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CASE REPORT

Myositis ossificans mimicking parosteal osteosarcoma : A case report and literature review

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Myossitis ossificans (MO) is an aberrant reparative process that causes benign heterotopic ossification in soft tissue. We report a case of MO presenting as a large mass located at the dorsal aspect of the distal thigh, with no history of trauma, with radiological and clinical features mimicking parosteal sarcoma. An incisional biopsy was performed and the mass was excised. The histological features identified the lesion as MO. In half of the cases, these ossifications may adhere to the periosteum. In these cases, the lesion is known as parosteal MO, which may be confused with a parosteal osteosarcoma. This parosteal MO seldom becomes malignant. We emphasize the importance of a differential diagnosis of MO, since these lesions may simulate tumours and lead to misdiagnosis.

Keywords : myositis ossificans ; parosteal osteosarcoma ; heterotopic ossification ; bone tumours.

INTRODUCTION

Myositis ossificans (MO), also known as heterotopic bone formation or heterotopic ossification, is characterized by the presence of bone in soft tissue where bone normally does not exist. The concept of MO was initially described by Von Dusch in 1868. The term, however, is a misnomer because the condition involves no muscle inflammation and the process is not limited to muscle. Although it primarily occurs in muscles, it may also form around ligaments, tendons, fasciae, aponeuroses and joint capsules.

MO is mostly precipitated by trauma such as fracture, total hip arthroplasty (THA), or direct muscular trauma, but it may also have a neurogenic cause, such as spinal cord or central nervous system injury. In addition, there is a rare hereditary form known as myositis ossificans progressiva. MO can also occur with no previous injury.

The clinical evolution, location, and radiological features are usually sufficient to arrive to the diagnosis of MO. However, in half of the cases, these ossifications may be deeper in the thigh and adhere to the periosteum (*11*). In these cases, the lesion is known as parosteal MO, which may be confused with parosteal osteosarcoma (POS). Parosteal MO seldom becomes malignant (*9*).

We report the case of a MO presenting as a large mass located deep at the dorsal aspect of the thigh,

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with no history of trauma, with radiological and clinical features mimicking parosteal osteosarcoma.

CASE REPORT

A 22-year-old Caucasian male, a motorbicycle mechanic, was admitted in our musculoskeletal oncology unit on December 2005 due to the presence of a mass located posteriorly at the distal part of his right thigh. Prior to admission, he had been admitted in another institution, where a marginal resection of the tumour had been performed. Two months after the surgery, the patient was referred to our institution owing to local recurrence. He denied a history of trauma or previous fracture to the extremity. His recent medical history was unremarkable and he denied any constitutional symptoms such as weight loss, fever or malaise. He referred that he had first become aware of the pain one month earlier and he had noticed a mass in his right thigh, which had progressively increased in size. Physical examination disclosed no evidence of either a palpable soft-tissue mass or adenopathies, but a scar from the previous surgery and a firm mass at the posterior aspect of the thigh. The mass seemed to be deeply seated in the muscle. There was increased warmth and a slight erythema over the area. Motor strength, distal pulses, and sensitive tests were normal. He ambulated with a normal gait but knee flexion was limited to 95°.

The x-ray imaging brought from the other institution revealed calcification within the thigh mass, which was denser peripherally than centrally. The image showed a thin area of decreased opacity separating the mass from the femoral shaft (Fig. 1A & 1B). Magnetic resonance imaging disclosed a mass which was well delineated by a hypo intense zone representing the mineralized component. The signal intensity from the mass core was heterogeneous (Fig. 1C). It was not possible to obtain the histological results from the resection specimen.

Plain radiographs taken upon admission in our unit revealed a 9 × 6 cm well-delimited completely ossified mass located at the posterior aspect of the distal femur, which seemed to be in close contact with the bone (Fig. 2A & 2B). Bone scintigraphy showed increased radioisotope uptake over the lesion (Fig. 3). Computed tomography scanning demonstrated the bone pattern of mineralization within the mass and revealed a focal, sessile attachment of the mass to the adjacent femoral cortex (Fig. 4). MRI revealed a 9 × 6 × 9 cm mass on the posterior cortex of the distal femur. The periphery of the lesion showed a hardly defined edge and a

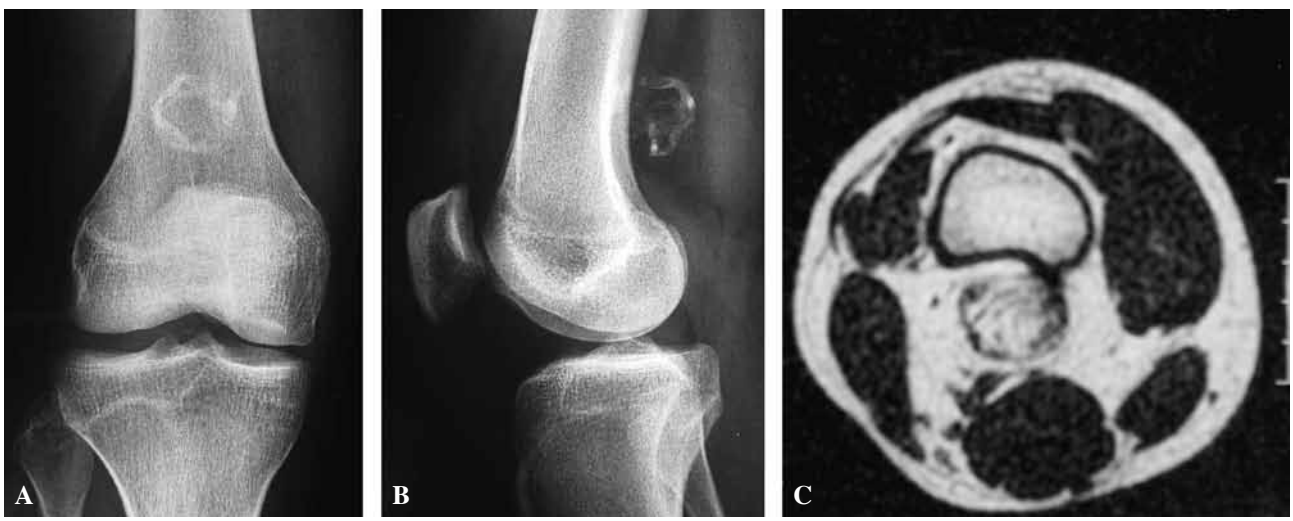

Fig. 1. — A & B : X-ray imaging revealed a calcification within the tigh mass which was denser peripherally than centrally. The image displayed a thin area of decreased opacity separating the mass from the femoral shaft. C : MRI disclosed a mass which was well delineated by a hypo intense zone representing the mineralized component. The signal intensity from the mass core was heterogeneous.

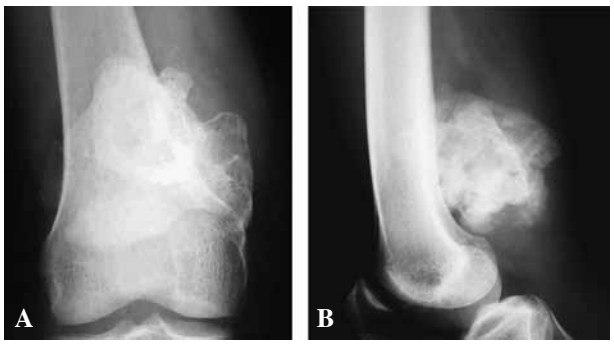

Fig. 2A & B. — Radiographs taken after the tumour recurred following the initial resection, showing a well-delineated completely ossified mass located at the posterior aspect of the distal femur, which appeared to be in close contact with the bone.

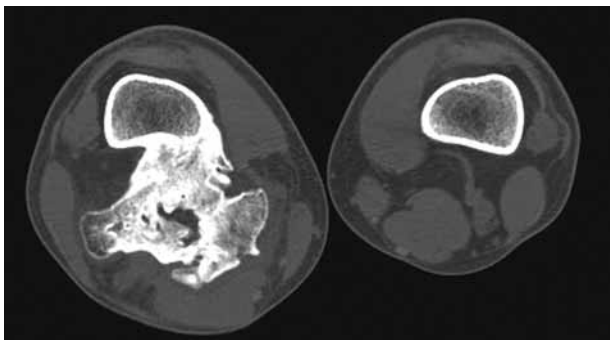

Fig. 4. — Computed tomography scanning demonstrated the bone pattern of mineralization within the mass and revealed a focal, sessile attachment of the mass to the adjacent femoral cortex.

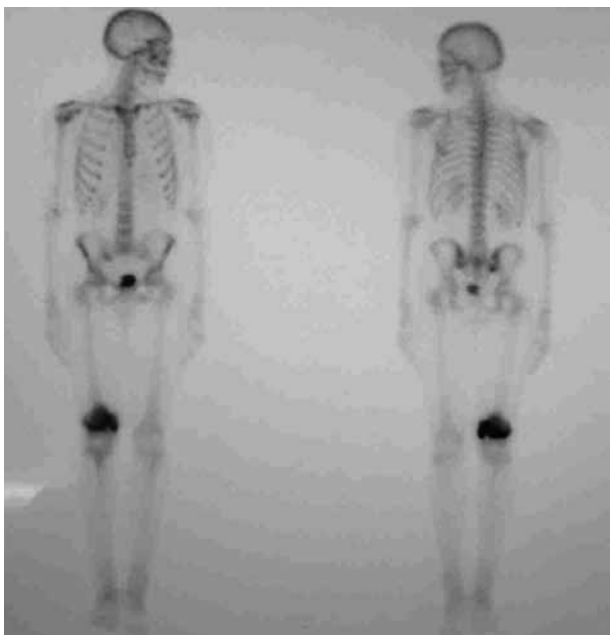

Fig. 3. — Bone scintigraphy identified the distal femur as an isolated region of increased radioisotope uptake.

heterogeneous enhancement of the mass periphery on the T1-weighted images after contrast administration (Fig. 5A). On T2-weighted images the mass appeared heterogeneous and had a markedly increased signal (Fig. 5B). The mass displaced the popliteal vessels posteriorly. There was a discernible clear fatty plane between the mass and the popliteal vessels and the tibial nerve on at least two axial sections. An intravenous digital subtraction angiogram was performed, showing a hypervascularized mass.

An incisional biopsy performed using partially the previous scar recovered a 4×0.7 cm cylinder of bone for histological analysis (Fig. 6) ; the latter showed no signs suggesting a malignant lesion. Three weeks later, the patient underwent resection of the mass. The resection specimen consisted of a $9 \times 6 \times 9$ cm mass which was partially covered by cartilage. The peripheral part of the specimen showed an endochondral ossification process from the surrounding hyaline cartilage. After sectioning the mass, a peripheral rimming of mature lamellar and woven bone was found. Moving towards the edge, there were islands of disorganized osteoid. In the centre of the specimen some trabeculae delimited multiple spaces occupied with an irregular mass of immature fibroblasts and fibroadipose tissue that contained blood vessels. The deepest intertrabecular spaces were occupied by myeloid bone marrow. No increased mitotic activity and no other histological feature suggesting malignancy were found. These histological findings were consistent with myositis ossificans.

The patient was followed-up every 3 months during the first year after surgery, then yearly, for five years to-date. He is currently free of symptoms, with no signs of tumour recurrence, and is professionally active.

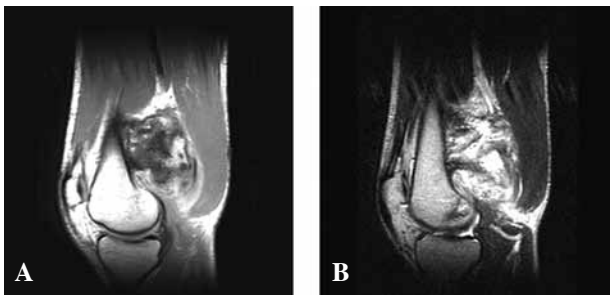

Fig. 5. — Magnetic resonance imaging revealed a mass on the posterior cortex of the distal femur. A) The periphery of the lesion showed a hardly defined edge and a heterogeneous enhancement of the mass periphery on T1-weighted images after contrast administration. B) On T2-weighted images the mass turned out to be heterogeneous and displayed a markedly increased signal. The mass displaced the popliteal vessels posteriorly.

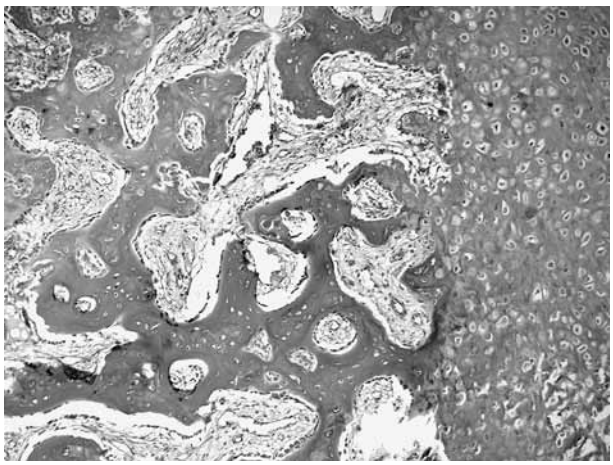

Fig. 6. — Histological study from the incisional biopsy showed a specimen that was partially covered by cartilage. The peripheral part of the specimen showed an endochondral ossification process from the surrounding hyaline cartilage.

DISCUSSION

Myositis ossificans (MO) was initially described in 1883 by Reidel as a reparative process that causes benign heterotopic ossification in soft tissue. Later in 1918, Déjerine and Ceillier reported that MO commonly occurred among soldiers who had experienced spinal cord trauma as combatants in World War I (*3*).

This aberrant reparative process that causes benign heterotopic ossification in soft tissue should not be confused with dystrophic and metabolic (also known as "metastatic") calcification, as may be observed with chronic hypercalcaemia or associated with renal osteodystrophy, connective tissue disease and hormonal imbalance. It may occur in morbid tissues and tumours as well. Metabolic calcification usually results in a generalized mineral deposition that includes visceral organs. It has been associated with abnormal calcium and/or phosphate levels.

The acquired form of MO is observed after spinal cord or central nervous system injuries. Therefore it is known as posttraumatic neurogenic MO. This type of MO often occurs among patients with recent spinal cord injury, frequently young adult patients of either sex. MO develops only in sites distal to the level where the spinal cord injury exists. Cranioencephalic trauma, brain strokes and brain tumours may also lead to MO (*5*). Besides, it also often appears after orthopaedic procedures such as arthroplasty surgery, joint fractures, joint dislocations or soft-tissue trauma. The quadriceps femoris and brachialis muscle have been described to be often involved (*18*).

In those clinical contexts (trauma, after surgery, spinal cord injury) radiographic changes may be diagnostic, but may not be apparent for 3 to 6 weeks. Plain radiographs typically show a floccular calcified density in soft tissues between the 2nd and 6th week from onset. After one year, a mature bony zone typically appears at the periphery of the lesion, which features a central lucency. Then the calcification becomes sharply circumscribed.

On the opposite, when there is no history of trauma, the diagnosis may be more difficult, as in the case reported. In this case, furthermore, the lesion is deeper in the thigh and seems to involve the periosteum of the femur. This lesion, called parosteal MO, may mimic parosteal osteosarcoma.

Half of all ossifications may be deeper in the thigh and adhere to the periosteum (*11*). In these cases, even though the history, physical examination and radiographic findings can be suggestive of MO, the diagnosis may be uncertain. Plain x-ray imaging may help in the initial differential diagnosis, but it

does not provide sufficient specificity to establish the definitive diagnosis of atraumatic MO. Neither myositis ossificans nor parosteal sarcoma do elevate the periosteum, since they both grow on its surface. This special feature may be useful in the differential diagnosis with other bone neoplasms (*16,19*).

Cross-sectional CT and specially MRI images are useful diagnostic tools, assessing extension of the tumour into the medullary cavity or identifying areas with dedifferentiation. When CT or MRI reveals extension of the lesion into the medullary cavity, it is mandatory to make the differential diagnosis with high-grade surface osteosarcomas. Reported studies have shown intramedullary extension in 8 to 59% of parosteal osteosarcomas. Higher-grade tumours have more frequent intramedullary involvement (*12*).

In MO, the histology shows that ossification has three different zones : the central undifferentiated zone, the surrounding zone of immature osteoid formation, and the peripheral zone with mature bone. At least 10 days after symptoms onset are required for these zones to become apparent. This is why biopsy of these lesions should be directed to the peripheral areas. Even if the biopsy is performed before 10 days have elapsed or if a biopsy sample is obtained from the central region, the specimen yields undifferentiated tissue which may resemble an osteosarcoma. Either an incisional or CT-guided biopsy can be used to rule out a high-grade lesion. In contrast to osteosarcoma, MO exhibits a zone pattern, the lesion has viable muscle fibres, and myositis ossificans does not invade surrounding tissue. A specimen obtained from a biopsy performed after ossification maturation reveals primarily mature lamellar bone. The histological features of lesions from patients with non-traumatic MO may lack the typical histological appearance of MO.

MO and osteogenic sarcoma may be difficult to differentiate (*1*). Malignant transformation of the ossified region can occur. Thus, it is worth remembering the different calcification pattern of both lesions. In osteosarcoma the calcification extends from center to periphery, whereas in MO the calcification first occurs in the periphery of the soft tissue mass. Moreover, in MO the calcification occurs in association with the bone's diaphysis, unlike osteogenic sarcoma's usual association with the metaphysis. Parosteal osteosarcomas do not elevate the periosteum since they grow on its surface (*4,10,11,17*).

Cases of MO have also rarely been described after burns, in sickle cell anaemia, haemophilia, tetanus, poliomyelitis, multiple sclerosis as well as toxic epidermal necrolysis (*6*). Finally, some cases of idiopathic MO occur with no recognized precipitating condition.

Inclan *et al* (*7*) differentiated tumoral calcinosis from the dystrophic and metabolic calcification. Tumoral calcinosis is a rare familiar disease characterized by solitary or multiple painless periarticular masses (*13*). The soft-tissue lesions of tumoral calcinosis are typically lobulated well-demarcated calcifications that are mostly distributed along the extensor surfaces of large joints. Tumoral calcinosis calcification features a typical appearance : amorphous, cystic and multilobulated that is located in a periarticular distribution, while the cystic appearance shows fluid-fluid levels. MO can be radiographically distinguished from calcinosis in tumoral calcinosis by its rapid evolution from faint calcification to organized cartilage and bone, and lack of lobular morphology. Late lesions, also called heterotopic ossification, are clearly different from tumoural calcinosis because of their organization into bone with a distinct cortex and medullary space.

Early in the course of the disease, MO may cause pain, fever, swelling, erythema and a mild decrease in the range of motion of the joint. In this early inflammatory phase, the condition may mimic cellulitis, thrombophlebitis, osteomyelitis or a tumour (*8,14*). Later, marked reduction in range of motion and ankylosis of the joint may occur. Clinicians often turn to conventional radiography followed by 3-phase radioisotope bone scanning to confirm the diagnosis of MO and establish both the extent and the metabolic activity of this ossifying lesion. Other complications of MO include peripheral nerve entrapment and pressure ulcers (*2*).

Surgical management is warranted only in patients with non-hereditary myositis ossificans and

only after maturation of the lesion. Surgery has been advocated when lesions mechanically interfere with joint movement or impinge on nerves. Surgery may also be indicated when diagnosis is uncertain. MO is likely to recur and possibly progress if resection is undertaken before the lesion has become mature. With a view toward avoiding recurrent MO and other operative complications, serial quantitative bone scans are used as an aid to schedule the surgical intervention. As treatment or prophylaxis for MO, either indomethacin, a bisphosphonate or local radiation have been recommended.

Protected weight bearing, avoidance of early heat, massage and stretching reduce the tendency to form ectopic bone. Treatment for myositis ossificans should be delayed until signs of maturation of the bone mass become manifest.

Although the various presentations of MO have been recognized for many years and numerous methods of diagnosis and treatment have been applied, the aetiology of MO and its prevention or treatment remain elusive.

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