Self-assessment corner

A rapidly enlarging keratinised ‘skull-cap’

LM Samuel, LM Matheson

A 73-year-old woman presented with a large mass on her scalp. It had appeared and rapidly enlarged over a number of weeks. She had a little discomfort and no systemic symptoms of note. There was no alopecia and there were no other lesions. The mass was biopsied and a computed tomography (CT) scan was performed to assess the depth of penetration (figure).

Questions

- 1 What is the most likely diagnosis?
- 2 List three other differential diagnoses.

Department of Clinical
Oncology, Western
General Hospital,
Crewe Road,
Edinburgh EH4 2XU,
UK

LM Samuel
LM Matheson

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Figure CT scan

Answers

QUESTION 1

An isolated rapidly enlarging keratinised lesion has to be a squamous carcinoma until proven otherwise.

QUESTION 2

The differential diagnoses are keratoacanthoma, cutaneous metastasis, basal cell carcinoma, Merkel cell tumour, plaque psoriasis and kerion from tinea capitis.

Although keratoacanthoma is possible, it would be difficult in a lesion of this size to exclude an invasive component of squamous carcinoma. A cutaneous deposit from a rapidly proliferating cancer such as leukaemia, lymphoma, small cell carcinoma or even Kaposi's sarcoma is possible, though a thick keratin cap in association with any of these would be unusual. Both basal cell carcinoma and Merkel cell tumour are possibilities but they do not usually enlarge rapidly over a number of weeks. Plaque psoriasis and a kerion can be scaly, but are usually associated with alopecia. One might also expect to find other lesions in psoriasis, and some itch in tinea capitis.

To our surprise, mycosis fungoides was the diagnosis from the biopsy. This would make the case a 'd'emblee' variant, presenting with *de novo* tumour. The CT scan (figure) confirmed the limited penetration of a cutaneous lesion, and highlighted the thickness and density of the keratin 'cap'. She was treated with megavoltage electron therapy which has the advantage of high surface dose but limited dose at depth. Subsequently the keratin cap required removal under a general anaesthetic and a skin graft.

Discussion

Mycosis fungoides was first described by Alibert in 1806, who had a patient with skin tumours resembling mushrooms. These lesions waxed and waned over about five years before the patient died from a 'fever'.

Mycosis fungoides is the best known member of a heterogeneous group of cutaneous T-cell non-Hodgkin's lymphomas. Su *et al*¹ have produced a clinico-pathological classification of cutaneous lymphomas (box 1). In common with all non-Hodgkin's lymphomas the incidence of mycosis fungoides is increasing, with about 1000 new cases per year now in the US. Most cases are diagnosed in their sixth decade, and the male:female ratio is 2:1.²

Clinical features and the different phases of mycosis fungoides

Clinical features (pre-mycotic phase)

- eczematous skin rashes for up to 10 years
- variable size and shape of patch-like lesions, often bathing trunk distribution
- may be pruritic and fail to respond to standard topical eczema therapies
- skin biopsies are often nonspecific

Clinical features (plaque phase)

- lesions become scaly and are sharply demarcated
- the shape is often annular but can be quite bizarre
- plaques may become ulcerated or exophytic

Clinical features (tumour phase)

- may arise in pre-existing plaques
- tumours usually reddish brown or purple
- may present with *de novo* tumours
- predilection for the face and body folds (eg, leonine facies)
- often ulcerate with subsequent secondary infection
- may transform into a high-grade lymphoma, with visceral and lymph node involvement

Box 2

Mycosis fungoides arises from helper T-cells (ie, CD4+ve). Genetic analysis of T-cell receptor gene rearrangements have confirmed the clonality of these lymphomas.³ With polymerase chain reaction technology it may be possible to detect clonality from skin biopsies in the early pre-mycotic phase of the disease which would aid diagnosis. The clinical features of the three different phases of mycosis fungoides are given in box 2. As was the case with our woman, patients can present with *de novo* tumours.

In a patient with suspected mycosis fungoides it is important to examine the skin carefully. Consultation with the dermatopathology team is essential before performing a biopsy to ensure that the maximum diagnostic information can be obtained (ie, histology, immunophenotyping, molecular genotyping, and electron microscopy). Other investigations to perform include a chest X-ray, full blood count and film for Sezary cells, electrolytes and liver function tests. An excision biopsy of any palpable lymphadenopathy should be considered, particularly as lymph node involvement correlates with prognosis.⁴ The staging system (box 3) is quite complex, but may give an indication of prognosis.

Clinicopathological subtypes of cutaneous T-cell lymphomas

Type	Cutaneous T-cell lymphoma (NHL)	Prognosis
I	mycosis fungoides, or 'classical'	chronic course
II	primary large cell NHL (ki-1+ve or -ve)	chronic course
III	primary angio-invasive	chronic course
IV	adult T-cell leukaemia/lymphoma (HTLV-1 associated)	poor outlook
V	secondary involvement by a peripheral T-cell NHL	poor outlook

NHL= non-Hodgkin's lymphoma

Box 1

The clinical staging of mycosis fungoides		
Stage	Clinical features	Median survival
I	generalised plaque, no extracutaneous disease	>12 years
II	cutaneous tumours, erythroderma, or lymph node positive	about 5 years
III	extracutaneous disease, involving viscera	<3 years

Box 3

Mycosis fungoides: therapeutic options
<i>Treatment (local)</i>   • topical: simple emollients (incl steroids); chemotherapy (nitrogen mustard)   • photochemotherapy: PUVA (oral agent, then total skin UVA exposure); photophoresis (UV to blood <i>ex vivo</i>)   • irradiation: megavoltage (MeV) electron beams (involved field); MeV electron beams (wide field, eg, total body); MeV photon beams (involved field, eg, lymph nodes)   <i>Treatment (systemic)</i>   • chemotherapy: single agent (eg, chlorambucil); combination (eg, COP regimen); biological response modifiers (eg, interferon)   • experimental: monoclonal antibody therapy; retinoids; cyclosporins

Box 4

Mycosis fungoides is not curable with current therapies but useful palliation can be

achieved. Various treatment options are available (box 4); the choice is clearly dependent on the clinical stage and status of the patient as well as the availability of the treatments.^{2,4}

As is the case with low grade non-Hodgkin’s lymphomas, ‘localised’ treatments or simple chemotherapy can keep patients well and symptom-free for a number of years. Though combination chemotherapy can achieve higher complete response rates, there is no evidence the long-term survival is increased.^{2,4} However, in the 10–20% of patients whose disease transforms to a high-grade lymphoma, more intensive therapy is required. As in other areas of cancer medicine, the development of more effective (in the long term) therapies are awaited.

Final diagnosis

Mycosis fungoides associated with a dense keratin ‘cap’.

Keywords: mycosis fungoides, computed tomography

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A complication of self-poisoning

CH Gerrand, MR Reddy, MA Waldram, M Simms

South Birmingham
Trauma Unit, The
General Hospital,
Birmingham B4 6NH,
UK
CH Gerrand
MR Reddy
MA Waldram

Selly Oak Hospital,
Birmingham B29 6JD,
UK
M Simms

A 30-year-old man presented 20 hours after taking 450 mg flupenthixol and an unknown quantity of methixene tablets. He was semi-conscious. His left arm and chest were blistered, oedematous and erythematous.

Questions

- 1 What is the underlying condition?
- 2 With which systemic disorder is it associated?
- 3 What investigations are appropriate?
- 4 How should it be managed?

Figure 1 Left arm