Letters to the Editor

Generally, our findings showing an association of medium-sized vessels in the dermis, consistent with CPAN. An ankle X-ray showed peristomial elevation and new bone formation affecting the tibia. MRI scan showed bilateral ankle effusions, marked tenosynovitis, disruption of the articular surfaces of both subtalar joints and a lesion in the metaphysis of the right tibia, thought to represent a medullary bone infarct possibly secondary to arteritis (Fig. 1). A synovial biopsy of the right ankle demonstrated extensive perivascular chronic inflammatory cell infiltrate with several foci of acute inflammation. There was a good initial response to prednisolone 30 mg daily, but a poor response to colchicine, hydroxychloroquine and dapsone. He was unable to tolerate azathioprine due to hypersensitivity and developed cushingoid side-effects on steroids. Treatment with cyclophosphamide 100 mg daily, prednisolone 7.5 mg daily and indomethacin 50 mg daily resulted in moderate improvement in his ankle pain and no recurrence of his cutaneous nodules.

Case 2 was a 51-yr-old male who presented in November 1994 with a 5-month history of tender erythematous nodules on both shins, and bilateral pitting ankle oedema. Active and passive movements were limited at both ankle joints. He subsequently developed a livedo pattern on both lower legs, and painful ulceration of his right shin. Laboratory investigations were unremarkable apart from an elevated ESR at 46 mm/h. Skin biopsy demonstrated a medium-vessel vasculitis.

**Cutaneous polyarteritis nodosa with seronegative arthritis**

Sir, Cutaneous polyarteritis nodosa (CPAN) is a localized cutaneous vascular disorder characterized by necrotizing arteritis of medium-sized vessels in the skin. We report two cases of CPAN associated with seronegative arthritis resulting in chronic pain and disability. Arthralgia has previously been reported in CPAN, but an association with arthritis has not been emphasized.

Case 1 was a 50-yr-old male who presented in November 1993 with a 4-yr history of recurrent tender erythematous papules, and an 8-yr history of arthralgia affecting his right ankle. Examination revealed a patchy violaceous eruption around his right ankle, papules on his foot, two ulcers over the lateral malleolus and bilateral pitting ankle oedema. Marked limitation of movement was noted at the ankle and subtalar joints.

Laboratory investigations were unremarkable apart from an elevated ESR at 72 mm/h. Skin biopsy demonstrated acute and chronic arteritis involving medium-sized vessels in the dermis, consistent with CPAN. An ankle X-ray showed peristomial elevation and new bone formation affecting the tibia. MRI scan showed bilateral ankle effusions, marked tenosynovitis, disruption of the articular surfaces of both subtalar joints and a lesion in the metaphysis of the right tibia, thought to represent a medullary bone infarct possibly secondary to arteritis (Fig. 1). A synovial biopsy of the right ankle demonstrated extensive perivascular chronic inflammatory cell infiltrate with several foci of acute inflammation. There was a good initial response to prednisolone 30 mg daily, but a poor response to colchicine, hydroxychloroquine and dapsone. He was unable to tolerate azathioprine due to hypersensitivity and developed cushingoid side-effects on steroids. Treatment with cyclophosphamide 100 mg daily, prednisolone 7.5 mg daily and indomethacin 50 mg daily resulted in moderate improvement in his ankle pain and no recurrence of his cutaneous nodules.
consistent with CPAN. Increased uptake affecting his left ankle was noted on bone scan. Treatment with dapsone 50 mg daily resulted in limited improvement, but dramatic improvement occurred following the introduction of oral prednisolone 20 mg daily. Azathioprine was commenced as a steroid-sparing agent. He subsequently developed a mononeuritis causing numbness along the lateral border of his left foot. On review 3 yr after diagnosis, he was well and off all medications.

CPAN is considered to be a distinct clinical entity characterized by a chronic prolonged course but an overall good prognosis [1]. In CPAN, systemic symptoms such as fever, myalgia, arthralgias, arthritis and peripheral neuritis may occur, but patients are usually normotensive and the lack of life-threatening organ involvement helps distinguish it from systemic PAN [1–3]. None of the 112 patients with CPAN reviewed went on to develop systemic disease during follow-up [1–3]. As in our cases, laboratory findings are usually unremarkable apart from an elevated ESR.

The characteristic cutaneous manifestations of CPAN are subcutaneous tender erythematous nodules. Areas in which groups of nodules occur often develop livedo reticularis. Nodules are unusual in systemic PAN and are, thus, a useful distinguishing feature. Lower limb ulceration is seen in up to 50% of patients with CPAN [1, 2] and was seen in both our patients. Peripheral neuropathy has been reported in 50% of patients. Mononeuropathy may also occur, but is seen more commonly in systemic PAN [1, 2].

The unusual feature in our patients was the severity of the arthritis, requiring aggressive treatment with immunosuppressants. Because of its potentially high toxicity, cyclophosphamide is reserved for severe forms of vasculitis unresponsive to other less toxic treatments. While arthralgias and non-destructive arthritis have been reported in CPAN, an association with severe arthritis has not been emphasized in the literature to date. In one review, arthralgias were reported in 12 of 23 cases, but radiological changes were minimal or absent in most [1]. Likewise, Kumar et al. [3] described arthralgias in 7 of 10 children with CPAN, but without radiological evidence of bony deformity. Neither arthralgias nor arthritis were seen in 14 patients with CPAN described by Borrie [4]. There is one report of a patient with CPAN and severe arthritis resulting in progressive joint destruction which required arthroplasty [5], and a patient with rapidly progressive non-destructive arthritis has also been described [6].

These two cases highlight the overlap between predominantly systemic and localized forms of PAN, and demonstrate that the course of CPAN may be a chronic, painful one requiring aggressive treatment, and is not as benign as previous reports have suggested [1]. Patients with CPAN may initially present with ankle pain and an awareness of the various cutaneous manifestations is important to enable early diagnosis of this condition.

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Accepted 14 May 1999

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