A self limiting tumour

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Case history

A 20 year old professional footballer presented with pain and swelling of the left thigh after a kick three weeks previously. He had sustained a number of injuries in the past including a previous calf haematoma because of the nature of his profession but none had been as severe.

Clinical examination revealed a normal gait. There was definite but diffuse swelling of the extensor compartment of the left thigh that measured 1 cm more in circumference compared with the normal right side at the level of the mid-thigh. It was tender but no discrete mass could be felt. There was also slight limitation of knee flexion but extension was full and no hip abnormality was detected. The patient was otherwise well with no constitutional symptoms. The rest of the physical examination was normal.

Plain radiographs of the left thigh were obtained. What abnormality does figure 1 show? What is the differential diagnosis?

Figure 1 Plain radiograph of the left thigh.

Figure 2 99m Technetium MDP scintigram of the thighs showing diffusion (A) and bone (B) phases. Increased uptake is seen in the juxtacortical lesion related to the left femur.

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Figure 3 Longitudinal (A) and axial (B) ultrasound images of the quadriceps muscles showing calcification in the periphery of the soft tissue lesion characteristic of the more mature outer zone (arrows) of early myositis ossificans.

Differential diagnosis

The clinical history points strongly to a post-traumatic sequel. This may have involved either the bone or soft tissues. Bony involvement may be attributable to a fracture or stress fracture. The first develops acutely after the injury whereas the second has a more insidious course and often develops at a later stage because of repeated weight bearing, for example, if the patient returns to training after the initial insult. A detailed history as to the time course of the symptoms in relation to the injury is particularly useful in this regard. In this patient, the normal gait suggests that a fracture or stress fracture is unlikely.

Post-traumatic soft tissue lesions include haematoma, myositis ossificans, or a muscle tear with or without associated muscle hernia-

tion. The diagnosis of muscle herniation can usually be confirmed clinically by demonstrating the presence of a soft tissue lump/mass when the patient contracts the affected muscle and its disappearance when the muscle is relaxed. In the absence of this, it is often difficult to distinguish a haematoma from early myositis ossificans or a muscle tear clinically.

Besides trauma, tumour is another consideration albeit a much less likely one. Not infrequently, patients with a tumour may not notice the lesion until trauma to the area directs their attention to it. This is especially so if the trauma results in haemorrhage within the tumour, which then gives rise to pain. In a patient of this age group, the tumours may include a variety of benign or malignant soft tissue tumours or even bone tumours such as osteosarcoma. An abscess or cellulitis is unlikely in this patient because of the lack of constitutional symptoms.

Plain radiographs are an excellent means for initial radiological evaluation. They provide good bony details and besides demonstrating obvious bone abnormalities, for example, fractures or bone tumours, they are also useful in detecting more subtle abnormalities such as early periosteal reaction that may be seen with stress fractures or some other pathologies. Soft tissue abnormalities on the other hand are less well demonstrated with plain radiographs and are often visualised as non-specific soft tissue swelling.

In this patient, the lateral radiograph of the left thigh shows soft tissue swelling adjacent to the femur with some calcification seen mainly at the periphery of the mass. No associated bony abnormality is detected. This appearance is highly suggestive of myositis ossificans. Occasionally, tumours, for example, parosteal osteosarcoma or synovial sarcoma may give rise to a calcified mass adjacent to bone. Generally the calcification tends to be more dense in the first and less well formed in the second compared with the calcification seen in this case.

Further investigations

The patient went on to have a 99m-technetium radionuclide scintigram as well as ultrasound and magnetic resonance imaging (MRI). The three phase scintigram (fig 2) shows increased activity corresponding to the lesion noted on the plain radiographs in the flow, blood-pool and delayed images. Ultrasonography confirmed the presence of a soft tissue mass with calcification seen mainly at its periphery (fig 3). A Doppler study performed at the same sitting showed the mass to be avascular with no Doppler signal detected within it. MRI (fig 4) revealed a mass of mixed signal with some subacute blood within it and some adjacent soft tissue oedema. No underlying bone abnormality was noted.

Diagnosis

Myositis ossificans (heterotopic bone formation).

Figure 4 Sagittal T1-spin echo before (A) and after (B) Gd-DTPA images showing a small area of haemorrhage in (A) with a much larger abnormality demonstrated after contrast administration. The before and after contrast axial images are depicted in (C) and (D) respectively.

Discussion

Myositis ossificans is a benign self limiting condition in which a mass of heterotopic bone forms within the soft tissues. Although commonly used, the designation myositis ossificans is misleading as the lesion is not inflammatory

in nature and muscle is not always involved. A number of other names have been proposed to describe this condition (for example, heterotopic bone formation, pseudomalignant osseous tumour of the soft tissue, extraosseous localised non-neoplastic bone and cartilage formation, myositis ossificans circumscripta and pseudomalignant myositis ossificans) but the term myositis ossificans has been retained in the WHO disease classification as it is widely used by clinicians and pathologists.

The condition often develops within one to two weeks after direct trauma to the area or unusual muscular exertion. In about half the cases however, no apparent history of antecedent trauma¹ can be elicited. It is more common in men and usually affects adolescents and young adults.² Although it may occur at virtually any site, the usual areas involved are those most susceptible to injury, typically the thigh (quadriceps femoris and adductor muscles), elbow (flexor muscles) and buttocks (gluteal muscles) and less often the shoulder and calf.^{3,4} In general, the proximal portion of the extremity is more frequently affected than the distal part.⁵

The initial pathological process comprises of muscle necrosis and haemorrhage after the trauma. This gives rise to a soft tissue mass that is often accompanied by pain, warmth and surrounding oedema. Occasionally, there may also be fever and an increased erythrocyte sedimentation rate causing clinical confusion with an abscess. The presence and activity of myositis ossificans can be detected and monitored by estimation of the alkaline phosphatase levels. However, this is non-specific.⁶ Histologically, marked proliferation of spindle cells with a well recognised zoning phenomenon is observed.⁷ The least differentiated tissue lies in the central zone where rapidly proliferating spindle cells of various shapes and sizes and even atypical mitoses are seen. Because of the marked cellular proliferation, biopsy specimens obtained from this zone may be easily confused with a sarcoma. Adjacent to this is a middle or intermediate zone where the osteoid is more organised in appearance and separated by a loose cellular stroma. The outer zone is the most mature and consists of well formed bone which may form a shell around the entire lesion.² Cartilage formation may also be present. These three zones need not necessarily be distinct nor do they always have a concentric distribution but their identification is important as it permits definite distinction from soft tissue or bone sarcomas that do not exhibit a similar zonal phenomenon.⁸

No calcification is present in the early phase but with time, by the third week, calcification appears first in the periphery followed by mature bone throughout the entire lesion.

The radiographic appearances reflect the underlying histological pattern of maturation. In the early phase, plain radiographs may be unremarkable or show non-specific soft tissue swelling. Occasionally periosteal reaction may be observed if the lesion is juxtacortical in location. Faint calcification is often visible within two to six weeks^{2,7} and at about six to

Figure 5 (A) Lateral radiograph of the left thigh at six months showing maturation of the myositis ossificans that has reduced significantly in size compared with figure 1. (B) The mature lesion is also seen on the T1 axial MR image showing the significant reduction in size.

eight weeks, a lacy pattern of new bone may form around the periphery of the mass.⁹ As the lesion matures, there is increasing calcification and ossification of the lesion. A radiolucent band is usually but not always present between the mass and the adjacent bone cortex. Complete maturation is usually reached in five to six months by which time the size of the mass would have been constant and may well start to reduce.

The zonal phenomenon is better appreciated on computed tomography¹⁰ where a rim of mineralisation is usually seen after four to six weeks with a centre of decreased attenuation noted. This classic computed tomographic appearance is not observed in early disease. Even when calcification is present, the diagnosis may not always be clear cut. The calcified rim appears to be remodelled in the axis of the forces of traction.¹¹ It may thus be incompletely formed or irregular and be confused with other

soft tissue tumours that calcify such as soft tissue sarcomas or haemangioma.

Ultrasonography may also demonstrate a zonal appearance¹² and has been shown to be useful in the early detection of myositis ossificans at a stage when there is little or no calcification seen on plain radiographs.¹³

Scintigraphy is highly sensitive in detecting myositis ossificans. There is intense uptake of the technetium isotope because of the profuse osteoblastic activity and bone formation. Unfortunately this is non-specific as soft tissue and bone tumours also show increased activity as do a number of other conditions.¹⁴

The MRI findings in myositis ossificans vary according to the stage of the disease. A spectrum of MRI appearances may be seen and in some cases may simulate a tumour.¹⁵ Early lesions tend to be inhomogenous and show increased signal intensity centrally on T2 weighted images probably related to the increased cellularity here. Fluid-fluid levels resulting from haemorrhage and surrounding soft tissue and even bone marrow oedema¹⁶ may also be observed. Curvilinear and irregular areas of decreased signal corresponding to calcification are noted later on although these are better visualised on computed tomography. Chronic lesions tend to be well defined, possess a border of low signal and contain fatty marrow.

The significance of myositis ossificans lies in the fact that it may be confused with a malignant tumour, usually a soft tissue sarcoma. This problem is especially acute in the early stages of the disease when the radiological findings may not be so clear cut and a strong index of suspicion is required. Accurate diagnosis is important as the management is completely different in the two conditions. In myositis ossificans, treatment is usually not required although oral bisphosphonates, which are potent inhibitors of calcification,¹⁷ have been shown to be effective in modifying the process of heterotopic ossification. Occasionally, surgical resection may be necessary if the mass causes functional impairment. In these cases, resection is best performed when the lesion has matured as rapid recurrence may occur after resection of an immature lesion.¹⁸ If left alone, the lesion may reduce in size with time and in some instances disappear.

Follow up

The patient was given Didronel (etidronate sodium 400 mg thrice daily) in the hope of preventing further calcification/ossification. Clinically, he showed symptomatic improvement and was able to return to full training and resume his competitive sports activities. Within six months, follow up plain radiographs (fig 5A), ultrasound and MRI (fig 5B) show a decrease in the size of the lesion with maturation.

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