CASE REPORT

Ischaemia due to a vascular malformation causing focal myositis

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SUMMARY

Summary Focal myositis secondary to an intramuscular vascular malformation has rarely been reported in the literature. We describe a 21-year-old woman presenting with left thigh pain. Imaging of the thigh muscles showed a vascular malformation and muscle biopsy demonstrated focal changes diagnosed initially as myositis. Ischaemia is thought to be the responsible mechanism.

BACKGROUND

The two broad categories of vascular anomalies are vascular neoplasms and vascular malformations (VMs), differentiated by the absence of endothelial proliferative activity in the latter.[1] VMs are further subcategorised into high flow (arteriovenous malformations and arteriovenous fistulas) and low flow (venous, lymphatic, capillary or a combination thereof).[2] VMs can occur in the brain, liver, lungs, uterus and the limbs including the subcutaneous tissues and muscles.[3 4] There are several case reports of VMs of the muscles of the limbs and the head and neck region,[5–9] but to our knowledge none with a biopsy proven myositis. Intramuscular VMs tend to occur in younger age groups, more commonly affecting the lower extremities.[10–12]

We describe a young woman presenting with focal myositis in association with an intramuscular VM with an emphasis on the muscle biopsy features.

CASE PRESENTATION

A 21-year-old woman presented to our clinic with a history of pain in the inner left thigh for 2 years. The pain was continuous, graded at 4/10 on the visual analogue pain scale, and was exacerbated by exercise and by prolonged sitting. She had limited mobility due to the pain. There was no history of visual disturbances, dysphagia, dysarthria, limbs weakness, backache, sphincteric dysfunction, trauma to the lower limb, fever, weight loss, skin rash, joint pain or change in urine colour. The patient has type 1 diabetes mellitus, well controlled on insulin and the last HbA1C was 7%. Her family history was unremarkable. The neurological examination was normal.

INVESTIGATIONS

The CK level was 45 IU/L (normal range: 29–165 IU/L). Rheumatological work-up was normal without evidence of vasculitis. Ultrasound (US) of the lower limb muscles showed a 4×1.3×1.5 cm, well defined ovoid hypoechoic lesion within the left adductor muscles with no internal vascularity. MRI of the left thigh showed a focal lesion, isointense on T1 and hyperintense on T2, within the adductor magnus muscle with surrounding oedema interpreted as myositis ossificans. Nerve conduction studies and electromyographic examination were normal. Biopsy of the left adductor muscle showed fasciitis, frequent degenerating, regenerating and necrotic fibres (perifascicular in some regions), chronic inflammatory infiltrates with a moderate number of T lymphocytes, often in the vicinity of blood vessels (figure 1). There were no ragged red fibres or inclusion bodies. Larger numbers of lymphocytes were present in the adjacent fibroconnective tissue. Electron microscopy showed no evidence of tubuloreticular inclusions in endothelial cells. A diagnosis of focal inflammatory myopathy was made and the patient was treated with methotrexate but did not improve after 4 months of therapy. A repeat MRI showed an isointense T1; hyperintense T2 lesion in the left adductor magnus muscle measuring 3.5×1.5×1.5 cm, with no muscle atrophy or fatty infiltration, suggestive of haemangioma (figure 2). Repeated US of the lower limb muscles showed an irregular solid hypoechoic mass in the left adductor magnus muscle measuring 3.3×1.7×1.2 cm. An intramuscular low-flow venous vascular malformation (VVM) was diagnosed and angiography was not deemed necessary in this patient.

DIFFERENTIAL DIAGNOSIS

- Polymyositis.
- Myositis ossificans.
- Infectious myositis.
- Primary or secondary muscle tumours.
- Idiopathic focal myositis.

TREATMENT

The patient was treated with sclerotherapy (figure 3) and the symptoms resolved.

OUTCOME AND FOLLOW-UP

The patient's symptoms resolved giving support to the VVM as the cause of the underlying symptoms.

DISCUSSION

The large and heterogeneous group of inflammatory myopathies includes the forms known as polymyositis, dermatomyositis and inclusion body myositis. Idiopathic focal myositis was first described in 1977 by Heffner *et al* who published 16 cases presenting with a rapidly enlarging focal muscle mass with inflammatory myopathic features

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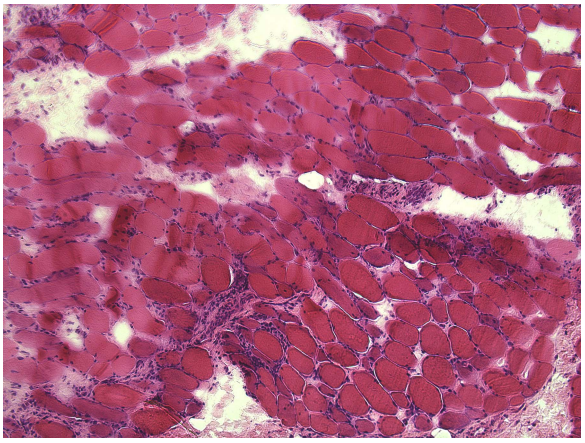

Figure 1 H&E-stained section (×10 magnification) reveals moderate infiltration by lymphocytes (confirmed by immunohistochemistry for CD45 and CD3 (not shown). Frequent severely atrophic fibres are present, often showing signs of regeneration.

on biopsy.[13] Generalisation of focal myositis is variable.[9 14] Ischaemia has long been implicated in the pathogenesis of muscle inflammation.[15 16] Most focal myositis cases reported in the literature are idiopathic, however, atheromatous emboli and diabetic angiopathy have been reported in association with acute focal myositis secondary to ischaemia.[16–18] Less than 1% of VMs occur in skeletal muscle.[19] In general, 90% of VMs—outside the central nervous system—are low flow (capillary, venous or lymphatic); the commonest among them are the VVMs.[20] VVMs are usually present at birth and females are more commonly affected.[10 21] Patients usually present with pain exacerbated by menses or pregnancy, swelling, bluish discolouration of the skin, bony deformities, fractures and intralesional bleeding, but some are asymptomatic.[9 10] Our patient presented only with pain. Perhaps a clue to the underlying aetiology was the worsening of pain with exercise and with sitting upright, perhaps due to mechanical compression of the malformation with obstruction of venous outflow, or due to higher blood flow

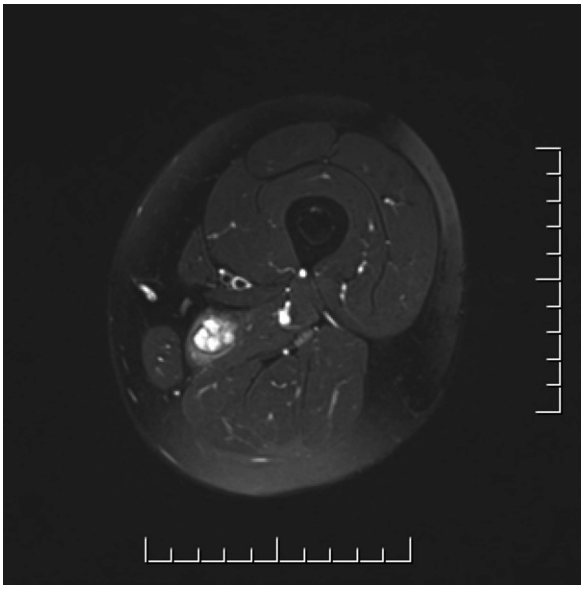

Figure 2 MRI of the left thigh showing T2 high-signal intensity lesion in the adductor magnus muscle.

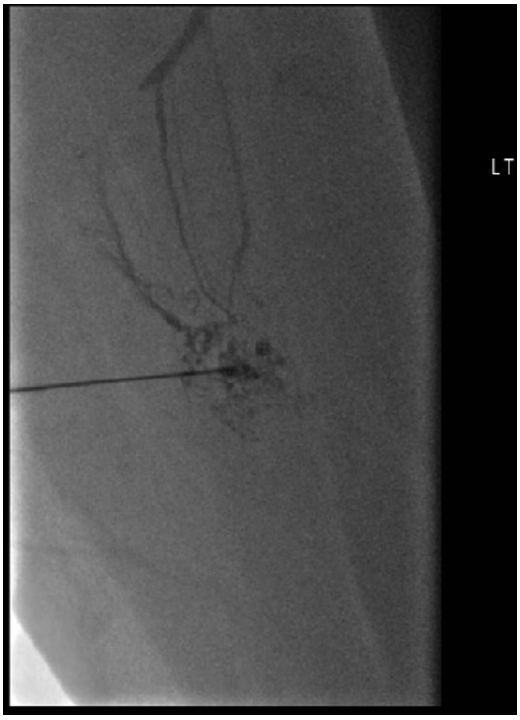

Figure 3 Ultrasound-guided sclerotherapy of the left thigh venous malformation.

rates with exercise. MRI is the imaging modality of choice for primary classification and localisation, in addition to post treatment follow-up; however, some authorities suggest that rigorous assessment requires an angiogram, although that did not appear to be necessary in our case.[22] US imaging has multiple disadvantages, being operator dependent with a limited field of view and restricted penetration.[10 23] Treatment is tailored to the specific patient. It includes: aspirin and compressive garments, sclerotherapy and surgical resection.[10] VMs are an important differential diagnostic consideration when assessing patients presenting with focal myositis in order to avoid unnecessary and potentially harmful treatments.

Learning points

- Consider vascular malformations (VMs) in the differential diagnosis of focal myositis.
- The most common VMs outside the central nervous system are VVMs.
- Exacerbation of pain by positions/activities that increase blood flow to the involved limb is a clue to the diagnosis.

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Competing interests None.

Patient consent Obtained.

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