Plain radiographs can rarely support a diagnosis of Meckel's diverticulum and contrast studies are often unrewarding (Miller *et al.* 1981). It was hoped that radio-pertechnetate scans would provide a reliable method of detection, but high false negative and positive results have been reported (Miller *et al.* 1981).

The preoperative diagnosis of complicated Meckel's diverticulum remains difficult and the long delay in arriving at an accurate diagnosis in our case partly reflects this.

Acknowledgment: We thank Mr W G Everett, Consultant Surgeon, Addenbrooke's Hospital, for permission to report this case.

References

Farthing M G
(1981) *British Journal of Surgery* **68,** 176
Grant A B F
(1981) *Australian and New Zealand Journal of Surgery* **51,** 77–78
Leslie M D
(1983) *British Journal of Surgery* **70,** 244
Miller K B, Naimark A, O'Connor J F & Bouras L
(1981) *Gastrointestinal Radiology* **6,** 209–215
Soltero M J & Bill A M
(1976) *American Journal of Surgery* **132,** 168–173
Spiro H M
(1977) Clinical Gastroenterology. 2nd edn. Macmillan, New York; p 419
Williams R S
(1981) *British Journal of Surgery* **68,** 477–480

Elephantine sarcoidosis presenting as ulcerating lymphoedema[1]

M F Mühlemann MB MRCP **N P J Walker** BSc MRCP
L B Tan DPhil MRCP **R H Champion** MB FRCP
Department of Dermatology
Addenbrooke's Hospital, Cambridge CB2 2QQ

Ulceration is an unusual but recognized manifestation of cutaneous sarcoidosis (Neill *et al.* 1984). Elephantiasis, with or without ulceration, is a much rarer manifestation with apparently only two reported cases (Ravindra Nathan *et al.* 1974, Sézary 1942). We report a patient who presented with grossly lymphoedematous legs and persistent ulcers which histologically showed non-caseating granulomata. Thorough investigation of the patient failed to show any unequivocal evidence of systemic involvement.

[1]Case presented to Section of Dermatology, 20 October 1983. Accepted 9 October 1984

Case report

A 59-year-old housewife developed non-pitting swelling of her right leg ten years prior to admission. For one year she had had intermittent ulceration of her left hallux, spreading slowly to involve the medial aspect of the left sole. Six months prior to admission the left leg, previously unaffected, became more swollen and the numerous small ulcers became rapidly worse and painful, spreading to the entire left foot and ankle. Apart from the cutaneous symptoms, she gave a history of exertional dyspnoea and non-irritating dryness of her eyes. Physical examination on admission showed bilateral 'elephantine' lymphoedema of the legs and close clusters of ulcers on the left foot and ankle (Figure 1). There were small erythematous macules just distal to the left knee. At that stage these did not show clinical evidence of infiltration. General examination was normal with no lymphadenopathy.

Swabs from ulcers grew *Staphylococcus aureus*, group B Streptococci, Coliforms and Pseudomonas. No fungi or mycobacteria were isolated from swabs or biopsies. Histological examination of biopsies from an ulcerated area and a cutaneous nodule, one of several which had developed around the left knee, showed non-caseating sarcoid-like granulomata. Mantoux 1 in 100 was negative, the Kveim test strongly positive. Chest X-ray, serum angiotensin-converting enzyme and pulmonary function tests were normal, but her IgG level was persistently elevated with oligoclonal banding. Gallium scan showed very slightly increased uptake, and therefore possible involvement, in the lacrimal and salivary glands, spleen and heart. Schirmer's test was slightly abnormal, conjunctival biopsy normal. A CT scan showed very slightly enlarged abdominal lymph nodes and a lymphangiogram was not done because the ulceration of the foot made it technically not feasible.

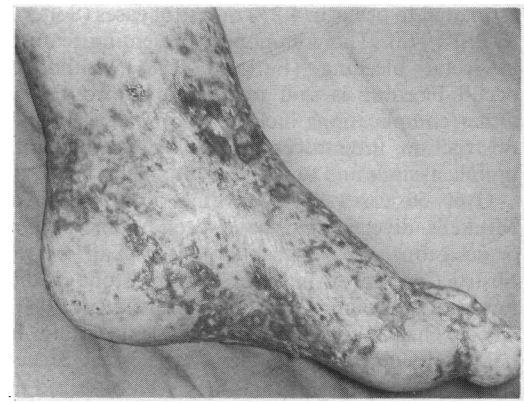

Figure 1. Extensive ulceration of oedematous left foot and ankle

0141–0768/85/030260–02/$01.00/0

The leg ulceration began to respond at first to rest, topical antiseptics and systemic antibiotics. Over many weeks some areas re-epithelialized but infiltrated dermal nodules became apparent. The nodules below the knee ulcerated and scar sites on the forearms became infiltrated. Further improvement over several weeks was disappointing until after the biopsy and the diagnosis of cutaneous sarcoidosis was made. Prednisolone 40 mg daily was started; the left leg oedema improved markedly in one month and the nodules flattened considerably. After six months this improvement has been maintained, but the ulcerating nodules continue to recur when the dose of prednisolone is reduced below 15 mg daily.

Discussion

When this woman presented we were interested in her unusual story of apparent asymmetrical enlargement of the lower legs and by the multiplicity and the chronicity of her ulcers. Despite the rarity of such cases, an initial diagnosis of chronic lymphoedema with secondary ulceration seemed to be supported by an initial improvement with bed rest, nursing care and antibiotics. Further progress then became extremely slow and alternative aetiologies were sought. The presence of non-caseating granulomata in the biopsy, at a time when clinically there were no granulomata, was a complete surprise. Thorough investigation has failed to confirm active sarcoidosis in any other organ though the gallium scan is suggestive. Subsequent introduction of systemic steroids led to the resolution of the infiltrate but only some decrease in size of the left leg and the right leg remains no less swollen than before.

In Sézary's (1942) case the patient had extensive organ involvement – skin, bone, lungs and lymph nodes – as well as asymmetrical lower limb oedema, more pronounced on the left, associated with erythematous lesions on the left shin. Unfortunately no further details of this patient are available but the published photograph shows no ulceration. In fact, ulceration is a rare feature of sarcoidosis with only 30 cases reported in the literature. The case reported by Ravindra Nathan *et al.* (1974) had persistent lymphoedema for almost 11 years before cutaneous and systemic manifestations of sarcoidosis became apparent. Their patient had multiple large ulcers on the legs, widely disseminated papular and nodular lesions and firm subcutaneous ulcerating nodules. Scar sites were infiltrated and there was evidence of eye involvement. Their patient responded dramatically to systemic steroids; the oedema had almost disappeared in two months.

There are marked similarities between these 3 patients, but what is not clear is whether the lymphoedema is a direct result of the sarcoidosis or whether this is an example of the Köbner phenomenon in patients with sarcoidosis and previously lymphoedematous legs. In our patient the sarcoidosis certainly presented in a most unusual way and the response to systemic steroids was dramatic. We believe that she is a third case of what Sézary called 'elephantine sarcoidosis'. Sarcoidosis should be considered as a cause of chronic, particularly asymmetrical, leg oedema with or without ulceration when obvious venous or lymphatic obstruction is absent.

References

Neill S M, Smith N P & Eady R A J
(1984) *Clinical and Experimental Dermatology* **9**, 277–279
Ravindra Nathan M P, Pinsker R, Chase P H & Elquezabel A
(1974) *Archives of Dermatology* **109**, 543–544
Sézary A
(1942) *La Semaine des Hopitaux de Paris* **18**, 45–52