults with cancer (2004) gives definitions of these two terms and tries to explain their distinctions and overlaps. However, there is still considerable confusion and unclear thinking about these concepts, with the important consequence that many patients are being referred inappropriately, too late or not at all to specialist services.

The Sheffield Model for Supportive Care clarifies this area and should be seen as the model which applies to North Trent Cancer Network (Ahmedzai, Walsh, *Seminars in Oncology*, 2001.)

In essence, supportive care is a wide range of specialist services which work as a ‘virtual team’ to help the patient (and family) cope with the effects of disease, of treatment-related side-effects (acute and long-term) and with the psychosocial and rehabilitation needs for both long-term survivors with cancer and those who are progressing.

Palliative care is a somewhat more restricted range of services, often configured as an actual team, which focuses on symptoms, psychological, social and spiritual issues for patients and their families, when the disease is progressive and will likely lead to death within 6-12 months.

In many acute settings, palliative care teams provide both supportive and end of life care; in community and hospices, they concentrate almost exclusively on end of life care. Specialists who contribute to supportive care for cancer patients, e.g. dieticians, SALT, other AHPs, pain clinic staff, are scattered across a hospital and are often not coordinated. They provide only limited input in community and hospices. It is ideal for cancer MDTs to have their own dedicated supportive care professionals, or at least dedicated sessions from a trust service.

All patients, regardless of the stage of disease or estimated prognosis, are candidates for supportive care and all those who are nearing the end of life are candidates for palliative care. The difficulty arises in identifying which patients need the different specialists’ inputs of supportive care at which stage. In North Trent we have developed a screening questionnaire – SPARC, which provides a ‘holistic’ assessment of a patient’s needs for symptom control, psychological, social and spiritual issues as well as needs for information, help with daily living, making plans, and other areas. It is recommended that this instrument is used by clinics and wards to identify patients who need supportive and palliative care. An alternative tool is the ‘Distress Thermometer’ – the North Trent Supportive and Palliative Care Group is producing guidelines to advise MDTs to choose one or other of these tools, as well as other more specific questionnaires for complex pain, psychological distress, etc.

Most acute settings in North Trent have a team of palliative care nurse specialists. Only 4 out of the 5 localities have consultant-level input into these teams.
Furthermore, only 3 of out 5 localities have consultants with regular sessions in hospices.

The Sheffield/Chesterfield/Rotherham localities have a 24/7 medical on-call service with first-on registrars (covering Sheffield and Chesterfield) and second-on consultants (covering all three localities). The consultants also provide an informal second-on call service for the specialist palliative care teams in Barnsley and Doncaster/Bassetlaw.

Ideally a member of each trust palliative care team should attend the Skin cancer MDT. However there is currently insufficient staff to support this. Moreover, MDTs are not always configured to pick up and discuss supportive and palliative care issues within the normal agenda. It is recommended that an alternative arrangement is made to cover this by:

   a. Routine use of a supportive care screening tool, e.g. SPARC or Distress Thermometer by all clinicians in both in-patient and out-patient settings.
   b. Clearly identified routes of referral between the Skin MDT, usually via the CNS but also via medical staff, to a named person in the local palliative care MDT.
   c. The ability to timetable discussion of complex supportive or palliative care issues for specific patients in the MDT meeting, e.g. to discuss palliative surgery, difficult pain or respiratory management, transfer to hospice or other settings.

It is recommended that advance care planning for all patients is started as soon as feasible after the diagnosis of an incurable cancer, including the patient's preferences for place of care in the terminal stage and the use or rejection of interventional medical support, e.g. artificial hydration, CPR.
7.0 Teenager and Young Adults

In January 2009 specific referral pathways were developed for teenagers (16-18 years) and young adults (19-24 years) into the TYA MDT. The Skin Cancer NSSG has agreed age appropriate referral into these pathways Set out in the embedded document below

Discussion took place at the April Skin Cancer NSSG meeting and the group agreed the referral pathways.

TYA MDT Pathway

Refer to:
Appendix 6 - TYA Clinical Referral Form (hard copy)
Appendix 7 – TYA MDT Sample Referral Procedure (hard copy)