Case report

Giant cell arteritis of the skin simulating erythema nodosum

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SUMMARY Cutaneous involvement in giant cell arteritis is quite uncommon. A patient is described who presented with pretibial skin lesions clinically indistinguishable from erythema nodosum which, on biopsy, showed subcutaneous panniculart giant cell vasculitis. Cutaneous manifestations of giant cell arteritis are subsequently reviewed.

Key words: temporal arteritis, tender nodules.

Cutaneous involvement in giant cell arteritis is uncommon.1, 2 Reported dermatological abnormalities are limited to either tendon nodules overlying inflamed superficial arteries or to presumed ischaemic sequelae such as purpura, necrotic ulcers, or gangrene.3-10 We describe a patient with giant cell arteritis who presented with lower extremity claudication and pretibial skin lesions clinically indistinguishable from erythema nodosum. Biopsy of one such lesion showed a subcutaneous panniculart giant cell vasculitis.

Case report

A 61 year old Caucasian housewife presented with a four month history of aching in her feet. One month before admission she had noted heaviness in her calves, which limited her walking to half a block. Initially the heaviness was relieved temporarily by rest, but it rapidly progressed to include nocturnal calf and foot pain. The patient was a non-smoker and denied fatigue, myalgias, fever, weight loss, headache, jaw or upper extremity claudication, or visual symptoms.

Two weeks before admission a physical examination was remarkable for bilateral femoral bruits and the absence of pulses in the left popliteal and both posterior tibial and dorsalis pedis vessels. By the time of admission the patient had developed six 1.5-2.0 cm, round, erythematous, tender nodules in the left pretibial region. Temporal arteries were normal to palpation. There had been no intervening illness or drug exposure.

Results of laboratory studies obtained at the time of admission included haemoglobin 133 g/l, white blood cell count 6×10⁹/l, platelet count 371×10⁹/l, erythrocyte sedimentation rate 47 mm/h (Westergren), urine analysis normal, and serum rheumatoid factor negative. The chest radiograph and the electrocardiograph were normal. Serum protein electrophoresis showed increased alpha-1, alpha-2, and beta globulins and a normal gamma-globulin band, consistent with an acute phase protein response. The lipoprotein panel was interpreted as Fredrickson’s type IIA hyperlipoproteinemia. Results of electromyography and nerve conduction studies of the legs were normal. An aortofemoral arteriogram showed extensive tapering and stenosis of the profunda femoral bilaterally, moderate stenosis at multiple levels in the superficial femoral arteries, and marked stenosis of the left popliteal artery. Below the level of the mid-calf no perfusion was demonstrated. Tapering of the vessels on the arteriogram suggested arteritis. Excisional biopsies of a pretibial lesion and of the left temporal artery were performed.

Histological examination of the excised portion of the left temporal artery showed an inflammatory
vasculopathy compatible with giant cell arteritis superimposed upon atherosclerotic vessels. Glucocorticoid therapy was initiated in the form of prednisone orally at 60 mg/day, tapered to 10 mg orally per day over the 18 months of follow up. The pretibial lesions receded between the fifth and eighth days of glucocorticoid therapy and have not recurred. Symptoms of claudication gradually diminished and after six months of therapy the patient could walk four to six blocks without pain. Segmental Doppler flow and measured studies comparing arm with ankle pressures improved from an index of 0.4 at time of diagnosis to 0.7 at six months (an index of 1.0 or greater is considered normal).

**PATHOLOGY**

Sections of the left temporal artery showed variable intimal thickening with narrowing of the luminal size and altered elastic tissue with aggregates of multinucleated giant cells. The musculature and adventitia of the vessel wall were segmentally involved, with a cellular infiltrate of lymphocytes, neutrophils, histiocytic cells, and giant cells, consistent with giant cell arteritis (Fig. 1).

The histological examination of the skin specimen from the leg showed foci of large sized blood vessels with obliterated lumina and thickened walls within the subcutaneous fat. These vessel walls were infiltrated with lymphocytes, histiocytes, polymorphonuclear leucocytes, and a few eosinophilic leucocytes, as well as multinucleated giant cells (Fig. 2B). In addition to arteritis and disruption of the elastic tissues, areas of calcification were noted within the vessel walls (Fig. 2A). The involvement was limited to vessels in the subcutaneous fat and septae, the overlying epidermis and dermis appearing histologically normal.

**Discussion**

Giant cell arteritis is a relatively common disease, particularly in the United States and Northern Europe. In a study of the population of Olmsted County, Minnesota, the annual incidence was 17.4 per 100 000 persons aged 50 years or older. In a large routine necropsy series from Sweden, in which sections of the aorta and temporal arteries were examined, the prevalence of giant cell arteritis was found to be 1.7%. Nevertheless, although the clinical spectrum of giant cell arteritis has unfolded to include renal, hepatic, synovial, neurological and constitutional features, cutaneous involvement is uncommonly reported. When skin changes do occur, they most frequently involve the scalp. In his review of dermatological manifestations of giant cell arteritis Hitch describes tenderness, erythema, oedema, bullae, ulceration, and necrosis in the distribution of the superficial temporal arteries. In the lower extremities, reported skin changes include purpura, ecchymoses, ulceration, and gangrene. Urticaria, lividity, and hyperpigmentation of the skin have also been described.

Our patient presented with claudication of the lower extremities and typical erythema nodosum-like lesions—round, indurated, tender, erythematous nodules 1.5–2.0 cm in diameter—on the pre-
tibial areas of her legs (consistent with the original description of erythema nodosum by Willan in 1808). In our patient, however, these lesions histologically showed a giant cell vasculitis of the subcutaneous and septal vessels not described in erythema nodosum. Lower extremity angiogram suggested large vessel arteritis, and a temporal artery biopsy showed a multinucleated giant cell vasculitis consistent with temporal arteritis.

Erythema nodosum is a disease of subcutaneous interlobular septae, with septal venulitis, hyalinisation of connective tissue septae, occasional radially arranged nodular granulomata, and a bland proliferative vasculopathy. Multinucleated giant cells may be seen in erythema nodosum, but they are primarily restricted to the septae of the subcutaneous fat lobules and are not seen infiltrating the vessels as in our patient. No granulomata were seen in our patient’s skin specimen, further distinguishing her lesions from classic erythema nodosum. Calcification in the area of the temporal artery has previously been described in giant cell arteritis.

Erythema nodosum has been reported to occur in association with Takayasu’s arteritis, an idiopathic inflammatory process with a predilection for the aortic arch and its branches, occurring primarily in young women of oriental extraction. In one patient with Takayasu’s arteritis and skin lesions suggestive of erythema nodosum a biopsy of a nodule showed ‘lesions of the small vessels of the subcutis which were reminiscent of polyarteritis nodosa’. Clinically our patient did not have Takayasu’s arteritis, as she was over 60 and had no involvement of the eyes, heart, or arms.

In summary, we present a patient with giant cell arteritis who developed skin lesions clinically identical to erythema nodosum which, on biopsy, were found to be subcutaneous multinucleated giant cell vasculitis. Administration of oral glucocorticoids yielded prompt resolution of the cutaneous lesions and a gradual but complete reduction of the lower extremity claudication symptoms.

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