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?
Diagnosis
Myositis ossificans (heterotopic bone formation).
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, extrasosseous 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 trauma can be elicited. It is more common in men and usually affects adolescents and young adults. 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. In general, the proximal portion of the extremity is more frequently affected than the distal part.

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 phosphotase levels. However, this is non-specific. Histologically, marked proliferation of spindle cells with a well recognised zoning phenomenon is observed. 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. 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.

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 and at about six to
eight weeks, a lacy pattern of new bone may form around the periphery of the mass. 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 tomography, 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. It may thus be incompletely formed or irregular and be confused with other soft tissue tumours that calcify such as soft tissue sarcomas or haemangiomata.

Ultrasoundography may also demonstrate a zonal appearance 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.

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.

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. 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 oedema 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 biphosphonates, which are potent inhibitors of calcification, 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. 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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