Letters to the Editor

Sarcoidosis with palpable nodular myositis, periostitis and large-vessel vasculitis simulating Takayasu’s arteritis

Sir, Both periostitis and palpable nodular myositis are rare forms of symptomatic involvement of bone and muscle in sarcoidosis [1, 2]. Vasculitis secondary to sarcoidosis has been reported infrequently in children and adults [3, 4]. We describe a young woman with periostitis, palpable nodular myositis and large-vessel vasculitis simulating Takayasu’s arteritis (TA) without pulmonary involvement.

In 1993, a 29-yr-old woman was admitted to our clinic with painful nodular swellings in her right thigh, left gluteal, dorsal and abdominal regions. She had pain in the left arm, and a 15 kg weight loss over 4 months. Physical examination of the patient revealed nodular swellings in the right quadriceps (10 × 12 cm), left gluteus maximus (6 × 5 cm) and her back. Several sites of erythema nodosum on the anterior surface of both legs, and a markedly diminished left radial pulse, were present. Laboratory results were as follows: haemoglobin 8.5 g/dl, white blood cell count (WBC) 12 200/mm³ with normal differential, platelets 641 000/mm³, erythrocyte sedimentation rate (ESR) 82 mm/h, calcium 11.4 mg/dl (normal: 9–11 mg/dl), serum IgG 5400 mg/dl (normal: 800–1700 mg/dl), IgA 645 mg/dl (normal: 85–490 mg/dl). Muscle enzymes, ANA, rheumatoid factor, VDRL, angiotensin converting enzyme, and complement components C3 and C4 were negative or within normal limits. A chest radiograph disclosed aortic calcifications. A radiographic survey of the skeleton defined periosteal reaction in the proximal right femur. A technetium (⁹⁹mTc MDP) scan showed avid uptake in the proximal right femur and the bottom of the right scapula. An abdominal CT scan revealed heterogeneous thickening in the erector spinae, quadratus lumbarum, psoas and abdominal muscles. Electroneuromyography, Doppler velocimetric study and a high-resolution CT of the lung were normal. Biopsy of tissue from the right quadriceps and its underlying periosteum revealed non-caseating granulomas and several giant cells, some of which were the ‘Langhans’ type (Fig. 1a). Kveim test limits. Aortography disclosed complete obstruction of the left subclavian, left vertebral and superior mesenteric arteries, and diffuse narrowing in the left common carotid artery (Fig. 2). A CT scan of the right femur revealed periosteal reaction in the proximal femur (Fig. 1b). Prednisolone was increased to 60 mg/day, and she was also started on azathioprine 150 mg/day. At the end of 9 months follow-up, her ESR decreased to 30 mm/h, and her left arm pain moderately improved. On the other hand, we did not observe any change in the periosteal reaction and peripheral pulses.

Sarcoidosis was diagnosed, and the patient began a regimen of prednisolone 40 mg/day. After 1 month of treatment, all complaints apart from the pain in the left arm had been resolved, and prednisolone was gradually tapered to 10 mg/day. In December 1997, the patient was admitted because of pain in the left arm associated with a lack of left brachial and radial pulses. Blood pressure could not be measured in the left arm. Her blood pressure in the right arm was 110/70 mmHg. A coarse murmur was heard over the left side of her neck. At that time, laboratory results were as follows: haemoglobin 11 g/dl, WBC 15 700/mm³, ESR 70 mm/h. A PTT, IgG and IgM anticardiolipin (aCL) antibodies, c-ANCA, p-ANCA, antithrombin III, protein C and S, and cholesterol levels were normal or within normal limits. Aortography disclosed complete obstruction of the left subclavian, left vertebral and superior mesenteric arteries, and diffuse narrowing in the left common carotid artery (Fig. 2). A CT scan of the right femur revealed periosteal reaction in the proximal femur (Fig. 1b). Prednisolone was increased to 60 mg/day, and she was also started on azathioprine 150 mg/day. At the end of 9 months follow-up, her ESR decreased to 30 mm/h, and her left arm pain moderately improved. On the other hand, we did not observe any change in the periosteal reaction and peripheral pulses.

The least common form of symptomatic sarcoid muscle disease is the palpable nodule, which may cause pain and stiffness with cramps [1]. Periosteal reaction is also rare and thought to preclude the diagnosis of osseous sarcoidosis [2, 5]. To our knowledge, only one case with periostitis has been reported previously [5]. In our case, although the left radial pulse was lacking...
at the onset of the disease, aortic vasculopathy was diagnosed 4 yr later. The differential diagnosis of a patient with large-vessel involvement includes arteriosclerosis, embolus and rheumatic disorders. These disorders are unlikely in view of the patient’s clinical and laboratory findings. Aortitis may also occur in chronic inflammatory conditions such as syphilis, fungal infections and tuberculosis, but these were ruled out by special histochemical stains of the granulomatous lesions and by the serological studies. Giant cell arteritis and TA are two specific conditions that may cause aortic wall inflammation; the former is usually seen in older patients and is often associated with polymyalgia rheumatica. Our patient was young, and the temporal artery was clinically intact. Aortic arch involvement indistinguishable from TA in sarcoidosis has been reported previously in an adolescent and an adult [6, 7]. Gedalia et al. [8] also described a patient with an abdominal aortic aneurysm associated with childhood sarcoidosis.

We conclude that periostitis may develop in patients with sarcoidosis, particularly those with nodular myositis. We also suggest that all patients with sarcoidosis be followed carefully for the development of occlusive arterial involvement.

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