

## Chronic dermal sinuses as a manifestation of histiocytosis X

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### Abstract

Two young patients presented with generalised lymphadenopathy, otorrhoea, otitis, and rash. Over the next few years chronically discharging sinuses began to form over enlarged nodes and histological appearances were typical of histiocytosis X. In neither case were micro-organisms isolated from the lesions, and in both patients healing occurred with immunosuppressive agents.

Chronic dermal sinus formation secondary to lymph node disease has never before been recorded as a manifestation of histiocytosis X. Histiocytosis X should therefore be considered in the differential diagnosis of "suppurative" lymphadenopathy so that appropriate treatment may be given without delay.

### Introduction

The clinical manifestations in histiocytosis X depend on the distribution of characteristic cellular infiltrates in the skin, bone, lymph nodes, and other organs.<sup>1</sup> We have recently seen two patients with chronic dermal sinus formation secondary to lymph node disease, which has not been reported in histiocytosis X.

### Case histories

*Case 1*—In September 1982 a 21 year old student presented with unilateral otorrhoea and deafness and lymphadenopathy in the supraclavicular, axillary, and inguinal regions. Over the next year he developed a persistent

mucoid discharge from dermal sinuses arising over enlarged supraclavicular and axillary nodes. Tests at his local hospital found no evidence of infection with mycobacteria, actinomyces, other fungi, or bacteria. Probing a supraclavicular sinus disclosed a track deep to sternomastoid. Biopsy of an axillary node showed "chronic inflammation." In August 1984 he was admitted to the John Radcliffe Hospital with chronically discharging dermal sinuses over the supraclavicular and axillary regions (figure) and a seborrhoeic rash over scalp, pinnae, and trunk. Appearances of tissue from the rash were typical of histiocytosis X.<sup>1</sup> Computed tomography showed a large erosion within the right mastoid process. The sinuses healed with prednisolone but later recurred when the dose was reduced. After adding vincristine the sinuses dried up and the mastoid lesion finally reduced in size.



Case 1. Chronically discharging dermal sinuses overlying supraclavicular nodes.

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*Case 2*—A 4 year old boy was referred to hospital because of persistent cervical, axillary, and inguinal lymphadenopathy; bilateral otitis externa and recurrent otorrhoea; and a macular rash over the buttocks. Biopsy of an inguinal node confirmed histiocytosis X. He was given prednisolone with a good initial response but six years later (1978) he again had generalised lymphadenopathy and multiple lytic lesions had appeared in the skull vault. Vinblastine was added to the prednisolone with some shrinkage of enlarged nodes. In 1979 a painful red indurated swelling appeared in the right posterior cervical triangle. There was massive adenopathy in both groins and

also in the left posterior triangle. The cervical "abscess" was drained. Histological appearances of tissue from the abscess wall were typical of histiocytosis X. After radiation the area healed but over the next three years there was intermittent mucopurulent discharge from the cutaneous sinus that had formed over the right occiput and right posterior cervical region. The problem was partially controlled by increasing the dose of prednisolone, and on two occasions there was a satisfactory response to etoposide. No organism was ever isolated.

### Discussion

Lymphadenopathy is well recognised in histiocytosis X<sup>2</sup> but formation of sinuses secondary to lymph node disease is hitherto unrecorded, even among 260 patients in three major studies.<sup>1, 3, 4</sup> Chronically discharging sinuses were the presenting feature in case 1 and occurred after many years of disease in case 2. These lesions should not be confused with ulcerating skin plaques, which are more superficial and unrelated to nodes. Bony erosions may also break through and form dermal sinuses, but in neither of our patients did bone lesions appear to underlie the affected skin.

Histiocytosis X should now be recognised in the differential diagnosis of "suppurative" lymphadenopathy so that appropriate treatment may be given. The diagnosis in case 1 was delayed for over a year because of the false impression of an infective process.

Similarly in case 2 with a patient receiving corticosteroids added infection with tuberculosis or actinomycosis was thought to be responsible. In neither case, however, was any micro-organism isolated, and in both patients healing occurred with immunosuppressive drugs.

Though spontaneous resolution of multisystem histiocytosis X is occasionally seen,<sup>5</sup> the response to steroids in this age group, as in young children, may be gratifying. Vincristine and etoposide are the most useful "second line" agents.

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### References

- 1 Nezelof C, Frileux-Herbert F, Cronier Sachat J. Disseminated histiocytosis X. *Cancer* 1979;44:1824-38.
- 2 Sajjad SM, Osborne B. Lymph node involvement by histiocytosis X. *Arch Pathol Lab Med* 1982;106:96-8.
- 3 Greenberger J, et al. Results of treatment of 127 patients with systemic histiocytosis. *Medicine (Baltimore)* 1981;60:311-37.
- 4 Avery M, et al. Course and prognosis of reticuloendotheliosis (eosinophilic granuloma, Schuller-Christian disease and Letterer-Siwe disease). *Am J Med* 1957;22:636-52.
- 5 Broadbent V, Davies EG, Heaf D, et al. Spontaneous remission of multi-system histiocytosis X. *Lancet* 1984;i:253-4.

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## Bone changes occurring spontaneously and caused by oestrogen in early postmenopausal women: a local or generalised phenomenon?

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### Abstract

Regional values of bone mineral content and bone mineral density were calculated from total body dual photon absorptiometry scans of 52 early postmenopausal women treated with oestrogen for one year and of 52 similar women treated with placebo. The six regions were head, arms, chest, spine, pelvis, and legs. In addition, bone mineral density of the lumbar spine was measured by dual photon absorptiometry and bone mineral content of the forearm by single photon absorptiometry, using separate special purpose scanners.

All regions were unchanged after one year of treatment with oestrogen, excluding the lumbar spine, for which values rose. Values for all regions except the lumbar spine fell significantly in the placebo group. The rates of loss ranged from 2% to 8%, with no significant differences among the regions.

It is concluded that loss of bone in the early menopause is a generalised phenomenon, affecting all parts of the skeleton. Furthermore, oestrogen prophylaxis for loss of bone is effective in all parts of the skeleton. Finally, it is suggested that the measurement of bone mineral content in the forearm should be used for clinical follow up of bone changes, as this method is superior to others in the ratio of change to precision.

### Introduction

We have developed a method for measuring total body bone mineral in vivo by dual photon absorptiometry.<sup>1</sup> This measurement has advantages over other estimates of bone mass. Firstly, it directly reflects the mineralisation of the whole skeleton and, secondly, it allows the bone mineral content of particular sites to be calculated.<sup>2, 3</sup> As the total skeleton and the bones in the distal forearm contain the same ratio of trabecular to cortical bone, the bone mineral content of the forearm is a good indicator of total body bone mineral in both normal subjects and patients with disorders of calcium metabolism.<sup>4, 6</sup>

It is not known whether the rate of bone mineral loss is the same in trabecular and cortical bone tissue or whether this loss occurs at the same rate in different parts of the skeleton.<sup>2, 7, 8</sup> The principal question, therefore, is where to measure bone mass for both follow up and diagnosis. It is also not known whether the effect of treatment with oestrogen is uniform throughout the skeleton.

The aim of this study was to compare spontaneous changes in different parts of the skeleton with those caused by oestrogen and to find a measuring site with optimum sensitivity for follow up in early postmenopausal women.

### Methods

Regional bone mineral content was measured by dual photon absorptiometry on a whole body scanner, originally developed in our laboratory to measure total body bone mineral.<sup>1</sup> The radiation source is  $3.7 \times 10^{10}$  Bq (1 Ci) gadolinium-153 with principal photo peaks at 44 keV and 100 keV. During the 90 minutes of scanning the subject lies supine with the source under the scanning table and the detector above. The subject is scanned in a rectilinear raster pattern with a transverse scan speed of 1 cm/s and longitudinal steps of 2.5 cm.<sup>1</sup> Total body bone mineral (and bone mineral content) is calibrated weekly against standard dry defatted bones. Bone mineral density is calculated by dividing by the projected skeletal area.<sup>3</sup> Calculation of the bone

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