Classical polyarteritis nodosa (PAN) is characterized by multisystemic vasculitis in the small and medium-sized arteries. Cutaneous PAN is the most common limited form of PAN and has a benign prognosis. Another limited form of PAN that is confined to calf muscles has been documented; 28 cases have been reported (1–6). In addition, 6 cases with coexistence of calf muscle PAN and skin lesions have been reported (2–4). These cases suggest that there are similarities between calf muscle-affecting PAN and cutaneous PAN. However, it is not known how lesions can coexist in different organs without systemic manifestation. We report here a patient with PAN with lesions limited to the calf muscles and the skin. Ectopic adipose tissue accommodating markedly damaged arteries was found in the calf soleus muscle.

CASE REPORT

A previously healthy 78-year-old Japanese woman had a 2-month history of a papulonodular eruption on the legs with pain in the calves. The eruption gradually extended over her legs with heavy pain, and subsequently a low-grade fever was noted. Her condition was tentatively diagnosed as cellulitis, but it did not respond to antibiotics. The patient was referred to us for further evaluation of her leg lesions. On examination, the patient had red papules and purpura/petechiae on the bilateral lower extremities (Fig. 1a). A skin biopsy specimen of an indurated papular lesion showed vasculitis of a medium-sized artery with fibrinoid deposition in the subcutaneous tissue (Fig. 2a). Laboratory examination revealed a high serum C-reactive protein level (8.7 mg/dl; normal < 0.3 mg/dl) and normal creatinine and creatine phosphokinase levels. Antinuclear antibody was positive at 1:320 (homogenous pattern), but antibodies to ss- and ds-DNA, anti-neutrophil cytoplasmic antibody and antiphospholipid antibody were negative.

Magnetic resonance imaging (MRI) of the legs revealed increased signal intensity in the calf muscles and the subcutaneous tissue in T2-weighted images, indicating inflammatory changes in these areas. T1-weighted images exhibited a high signal intensity area in the left soleus muscle (Fig. 1b), suggesting the presence of ectopic adipose tissue within the muscle. A biopsy specimen from the abnormal area of the soleus muscle confirmed the presence of ectopic adipose tissue; there were vasculitic changes in medium-sized vessels located in the ectopic fat tissue with a severe perivascular inflammatory infiltrate (Fig. 2b). Higher magnification disclosed a marked infiltrate of neu-
trophils, eosinophils and histiocytes in and around the arterial walls. Remarkable disruption of the internal elastic lamina of the arteries was revealed by elastica van Gieson staining (Fig. 2c). There was no vasculitis in the surrounding muscle tissue.

The condition was diagnosed as PAN limited to the calf muscles and the skin, because of the absence of clinical or laboratory signs of systemic involvement. The patient was treated with oral prednisolone 20 mg daily, and the eruption and pain improved immediately. The corticosteroid was tapered over a few weeks. Currently, 8 months later, the patient has had no recurrence of the symptoms or CRP elevation at a dose of prednisolone 5 mg daily.

DISCUSSION

To our knowledge, this is the first case demonstrating the occurrence of PAN in the ectopic adipose tissue of skeletal muscles. The patient’s calf pain was attributable to the vascular changes in the ectopic fat tissue, but not to primary myositis. Since the patient had no severe systemic symptoms, the vasculitis seems to be confined to the skin and the calf muscle fat tissue. In this context, the vessels located in the adipose tissues of the involved skin and those in the ectopic fat may be identically damaged in our patient. Calf muscles contain intermuscular adipose tissue at higher levels than do other muscles of the lower legs (7), suggesting that PAN is prone to occur in calf muscles. The case reported here raises the possibility that arteries located in the ectopic adipose tissues might be affected in reported cases of PAN limited to calf muscles or to calf muscles and skin.

REFERENCES