The clinical staging of mycosis fungoides

<table>
<thead>
<tr>
<th>Stage</th>
<th>Clinical features</th>
<th>Median survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>generalised plaque, no extracutaneous disease</td>
<td>&gt;12 years</td>
</tr>
<tr>
<td>II</td>
<td>cutaneous tumours, erythroderma, or lymph node positive</td>
<td>about 5 years</td>
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<tr>
<td>III</td>
<td>extracutaneous disease, involving viscera</td>
<td>&lt;3 years</td>
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</tbody>
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Box 3

**Mycosis fungoides: therapeutic options**

**Treatment (local)**
- topical: simple emollients (incl steroids);
- chemotherapy (nitrogen mustard)
- photochemotherapy: PUVA (oral agent, then total skin UVA exposure); photopheresis (UV to blood *ex vivo*)
- irradiation: megavoltage (MeV) electron beams (involved field); MeV electron beams (wide field, eg, total body); MeV photon beams (involved field, eg, lymph nodes)

**Treatment (systemic)**
- 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**

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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
Answers

QUESTION 1
The underlying condition is compartment syndrome, involving all the compartments of the arm and forearm.

QUESTION 2
Where there is rhabdomyolysis, there can be an associated crush syndrome.

QUESTION 3
Measurement of intracompartmental pressure, serum electrolytes, blood gases and a full blood count are appropriate.

QUESTION 4
Immediate medical management in an unconscious patient should be directed at maintaining an airway and supporting the circulation. Hypovolaemic shock and oliguria may complicate crush syndrome. Raised intracompartmental pressure should be treated by urgent decompressive fasciotomy. Underlying muscle necrosis may necessitate amputation.

Learning points

- skin changes after unconsciousness may indicate raised compartment pressure
- consider coexisting crush syndrome

Erythema and blistering occur in areas which have been directly compressed. There may be raised compartment pressure under these skin changes; this appearance can be mistaken for a burn, infection or venous thrombosis and delay in diagnosis is common.

A conscious patient with compartment syndrome typically presents with pain disproportionate to the clinical situation, pain on passive movement of affected muscles, altered sensation distally and tenseness of the involved compartment. Pulses are usually normal. These features are masked in an unconscious patient.

Compartment pressure can be measured using a split central venous catheter attached to a pressure transducer. Commercial devices specifically designed for this purpose are available. Although it has been suggested that a compartment pressure of 30 mmHg or more demands fasciotomy, it is likely that the difference between compartment pressure and systolic blood pressure is the most important determinant of tissue perfusion. Where this is less than 30 mmHg, urgent fasciotomy is indicated.

Crush syndrome, first described in the London Blitz, is the systemic manifestation of rhabdomyolysis. There is myoglobinemia, hyperphosphatemia, hyperkalaemia, hyperuricemia, metabolic acidosis, intravascular volume depletion and coagulation defects. Acute renal failure may follow.

It is imperative to exclude compartment syndrome in all patients with limb symptoms following an overdose. Successful management depends upon early recognition and urgent decompressive fasciotomy.

Final diagnosis
Compartment syndrome.

Keywords: compartment syndrome, crush syndrome, self-poisoning, fasciotomy