Unexplained inflammation and deformity of the foot

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***Hospital Medicine* is delighted to introduce the first in an occasional series of teaching case reports. We hope that readers will find these educational and useful for their own professional development and also for teaching purposes.**

CASE REPORT

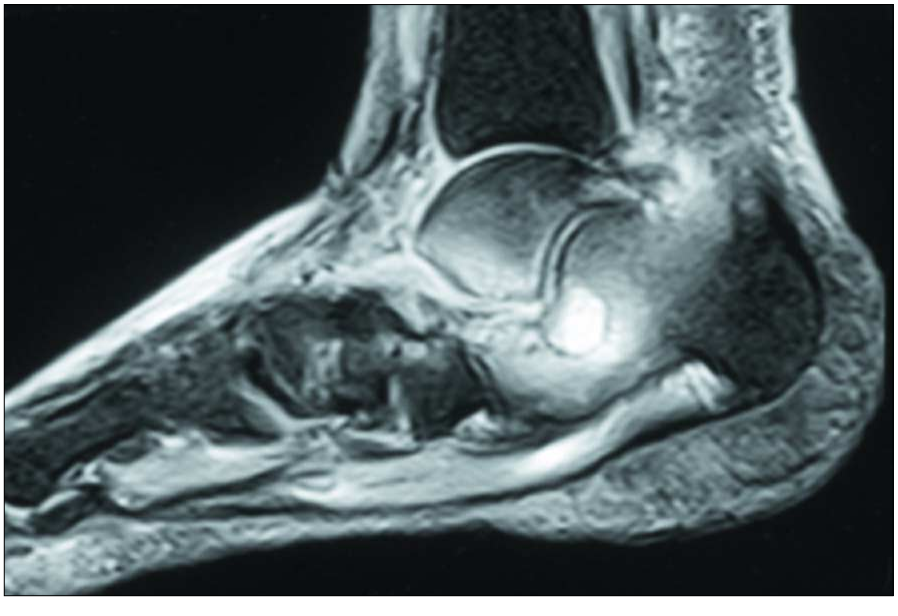
A 64-year-old Caucasian man was referred to a specialist diabetic foot clinic with unexplained deformity of the left foot. The problem had started an indeterminate time earlier – possibly 2 weeks. There was no history of trauma or of any underlying significant medical problem. The foot was displaced laterally, suggesting distortion of the tarsal bones. The skin was intact. There was generalized inflammation with swelling of the lower leg, and the skin was noticeably warmer than on the other side. There was some discomfort but no frank pain. Peripheral circulation was intact but there were signs of distal symmetrical sensory neuropathy, with loss of soft touch, pin prick and vibration sense. Ankle jerks were missing. Proprioception and deep pain sensation were preserved. There were no signs of other neurological or systemic disease, including sepsis, and he was taking no medication. Plain X-ray of the foot confirmed fracture dislocation of the calcaneum, and there was loss of fat signal, with enhancement of T2-weighted images on magnetic resonance imaging (*Figure 1*). Diabetes was excluded by 75 g glucose tolerance test.

1. What is the most likely diagnosis?
2. What specific causes should be excluded?
3. What is the main principle of management?
4. What additional pharmacotherapy might be considered?
5. How can the condition be differentiated from osteomyelitis?

INTRODUCTION

A doctor will not be able to diagnose a condition unless it is first suspected, and suspicion depends on expectation. If a doctor has never been taught to expect certain disorders, then he/she will never consider them in routine clinical practice. This is illustrated by the present case – in which an uncommon, but not rare, disorder would not normally be considered by those managing the patient. Failure to make the correct early diagnosis of this particular condition can have catastrophic consequences.

Figure 1. T2-weighted magnetic resonance image showing increased signal most marked in the calcaneum.



COMMENTARY

For any person presenting with spontaneous fracture and/or dislocation of the foot (or, less commonly, the leg) in association with distal symmetrical neuropathy, the most likely diagnosis is Charcot neuropathic osteoarthropathy (Charcot foot). In industrialized countries the commonest cause by far is diabetes. The other classical causes are leprosy, tabes dorsalis, spinal cord injury and syringomyelia, but the cause of the neuropathy may be obscure, as proved to be the case in this patient (Jeffcoate et al, 2000; Rajbhandari et al, 2002).

The pathogenesis of this condition seems to centre on disturbed microvascular circulation, with arteriolar-venular shunting, and associated osteopenia. These processes have been well described in diabetic neuropathy, and the condition has a number of features in common with reflex sympathetic dystrophy (RSD; otherwise known as complex regional pain syndrome-1, CRPS-1) (Jeffcoate et al, 2000). The deformity develops as a result of fractures which occur within the weakened bone, leading to increased loading on adjacent bones, further fractures and dislocation, leading potentially to progressive distortion of the structure of the foot (*Figure 2*). Even though loss of protective sensation is not usually the primary cause, the relative lack of pain can deceive both the patient and the doctor into thinking that the problem is confined to the soft tissues. Early diagnosis is essential because all loading must be taken off the foot to prevent further bone damage.

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Figure 2. Generalized painless inflammation of the lower part of the leg, with distortion of the tarsal bones and lateral displacement of the forefoot.

If there is associated ulceration of the skin (arising from unnoticed trauma and/or increased loading caused by the altered shape off the foot), it can be impossible to exclude osteomyelitis either as a primary cause or a complication. Systemic signs of infection are usually missing, and the results of investigations, including magnetic resonance imaging, are generally unhelpful. The only specific test which may be diagnostic is a labeled leucocyte scan, but this can produce both false positive and false negative results.

MANAGEMENT

The foot must be immobilized – preferably in a total contact cast – and the patient urged not to put any weight on it. Effective weight-sparing may need to be maintained for many months. The use of intravenous bisphosphonate has been shown to reduce discomfort, skin temperature and markers of bone turnover (as in RSD), and although it has not been shown to have any long-term benefit, it is used by many in the field (Jude et al, 2001). Although unlikely, there is a theoretical risk that the use of bisphosphonates may impair reossification. The condition recurs in the contralateral foot in 20% of cases. With or without bisphosphonates the condition eventually resolves, the bones enter a sclerotic phase and the signs of inflammation resolve (*Figure 3*). The patient can return slowly to weight bearing, and fitted footwear should be provided to reduce the risk of ulceration. It is possible that the isolated metatarsal fractures which can occur in neuropathies are a manifestation of the same process. HM

Figure 3. The appearance of the limb after 4 years, showing resolution of the inflammation but with residual distortion of the hindfoot.

Jeffcoate W, Lima J, Nobrega L (2000) The Charcot foot. *Diabet Med* **17:** 253–8

Jude EB, Selby PL, Burgess J et al (2001) Bisphosphonates in the treatment of Charcot neuroarthropathy: a double-blind randomised controlled trial. *Diabetologia* **44:** 2032–7

Rajbhandari SM, Jenkins RC, Davies C, Tesfaye S (2002) Charcot neuroarthropathy in diabetes mellitus. *Diabetologia* **45:** 1085–96

TEACHING POINTS

- Charcot foot can complicate any cause of distal symmetrical neuropathy.
- Enforced weight sparing is essential while the bones remain weak.
- The diagnosis should be excluded in any person with diabetes and neuropathy who presents with redness and swelling of the foot.
- In the absence of a significant history the diagnosis is much more likely than sprain, cellulitis or deep venous thrombosis, and the consequences of missing it are considerable. The patient should be assumed to have a Charcot foot, and managed accordingly, until proved otherwise.
- The diagnosis of acute Charcot is frequently missed in emergency departments and fracture clinics, causing irreversible distortion of the foot, ulceration and secondary osteomyelitis leading, often, to below knee amputation.