4 Burry JN, Lawrence JR. Phototoxic blisters from high frusemide dosage. *Br J Dermatol* 1976;**94**:495-9
5 Leitao EA, Person JR. Bumetanide-induced pseudoporphyria. *J Am Acad Dermatol* 1990;**23**:129-30
6 Baker EJ, Reed KD, Dixon SL. Chlorthalidone-induced pseudoporphyria: clinical and microscopic findings of a case. *J Am Acad Dermatol* 1989;**21**:1026-9
7 Epstein JH, Tuffanelli DL, Seibert JS, Epstein WL. Porphyria-like cutaneous changes induced by tetracycline hydrochloride phtotosensitization. *Arch Dermatol* 1976;**112**:661-6
8 McDonagh AJG, Harrington CI. Pseudoporphyria complicating etretinate therapy. *Clin Exp Dermatol* 1989;**14**:437-8
9 Epstein JH. Pseudoporphyria and UVA suntan salons. *Photochem Photobiol* 1987;**45**(suppl):40S
10 Farr PM, Marks JM, Diffey BL, Ince P. Skin fragility and blistering due to use of sunbeds. *Br J Dermatol* 1988;**296**: 1708-9
11 Stenberg A. Pseudoporphyria and sunbeds. *Acta Dermatol Venereol (Stockh)* 1990;**70**:354-6
12 Harber LC, Bickers DR. Porphyria and pseudoporphyria. *J Invest Dermatol* 1984;**82**:207-9
13 Gilchrest B, Rowe JW, Mihm MC Jr. Bullous dermatosis of hemodialysis. *Ann Intern Med* 1975;**83**:480-3

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Pachydermoperiostosis mimicking acromegaly

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Keywords: pachydermoperiostosis; acromegaly; clubbing

Pachydermoperiostosis is a rare condition of unknown aetiology, in which the principal manifestations are digital clubbing and periostitis. Other abnormalities include coarsening of the facial features and spade-like enlargement of the hands and feet. These may give rise to appearances resembling acromegaly. To draw attention to this diagnostic pitfall, we describe a patient with pachydermoperiostosis who was misdiagnosed as acromegalic on several occasions. The clinician should be alerted to the true diagnosis by the presence of clubbing, despite the similarity of other somatic features.

Case report

A 38-year-old man was admitted through casualty complaining of painful, swollen ankles of uncertain duration. He smelt strongly of alcohol and little further history was forthcoming.

On examination he had a prominent jaw, coarse features (Figure 1), including a huge tongue, and massive hands and feet. He had clubbing of digits and tender swollen ankles without cellulitis or calf tenderness. Cardiovascular, respiratory and abdominal examinations were normal. Neurological examination revealed disorientation, but no focal neurological signs.

Acromegaly had been diagnosed at other hospitals initially in 1983, and subsequently in 1986, when he had presented with painful swollen ankles and sweating of the hands and feet. Investigations found to be normal at that time included: plain radiographs of the skull (including pituitary fossa), chest and hands, haematology, biochemistry, thyroid function tests, cortisol, glucose tolerance test and computed tomography of the brain. Ankle radiographs were reported to show periosteal reaction on the medial aspects of the tibiae. Despite these investigations acromegaly was diagnosed in view of his striking appearance, although no treatment was recommended. The finger clubbing was thought to be familial.

The acromegalic features were again noted in 1989 during an admission for psychiatric assessment. On this occasion computed tomography of the pituitary fossa was normal, and basal growth hormone levels were at the lower end of the normal range (0.5 mU/l; range 0-10.0 mU/l). Other basal pituitary hormone levels were also normal, with the exception of FSH, slightly elevated at 9.9 mU/l (range 0.5-5.0 mU/l). Testosterone levels were normal. Cytogenetic studies showed a normal karyotype.

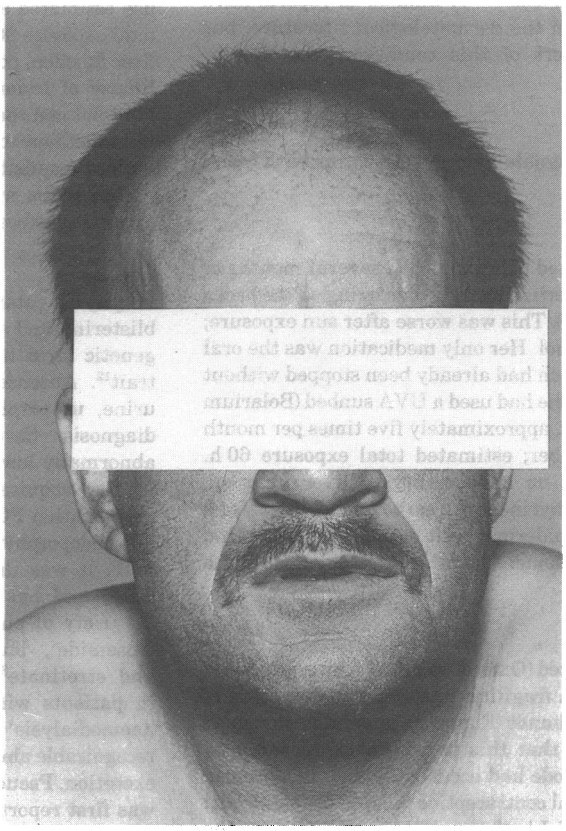

Figure 1. The patient's facial appearance

During the current hospital admission radiographs of the ankles showed a striking periosteal reaction bilaterally along the borders of the distal tibiae and fibulae (Figure 2). Other investigations included normal haematology and plasma biochemistry. Chest radiology showed cardiomegaly and normal lungs.

Pachydermoperiostosis was diagnosed because of the patient's appearance, the prolonged duration of the digital clubbing, and his recurrent symptoms from hypertrophic osteoarthropathy, possibly alcohol-related. The patient's symptoms settled spontaneously and he was able to be discharged well 48 hours later.

Discussion

This case illustrates how appearances of pachydermoperiostosis may mimic those of acromegaly and cause difficulty in differentiating between the two conditions.

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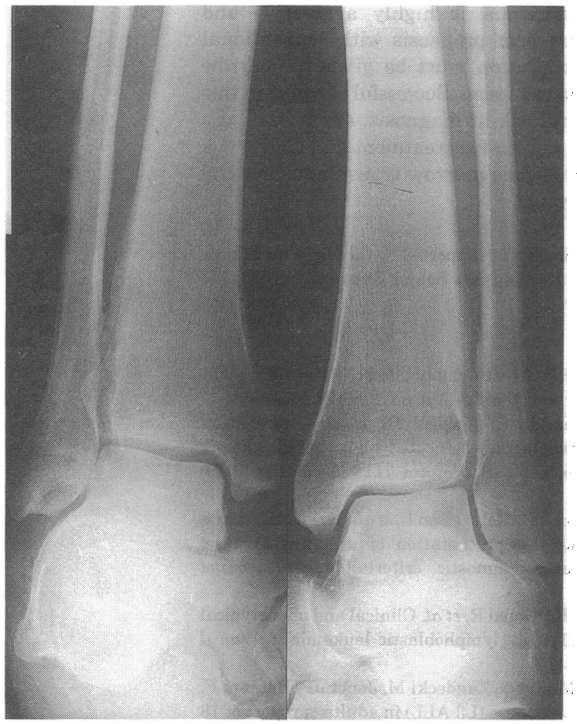

Figure 2. Ankle radiographs showing pronounced periosteal reaction at the distal end of the tibia and fibula bilaterally

In the patient reported an incorrect diagnosis was made at three separate hospitals in view of his striking appearance, despite the lack of supporting investigations. The clue to the correct diagnosis lay in the presence of hypertrophic osteoarthropathy with digital clubbing, which is not a reported feature of acromegaly.

Pachydermoperiostosis (Touraine-Solente-Golé syndrome) is an hereditary condition, apparently transmitted by an autosomal dominant gene with variable expression. Symptoms usually begin in teenage years, and boys are considerably more affected than girls[1]. Recognized clinical features illustrated by the reported case include the insidious onset of clubbing and enlargement of the hands and feet, with cylindrical thickening of the forearms and legs, of both bony and soft tissue origin[2]. Symptoms comprise reduced dexterity of the hands, and vague bone and joint pains[3], which may be exacerbated by alcohol[4]. Thickening and furrowing of the facial skin may give rise to a 'leonine' appearance, seborrhoea is common, and excess sweating, particularly of the hands and feet, may occur. Additional manifestations not evident in this case include intermittent joint effusions and acrolysis of the distal phalanges[5].

Diagnosis of pachydermoperiostosis clinically not only prevents excessively detailed endocrine investigation, but also makes an exhaustive search for an alternative cause of the patient's clubbing unnecessary. In conjunction with the clinical findings, other useful diagnostic pointers are the duration of clubbing, and the presence of a family history.

In conclusion, although both acromegaly and pachydermoperiostosis are infrequently encountered, avoidance of diagnostic confusion is important because of the prognostic and therapeutic implications. Awareness of the significance of clubbing under these circumstances is likely to prevent misdiagnosis.

References

1 Rimoin DL. Pachydermoperiostosis (idiopathic clubbing and periostosis): Genetic and physiological considerations. *N Engl J Med* 1965;272:923-31
2 Touraine A, Solente A, Gole L. Un syndrome osteodermatopathique. La pachydermie plicaturee avec pachyperiostose des extremites. *Presse Med* 1935;43:1820-24
3 Vogl A, Goldfischer S. Pachydermoperiostosis. Primary or idiopathic hypertrophic osteoarthropathy. *Am J Med* 1962; 33:166-87
4 Mueller M, Trevarthen D. Pachydermoperiostosis: arthropathy aggravated by episodic alcohol abuse. *J Rheumatol* 1981;8:862-4
5 Hedayati H, Barmad R, Skosey JL. Acrolysis in pachydermoperiostosis. *Arch Intern Med* 1980;140:1087-8

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Prolonged disease-free survival in an adult presenting with Burkitt-type acute lymphoblastic leukaemia and CNS disease

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Keywords: syngeneic bone marrow transplantation; Burkitt type acute lymphoblastic leukaemia, CNS disease

Burkitt-type acute lymphoblastic leukaemia (L3 ALL) is an uncommon disorder which carries a particularly poor prognosis[1]. We report long-term disease free survival in a 31-year-old woman with L3 ALL treated with conventional chemotherapy and syngeneic bone marrow transplantation.

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Case report

A 31-year-old previously well black woman presented with a 4-week history of swollen, bleeding gums and constitutional symptoms. Two weeks before admission she noticed diplopia and one week later discovered a right neck mass.

On examination she was an ill, thin, black woman with a right ptosis, swollen, tender gums and a firm, mobile, nontender 2×2 cm mass in the right anterior cervical chain. Neurological examination revealed bilateral Horner's syndrome, third and sixth cranial nerve palsies on the right, and a partial fifth cranial nerve palsy on the left.

Haemoglobin was 120 g/l, the white blood cell count 24.1×10^9/l, with 12% lymphoblasts with clefted nuclei and platelets were 46×10^9/l.

A marrow aspirate was packed and infiltrated with a homogeneous population of large blasts, typical of Burkitt's lymphoma, with deep blue cytoplasm and large round to oval nuclei with 1-2 prominent nucleoli. Intracytoplasmic and intranuclear vacuolation as well as nuclear clefts were present. Staining with Sudan Black, PAS and nonspecific esterase was negative. Cytogenetic analysis was normal.

Computerized tomography of the brain and a lumbar puncture were normal. All body fluid cultures were negative.

A diagnosis of ALL, Burkitt-type or L3, was made. Remission was successfully induced with vincristine, daunorubicin, prednisone and L-asparaginase[2]. Cranial irradiation (2400 cGy in 12 fractions) and six doses of intrathecal methotrexate 15 mg were also given. Neurological status was normal at discharge.

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