Kent & Medway Cancer Collaborative

Cutaneous Lymphoma

Pathway of Care

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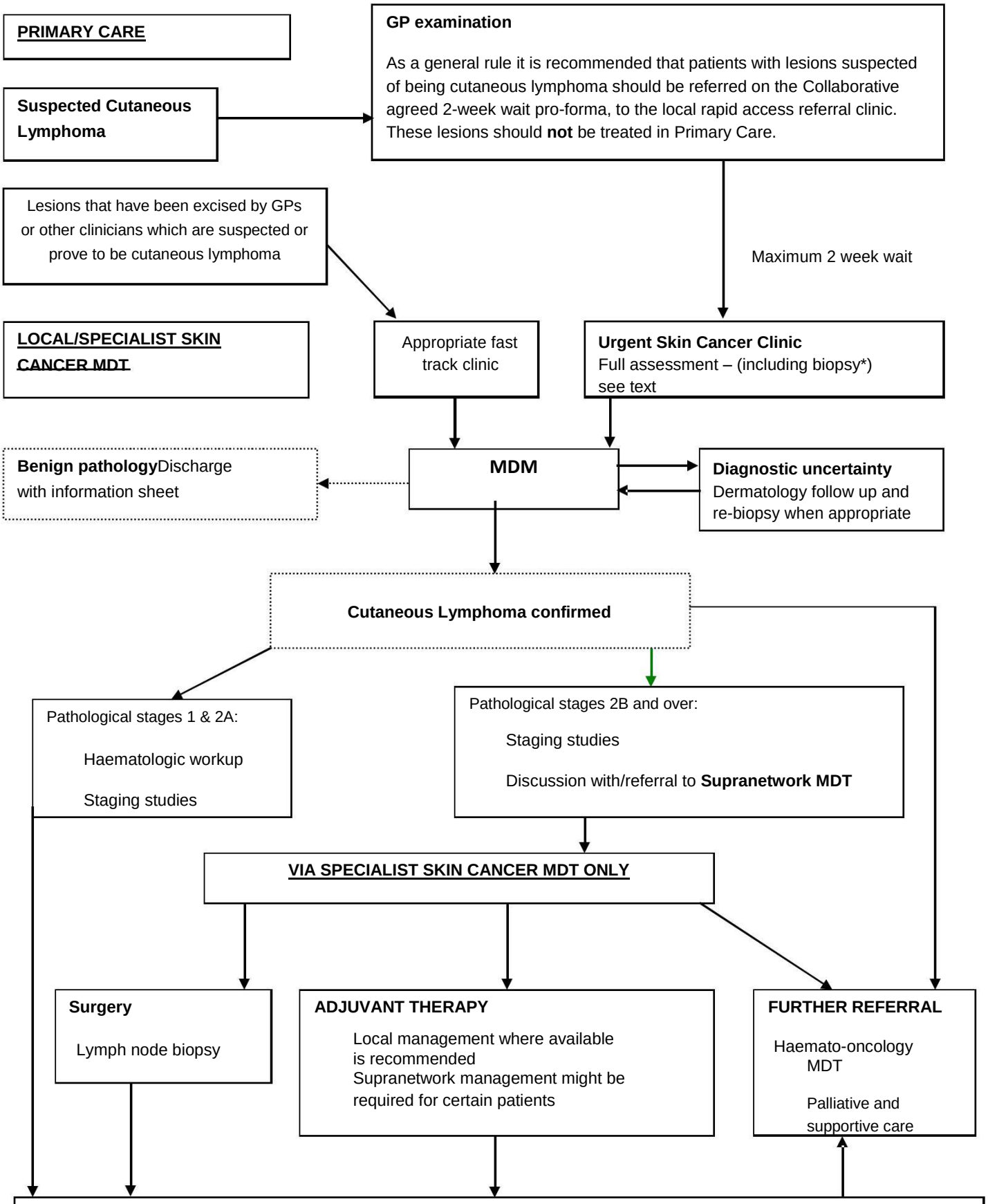
Contents

1.0 Pathway Overview



2.0 Process and Terminology

2.1 Scope

This document sets out the pathway of care for patients with suspected/proven Cutaneous Lymphoma. It has been updated to ensure compliance with the 2006 NICE Improving Outcomes Guidance (IOG) for people with Skin Tumours including Melanoma (IOG), and the Peer Review Measures (2010). The IOG expects Kent & Medway to provide four levels of care for patients with Cutaneous Lymphoma.

These are:-

1. Primary Care
2. Local MDT (LSMDT)
3. Specialist MDT (SSMDT)
4. Supranetwork MDT

Based on the population of Kent and Medway, the Skin Cancer Tumour Site Specific Group (TSSG) is proposing that there are two skin cancer MDTs in Kent and Medway that function at both Local and Specialist level, working to one set of protocols and policies agreed by the TSSG and its sub groups.

This document describes the process/care available at each level and is compliant with the 2006 IOG and the Peer Review Measures (2010).

2.2 Tumours

The scope of the pathway of care is confined to:-

Primary Cutaneous Lymphoma:-

- T cell
- B cell

2.3 Referral Guidelines and Processes

General practitioners are encouraged to refer patients with a suspected cutaneous lymphoma as a matter of urgency.

Patients with suspected cutaneous lymphoma should be referred under National cancer 2-Week Wait rules (2-WW).

The Cutaneous Lymphoma sub-group recommend that lesions thought to be cutaneous lymphoma should not be biopsiedin Primary Care.

Lesions biopsied in Primary Care that prove to be cutaneous lymphoma should be referred urgently under the 2-week wait rule. A copy of the histology report should either be faxed with the referral letter or accompany the patient to the clinic appointment. If a photograph of the pre-biopsied lesion was taken, a copy should accompany the patient if possible.

These patients will be discussed at the multi-disciplinary meeting (MDM) in the first instance.

2.4 IOG Recommendations

As per Skin Cancer Measures

SUPRANETWORK T-CELL CUTANEOUS LYMPHOMA MDT FOR TOTAL SURFACE ELECTRON BEAM THERAPY (TSEBT).

The Kent & Medway Cancer Collaborative (KMCC), in consultation with the Skin Cancer TSSG, has agreed with the specialist commissioners the guideline which specifies:-

That all cases of nodular mycosis fungoides (stage 11B or over), should be referred for discussion and consideration of Total Surface Electron Beam Therapy (TSEBT).

That the Skin Cancer Unit, St. John's Institute of Dermatology, Guy's and St. Thomas' NHS Foundation Trust, is the named Supranetwork MDT used by Kent & Medway for this function.

SUPRANETWORK T-CELL CUTANEOUS LYMPHOMA MDT REFERRAL FOR PHOTOPHERESIS

The KMCC, in consultation with the Skin Cancer TSSG, has agreed with the specialist commissioners the guideline which specifies:-

That all cases of erythrodermic cutaneous T-cell cutaneous lymphoma, stages 3 and 4, having both skin involvement and circulating T-cell clonal cells, will be discussed with the clinician in charge of a named photopheresis facility for potential referral and treatment by photopheresis.

That the Skin Cancer Unit, St. John's Institute of Dermatology, Guy's and St. Thomas' NHS Foundation Trust, is the named Supranetwork MDT used by Kent & Medway for this function.

2.5 Local Agreement with Supranetwork MDT

Following discussions with the lead of the Supranetwork MDT at a Lymphoma sub-group meeting held on 26th February 2009, desired and additional non-compulsory recommendations have been suggested:-

Development of improved communication and exchange links are encouraged.

That all appropriate management and follow up will be undertaken locally.

That with the diagnosis of CTCL/CBCL all patients (all grades) details may be logged with the supranetwork.

- Even stages 1 to 11A may be registered for data collection purposes.

Where histology has been referred to a tertiary institution for a second opinion:-

- The SSMDT will be informed of the event.
- The SSMDT may wish to inform the supranetwork to aid clinicopathological correlation and receive the information from the MDT discussion of the case.

These recommendations will be annually reviewed to assess value and revise as appropriate.

3.0 Rapid Access Referrals

Patients should be seen within 2 weeks of referral.

Patients will:-

- Be reassured if clinically and histologically benign.
- Undergo photography of lesion for recording if indicated.
- Undergo thorough examination including assessment of lymph node status.
- Undergo ex/incisional biopsy of suspicious skin lesions.

The visit will be fully documented using the agreed pro-forma. The patient will receive "discharge" information.

4.0 Cellular Pathology

All KMCC reporting pathologists follow The Royal College of Pathologists Histopathology Reporting on Cancers guidelines – a copy of which is available through the KMCC website:- http://kmcc.nhs.uk/tumour-sites/sub-groups-or-cross-cutting-groups/pathology-group/

It is expected that pathology departments will adhere to guidelines on specimen handling described in the Royal College of Pathology Minimum Data Set for Skin Cancer (RCP MDS).

In order to comply with the Department of Health (DoH) 31 day (referral to diagnosis) target, histology departments are obliged to report on pigmented lesions within 2 weeks of biopsies taken at Urgent Skin Cancer Clinics.

The Skin Cancer TSSG will monitor adherence to both the guidelines set out in the RCP MDS and the 31 day targets through process of regular audit.

5.0 Specialist Skin Cancer Multi-Disciplinary Team Meeting (SSMDT)

All patients will be discussed at site specific multi-disciplinary meetings (SSMDTs).

These will normally be held weekly.

Patients may be discussed at more than one MDM as they progress along their pathway of Care (PoC).

The multi-disciplinary team will include the required IOG stated personnel.

5.1 Data

Collection of data at each stage of the pathway is the responsibility of the team looking after the patient at that time. The minimum dataset agreed by the TSSG will be a combination of those data items that meet national requirements, and additional items as agreed by the TSSG.

National data requirements will include:

Cancer Waiting Times monitoring, including Going Further on Cancer Waits. The data items required will be as defined in ISB0147 at the time of referral and/or treatment.

Details of the Cancer Waiting Times dataset are available from:
http://nww.connectingforhealth.nhs.uk/nhais/cancerwaiting/documentation

Cancer Waiting Times data will be submitted according to the timetable set out in the National Contract for Acute Services.

The Cancer Outcomes and Services Dataset. The data items will be as defined in ISB1521, and any subsequent versions, at the time of diagnosis and/or treatment. The requirement will include those fields listed in the "Core" section of the dataset, and any additional tumour site specific sections, as applicable.

Details of the COSD are available from:
http://www.ncin.org.uk/collecting_and_using_data/data_collection/cosd.aspx

Cancer Registration and Cancer Outcomes and Services (COSD) data will be submitted according to the timetable set out by National Cancer Registration Service (NCRS).

Where applicable, teams will also collect additional data items as defined in any corresponding National Clinical Audit Support Programme (NCASP) audit dataset.

Details of these datasets are available from:
http://www.ic.nhs.uk/services/national-clinical-audit-support-programme-ncasp/cancer

Data for NCASP audits will be submitted, where applicable, according to timetables as agreed by the TSSG, and within the overall submission deadlines for each audit.

Submission of data to meet these national requirements will be the responsibility of each individual Trust.

Note that these standards are subject to variation from time to time, and where these requirements change, the data items required to be collected by the team will also change in line with national requirements.

Local data requirements will include any additional data items as agreed by the TSSG. These must be selected to avoid overlap with any existing data items, and where possible must use standard coding as defined in the NHS Data Dictionary.

Where possible and applicable, InfoFlex will be used for the collection and storage of data.

Additional areas of the COSD, relating to pathology, radiotherapy, SACT, diagnostic imaging and basic procedure details will feed into the dataset from other nationally mandated sources. It is the responsibility of each team to ensure that the whole of the relevant dataset is collected, and it is acknowledged that this may come from a variety of sources.

6.0 Supranetwork MDT

6.1 Skin Cancer Unit, St. John's Institute of Dermatology

Cutaneous lymphoma:

The unit acts as the specialist centre for the South London Skin Tumour network, and provides a local, regional and national service for the diagnosis and treatment of cutaneous melanoma and primary cutaneous lymphoma as well as rarer skin cancers (Kaposi sarcoma, Angiosarcoma, Merkel cell carcinoma).

The multidisciplinary team includes dermatologists, plastic surgeons, both medical and clinical oncologists and clinical nurse specialists and is supported by a dedicated dermatopathology department, skin radiotherapy service including TSEBT, dedicated day unit for chemotherapy, clinical trials research team and a molecular diagnostics laboratory.

Patients for diagnostic assessment of any possible primary cutaneous lymphoma (primary cutaneous B and T-cell lymphomas and other cutaneous haematologic malignancies).

Patients with primary cutaneous lymphomas for management advice/staging.

Patients with all stages and subtypes of primary cutaneous lymphomas for entry into clinical trials.

The unit offers a wide range of treatments for all stages of disease including ECP, PUVA and TSEBT. They also undertake clinical trials for novel therapies.

6.2 Skin Tumour Molecular Diagnostics Lab, St. John's Institute of Dermatology

The molecular diagnostics laboratory plays a key role in the diagnosis and treatment of cutaneous lymphoma.

Translational research is fundamental to the laboratory and they are continually working towards introducing new molecular tests to aid in the diagnosis, prognosis and treatment of cutaneous lymphoma.

7.0 Diagnosis and Management

7.1 Staging

Staging Classification Description

7.1.1 Stage TNM

IA 1 0 0
IB 2 0 0
IIA 1–2 1 0
IIB 3 0–1 0
III 4 0–1 0 IVA
1–4 2–3 0 IVB
1–4 0–3 1

T: Skin
T0 Lesions clinically and/or histopathologically suggestive of CTCL
T1 Limited plaques, papules, or eczematous patches covering <10% of skin surface
T2 Generalized plaques, papules, or erythematous patches covering ≥10% of skin surface
T3 Cutaneous tumours
T4 Generalized erythroderma

N: Lymph Nodes
N0 No palpable lymphadenopathy, lymph node pathology negative for CTCL
N1 Palpable lymphadenopathy; lymph node pathology negative for CTCL
N2 No palpable lymphadenopathy, lymph node pathology positive for CTCL
N3 Palpable lymphadenopathy, lymph node pathology positive for CTCL

M: Viscera
M0 No visceral organ involvement
M1 Visceral organ involvement, pathology present
B: Blood
B0 Atypical circulating cells not present (<5%)
B1 Atypical circulating cells present (≥5%)

Adapted from Bunn PA Jr, Lamberg SI. Report of the Committee on Staging and Classification of Cutaneous T-cell Cutaneous lymphomas. Cancer Treat Rep. 1979;63:725-728.

7.2 Investigations

Recommendations: Initial assessment
Repeated skin biopsies (ellipse rather than punch) are often required to confirm a diagnosis of CTCL
- Histology, immunophenotypic and preferably TCR gene analysis should be performed on all tissue samples (ideally molecular studies require fresh tissue)
- All patients (with the possible exception of early stage mycosis fungoides (stage IA) and cutaneous lymphomatoid papulosis) should ideally be reviewed by an appropriate MDT for confirmation of the diagnosis and to establish a management strategy
- Initial staging CT scans are required in all patients with the exception of those with early stages of mycosis fungoides (stage IA/IB) and cutaneous lymphomatoid papulosis
- At diagnosis peripheral blood samples should be analysed for total white cell, lymphocyte and Sézary cell counts, serum LDH, liver and renal function, lymphocyte subsets, CD4/CD8 ratios, HTLV-I serology and, preferably, TCR gene analysis
- Bone marrow aspirate or trephine biopsies may be required for CTCL variants (with the exception of cutaneous lymphomatoid papulosis) and may also be appropriate for those with late stages of mycosis fungoides (stage IIB or above). Grade A/level III

Note: Imaging guidelines can be located in the KMCC agreed document located on the KMCC website on the following link: http://kmcc.nhs.uk/tumour-sites/sub-groups-or-cross-cutting-groups/diagnostics-group/

The designated Supranetwork MDT will act as the "gate-keepers" for adjuvant therapy.

The Kent and Medway Skin Cancer TSSG will only support adjuvant therapies undertaken in the context of current controlled clinical trials. Such treatments will only be undertaken in the context of designated Supranetwork MDTs.

8.0 Treatment

8.1 Definitive Treatment

Once the diagnosis is made, medical management will be dictated by the exact tumour type. Many patients with CTCL of the mycosis fungoides type require only emollients and occasional use of potent topical corticosteroids ointment for patch disease.

The disease usually follows a chronic relapsing course. Widespread disease frequently responds to PUVA. This is available in both skin departments in Kent.

Development of plaques can be managed in the same way as patch stage and can induce regression of lesions. Radiotherapy and topical nitrogen mustard may be required on occasion. These treatments are more commonly used for the tumoural stage of the disease. Therapies such as total body superficial electron beam (TBSEBT), oral chemotherapeutic and combination regimes are reserved for disease unresponsive to the above measures.

Patients with Sézary syndrome usually require more intensive therapies, including extra corporeal photopheresis (ECP) which is only available at specialist centres. Pruritus can be severe in this type of CTCL. Infections are more common and require early treatment with antibiotics where appropriate:

Skin-directed therapy (topical therapy, superficial radiotherapy and phototherapy) is appropriate treatment for patients with early stages of mycosis fungoides (stages IA–IIA) with the choice of therapy dependent on the extent of cutaneous disease and plaque thickness (Grade A／level I)

Combined PUVA and a-interferon therapy can be effective for patients with resistant early-stage disease (stage IB–IIA) (Grade A／level IIi)

Patients with later stages of mycosis fungoides (stage IIB or higher) will require some form of systemic therapy PUVA ± a-interferon, ECP ± a-interferon, methotrexate, TSEB, bexarotene, denileukin diftitox, chemotherapy, alemtuzumab (Grade A／level IIii)

CTCL is a very radiosensitive malignancy and several fractions (2–3) of low energy (80–120 kV) superficial radiotherapy are appropriate for many patients (Grade A／level IIii)

Chemotherapy regimens in advanced stages of mycosis fungoides generally achieve complete responses in the region of 30% but these are short-lived (Grade B／level IIii)

Erythrodermic CTCL patients should be considered for immunotherapy and ECP as responses to chemotherapy are generally poor (Grade A／level IIii)

TSEB therapy is an effective treatment for stage IB and stage III mycosis fungoides but is not sufficient alone for stage IIB disease or those with significant haematological involvement (Grade A／ level IIi)

New agents such as bexarotene and denileukin diftitox offer important therapeutic alternatives which are currently being evaluated (Grade A／level IIii)

In treatment-resistant cases of late stage disease palliative radiotherapy and／or chemotherapy may produce a significant short-term benefit but the patient's quality of life should always be given priority (Grade B Level III)

All patients and especially those with late stages of disease (> IIA) should be considered for entry into well designed randomised controlled clinical trials.

Strength of recommendations

A There is good evidence to support the use of the procedure.
B There is fair evidence to support the use of the procedure.
C There is poor evidence to support the use of the procedure.
D There is fair evidence to support the rejection of the use of the procedure.
E There is good evidence to support the rejection of the use of the procedure.

Type of evidence

I Evidence obtained from at least one properly designed, randomized controlled trial. **II-i** Evidence obtained from well-designed controlled trials without randomization
II-ii Evidence obtained from well-designed cohort or case–control analytic studies, preferably from more than one centre or research group.

II-iii Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments (such as the introduction of penicillin treatment in the 1940s) could also be regarded as this type of evidence.
III Opinions of respected authorities based on clinical experience, descriptive studies or reports of expert committees.
IV Evidence inadequate owing to problems of methodology (e.g. sample size, or length or comprehensiveness of follow-up or conflicts of evidence).

8.2 Treatment by Supranetwork MDT

- As per their expert recommendations – will be communicated to the referring clinician.
- Shared care with locally administered treatment where possible is the aim.

9.0 Children and Young People

Children and Young People (CYP) with Cutaneous Lymphoma will be treated in accordance with principles set out in the CPY IOG.

All children and Young People up to the age of 18 must be referred to the CYP Principle Treatment Centre, which for Kent & Medway is based in the Royal Marsden Hospital.

All Young People between 16 and 24 years of age must be offered a referral to the CYP Treatment Centre.

Referral to a CYP Principle Treatment Centre does not necessarily mean that treatment will be undertaken at that centre shared care management protocols may allow some treatments to be undertaken locally.

10.0 Oncology Provision

The Non-Surgical Oncological management of all patients with cutaneous lymphoma as defined by the Non-Surgical Oncology Sub Group (NOG) of the Skin Cancer TSSG is set out in the document **"The Oncological Treatment of Skin Cancer".**

Note: *This is located on the KMCC website:*

http://kmcc.nhs.uk/tumour-sites/tumour-site-specific-information/dermatology-tssg/

11.0 Follow Up

BAD guidelines exist for the management and follow-up of patients diagnosed with cutaneous lymphoma.

The key purpose of follow up is patient education
- There is good evidence that educated patients detect their own recurrence
- Patients who do detect recurrence should be fast tracked back to the MDT
- Patients will have access to a clinical nurse specialist for support and advice
- Patients will have access to appropriate information and should always be given relevant "discharge" information

Long-term follow up should be undertaken at the discretion of clinicians and only after discussion and in agreement with patients. Follow up could be nurse led.

When the MDT has agreed that adjuvant therapy is appropriate, follow up will be “tailor-made” to suit the needs of the patient.

The Skin Cancer TSSG will review this pathway of care annually or when new evidence becomes available – whichever is soonest. The Skin Cancer TSSG will also audit adherence to this pathway of care.

12.0 Supportive and palliative Care

12.1 Prognosis

Prognosis in mycosis fungoides (and clinical variants) is related to age at presentation (worse if > 60 years), to the stage of the disease and possibly to the presence of a peripheral blood T-cell clone; some mycosis fungoides clinical variants may have a better prognosis

In Sézary syndrome the median survival is 32 months from diagnosis

Primary cutaneous CD30+ lymphoproliferative disorders without peripheral nodal disease have an excellent prognosis (range 96–100% 5-year survival)

The prognosis of other types of CTCL is generally poor with the frequent development of systemic disease.

12.2 Care Pathway

All patients with cutaneous lymphoma will have access to appropriate specialist palliative care and support at every stage of the patient journey.

Open and frank discussions with patients should take place with patients at all stages of their journey so that patients are not confused about their prognosis or have unrealistic expectations of any of the forms of treatment offered to them.

Relatives and carers will need to be supported and given appropriate information. However, in accordance with the recommendations set out in other Improving Outcomes Guidance, relatives and carers should not be given information different to that given to the patient.

Frail and terminally ill patients with cutaneous lymphoma should always be discussed with the specialist palliative care team.

Palliative care provision should available for all patients:

Hospital teams, including the Clinical Nurse Specialists for cutaneous lymphoma patients Primary Health Care Team would provide for palliative care at home

General Practitioner should be informed within 24 hours of the diagnosis and treatment plan.

The management of symptoms, psychological, social and spiritual issues, the communication of the diagnosis, and any associated problems, should be within the domain of all health care professionals.

Referral to specialist palliative care services should be considered when these issues have not been resolved and in particular for patients with:

Complex symptom management issues

Difficulties in adjusting to the diagnosis or disease progression

Psychological and family issues – such as communication problems within the family

Spiritual issues – such as the challenging of belief system/faith/cultural values as a result of the cancer.

Consideration of specialist palliative care or support should be given throughout the patient pathway, particularly:

At the Multi-disciplinary Team Meeting
When no active treatment is considered After active treatment
At relapse
In the terminal stages

13.0 Personnel and Contact information

A comprehensive, up to date list of MDM contact details can be found on the KMCC website via the following link: http://kmcc.nhs.uk/tumour-sites/terms-of-reference/

14.0 Glossary

Acronyms in common use throughout KMCC documentation:-

CNB	Cancer Network Board
CYP	Children and Young People (in relation to the IOG)
DCCAG	Diagnostic Cross Cutting Advisory Group
DOG	Disease Orientated Group (NSSG/TSSG/TWG)
DVH	Darent Valley Hospital
EK	East Kent
EKHUFT	East Kent Hospital University Foundation Trust
HoP	High Level Operational Policy
IOSC	Improving Outcomes: A Strategy for Cancer
K&C	Kent and Canterbury Hospital, Canterbury (EKHUFT)
KCH	Kings College Hospital
KMCN	Kent and Medway Cancer Network
KMCC	Kent & Medway Cancer Collaborative
KMCRN	Kent and Medway Cancer Research Network
LSESN	London and South East Sarcoma Network
MFT	Medway Foundation Trust
MTW	Maidstone and Tunbridge Wells NHS Trust
NOG	Non- Surgical Oncology Group *(Permanent oncologist sub group of the TSSGs with specific responsibility for chemo/rad pathways and advice to the TSSG, KMCC and geographical locations on new drugs).*
PoC	Pathway of Care *(KMCC agreed disease site specific clinical guidelines)*
QEQM	Queen Elizabeth the Queen Mother Hospital, Margate (EKHUFT)
QoL	Quality of Life
RAT	Research and Trials Group *(Permanent sub-group of the TSSGs with a specific responsibility for taking forward the clinical trials agenda).*
RMH	Royal Marsden Hospital
RNOH	Royal National Orthopaedic Hospital
QVH	Queen Victoria Foundation Trust Hospital, East Grinstead
UCLH	University College Hospital London
WHH	William Harvey Hospital, Ashford (EKHUFT)
WK	West Kent

15.0 Documentation and Administration

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