Painful muscular atrophy of the leg

Sir, We read with great interest the article by Garcia-Porrua et al. on panarteritis nodosa (PAN) limited to the calf and mimicking deep venous thrombosis. We want to report a similar case, but with a more protracted course.

A 37-yr-old woman was admitted with a 10-yr history of pain and swelling of her left calf followed by progressive muscular atrophy and shrinkage resulting in a talipes equinus that required surgical revision. Previous differential diagnoses had included Paget’s disease (because of increased technetium-99m uptake in a bone scan of the tibia, Fig. 1A) and myositis ossificans [based on radiographic findings of periostitis and bone apposition of the left tibia, (Fig. 1B), muscular oedema in magnetic resonance imaging (MRI), and a myositis pattern with axonal degeneration of the tibial and peroneal nerves on electromyography]. Past medical history revealed radiation therapy of a large haemangiomma of the left thigh in early childhood and several episodes of neuritis nervi optici. There was no history of trauma.

On examination, her left lower leg appeared markedly atrophic, the circumference being reduced by more than 60%. The gastrocnemius and peroneus muscles were fibrotic. Skin and subcutaneous tissue were sclerotic and tender, but no oedema or epidermal atrophy were present. On the forefoot prominent livedo racemosa discoloration of the skin was observed (Fig. 1D, E). Pulses of the a. poplitea and a. tibialis anterior were weakly palpable.

The leg was aching at rest and active and passive motion of the left ankle were restricted and painful, rendering walking difficult. Systemic signs were absent. Laboratory investigations revealed a markedly elevated erythrocyte sedimentation rate of 80 mm h and moderate leucocytosis (12 x 10^9 l); antinuclear, anti-DNA, antiphospholipid and antineutrophil cytoplasmic antibodies and hepatitis serology were all negative. Liver and renal function tests as well as muscle enzymes were within the normal range. Arteriography revealed an abrupt occlusion of the left popliteal artery with screwdriver-like collateral vessels. MRI investigations disclosed, as expected, muscular and adipose tissue fibrosis of the left calf (Fig. 1C). Furthermore, in the right leg, acute myositis of the peroneus muscle was detected which corresponded to a painful swelling of the anterior aspects of the right calf. Re-evaluation of a muscle biopsy performed 9 yr earlier revealed vasculitis of medium-sized arteries. A diagnosis of PAN limited to the calves was made. Systemic corticosteroid treatment (methylprednisolone at 1 mg/kg per day) resulted in decreased pain and swelling of the right lower leg. While tapering the dose, a crop of painful erythematous nodules arose on both calves, the left forefoot (Fig. 1D, E) and both forearms. Again biopsy revealed medium-sized vessel vasculitis with no detectable immunoglobulin deposits. Consequently, a combination regimen of methylprednisolone (1 mg/kg per day) and cyclophosphamide (2 mg/kg body weight per day orally) was instituted. Skin nodules, pain and swelling disappeared within a few weeks and the steroid dose was tapered. The patient remained symptom free on a maintenance
therapy of methylprednisolone (2 mg/day) and cyclophosphamide (2 mg/kg per day) for 2 years.

Limited forms of PAN, in particular of the skin, are known to occur [2–5] and are held to have a better prognosis. PAN localized to the calf is a clinically typical but excessively rare entity presenting as a painfully indurated calf with features of a pseudotumour of the muscle [6]. Differentiation from genuine neoplasms rests on the proof of oedema and myositis by MRI [6] and on the diagnostic vascular pathology on muscle biopsy and arteriography. Another hallmark of this disease is new bone formation similar to hypertrophic osteoarthropathy, as first described in 1956 by Saville [7]. With respect to the livedo reticularis discoloration of the skin, dermatological differential diagnoses include nodular vasculitis and livedoid vasculitis.

Our patient exhibited some unusual features of PAN of the calf: a particularly protracted course resulting in marked muscular atrophy, which had a severe impact on quality of life, and the presence of moderate systemic manifestations of PAN, such as lesions of the upper extremities and episodes of neuritis nervi optici. It is a matter of speculation whether radiation therapy of the large haemangioma on her left thigh in childhood had an influence on the pronounced asymmetry of the disease process in the lower legs.

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FIG. 1. (A) Technetium scan with increased uptake in the left leg, (B) radiograph showing periosteal bone apposition, (C) MRI with fibrosis of soft tissue, (D, E) erythematous nodules, livedo racemosa pattern and atrophy of left leg.