

Case Resolution

Ricardo Trueba

Magnetic Resonance and Computed Tomography Department, Grupo Médico Rostagno, Diagnóstico por Imágenes (Buenos Aires, Argentina)

See case presentation on page 289.

DIAGNOSIS: Myositis ossificans

DISCUSSION

Myositis ossificans is the most common form of heterotopic ossification, usually within large muscles. Its importance stems in large part from its ability to mimic more aggressive pathological processes. In more than 75% of cases, this disease is associated with trauma or a history of repetitive microtrauma. To a lesser extent, it is related to surgical procedures. It is also detected in patients with long ICU stays for head trauma, spinal trauma or neurological disorders, even patients with severe burns. The most common presentation sites are: elbow, shoulder, pelvis and thighs. It can also involve muscles, tendons, ligaments and aponeurosis.

Some conditions are related to myositis ossificans or share a similar name, including:

- Circumscribed myositis ossificans: it refers to new bone that usually appears after trauma (Figures 4 and 5).

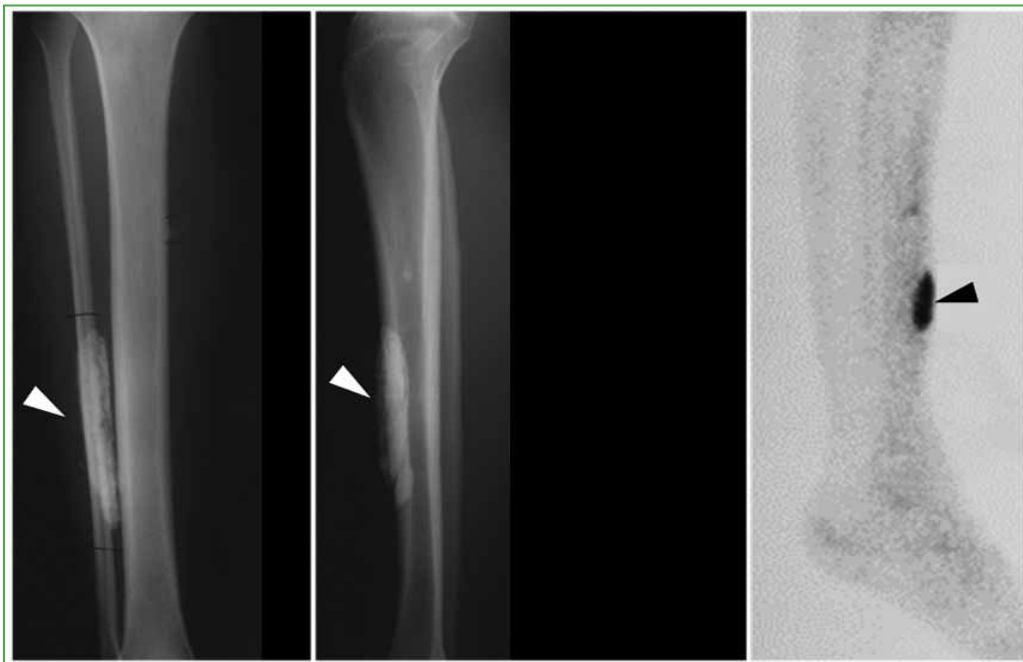


Figure 4. Myositis ossificans in the mature phase. Leg radiographs show tibial calcification adjacent to the shaft and bone scan uptake.

RICARDO TRUEBA, MD • ricardotrueba@gmail.com  <https://orcid.org/0000-0001-7908-817X>

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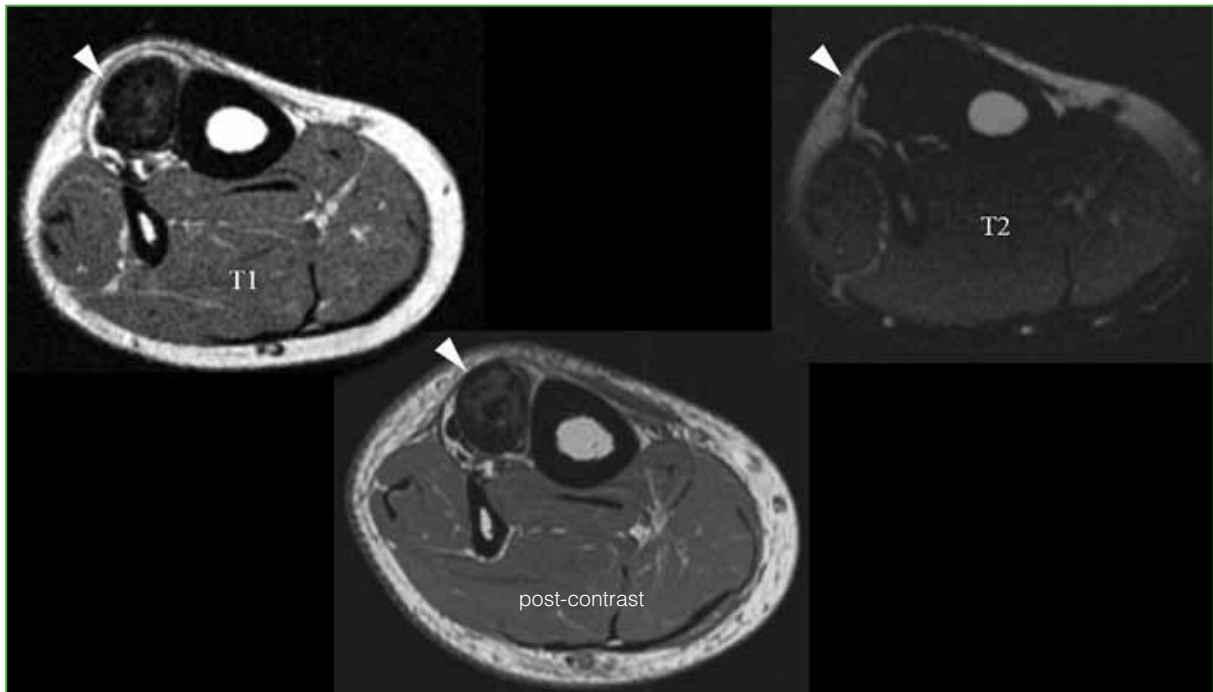


Figure 5. MRI of the leg with contrast medium. In the T1- and T2-weighted sequences, a low signal image is observed adjacent to the tibial shaft with little post-contrast enhancement.

- Myositis ossificans progressiva: it is a rare inherited disorder characterized by fibrosis and ossification of muscles, tendons and ligaments at multiple sites; it is disabling and ultimately fatal (**Figure 6**).
- Panniculitis ossificans: similar to myositis ossificans, but affects the subcutaneous tissues.

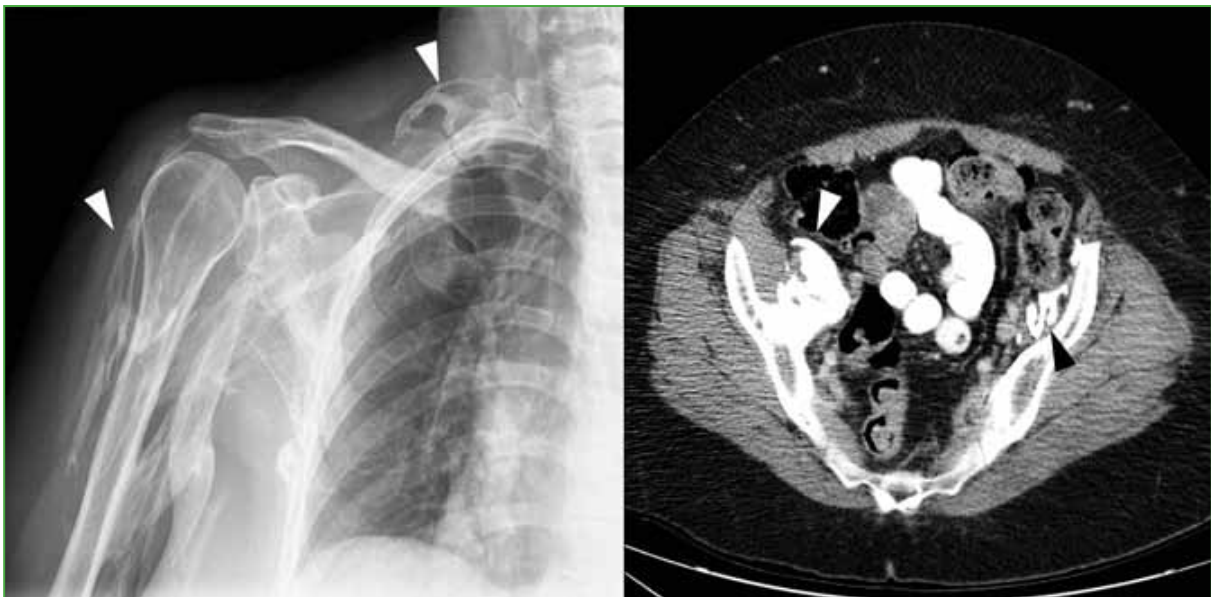


Figure 6. Myositis ossificans progressiva. On the right shoulder radiograph, multiple soft tissue calcifications are recognized, and on computed tomography, progressive calcifications are seen in the psoas.

Most cases of myositis ossificans occur as a result of trauma, and therefore the primary demographic is young adults. Another group especially prone to myositis ossificans are paraplegic patients, generally without evidence of trauma.

It typically presents as a painful, tender, enlarging mass, which in 80% of cases is located in the large muscles of the extremities, often following recognized local trauma, although a definite traumatic event is not always recalled. In paraplegic patients, recognized traumatic events are often absent, and the disease occurs especially around the knees and hips. Myositis ossificans is essentially a metaplasia of the intramuscular connective tissue that results in extraosseous bone formation (without inflammation).

There are three well-described histopathological stages:

1. First month: the tissue injury causes organizing granulation tissue with fibroblastic and osteoblastic differentiation and osteoid formation.
2. Second month: mineralized osteoid matrix develops with immature lamellar bone.
3. Third month: immature bone progresses to mature lamellar cortical and trabecular bone.

Unfortunately, the histological appearance of myositis ossificans may appear similar to that of osteosarcoma and therefore can lead to inappropriate management.

The typical radiographic appearance of myositis ossificans is a circumferential calcification with a lucent center and a radiolucent cleft that separates the lesion from the cortex of the adjacent bone. On plain radiographs, initially, there is no calcification, but there may be soft tissue inflammation. Calcification usually manifests within 2-6 weeks, and the lesion reaches the classic well-circumscribed peripherally calcified appearance by two months. Over the following four months or so, the calcifications usually become smaller and denser. The radiolucent cleft can be difficult to see on plain radiographs.

The appearance on computed tomography images is similar to that of conventional radiography, demonstrating mineralization proceeding from the outer margins towards the center. The cleft between the calcifications and the subjacent bone is usually visible. The peripheral rim of mineralization is visible within 4-6 weeks.

The appearance on MRI changes with the age of the lesion. Early features can be misleading because the peripheral calcification is poorly visualized and soft tissue edema can extend beyond the often inapparent calcific rim.

- T1: ill-defined isointense to muscle mass.
- T2: periphery: high signal (edema) observed up to eight weeks; central: heterogeneous high signal, due to high proliferating cellularity and cartilaginous components. Fluid-fluid levels have been reported and attributed to previous hemorrhage.
- T1 with contrast medium: enhancement is often present.

Late features mimic bone:

- T1
 - periphery: low signal (mature lamellar bone)
 - central: intermediate to high signal (bone marrow)
- T2
 - periphery: low signal (mature lamellar bone)
 - central: intermediate to high signal (bone marrow)
- T1 with contrast medium: generally none in mature lesions.

In the early stages of lesion development, nonspecific increased bone scan uptake is observed, gradually decreasing as the lesion matures.

FDG PET can demonstrate intense uptake mimicking high-grade lesions.

Myositis ossificans is benign and there is no compelling evidence that malignant degeneration ever occurs. As such, treatment is reserved for symptomatic lesions, and surgical resection is usually curative.

DIFFERENTIAL DIAGNOSIS

In imaging studies, the differential diagnosis should include:

- soft tissue sarcomas, including malignant histiocytoma or synovial sarcoma. There is no traumatic history (Figure 7).
- Paraosteal osteosarcoma: it calcifies in the center and continues towards the periphery, and affects the metaphyseal region more (Figure 8).

Treatment of this disease seeks to improve symptoms and may include radiation therapy, non-steroidal analgesics that inhibit the formation of calcifications, and surgery. It is important to clarify that, when indicating surgery, it is necessary to add the three strategies mentioned.

Radiotherapy: This treatment is essential to control and even reduce the masses related to myositis ossificans. It is used both in the prevention of surgeries that could potentially generate myositis ossificans, and in the surgical treatment of the extraction of masses of myositis ossificans to prevent their recurrence. There is no consensus on the dose, but, in general, it depends on the size, the location, and if it is preventive or adjuvant.

Nonsteroidal anti-inflammatory drugs: They are the drugs most used in the prophylaxis of myositis ossificans. Indomethacin is the gold standard, 25 mg are given 3 times a day for 6 weeks.

New studies show good results with COX-2 inhibitors, such as celecoxib, for the prophylaxis of myositis ossificans.

The surgical indication to remove myositis ossificans is directly related to the symptoms and how these affect the quality of life of the patient.

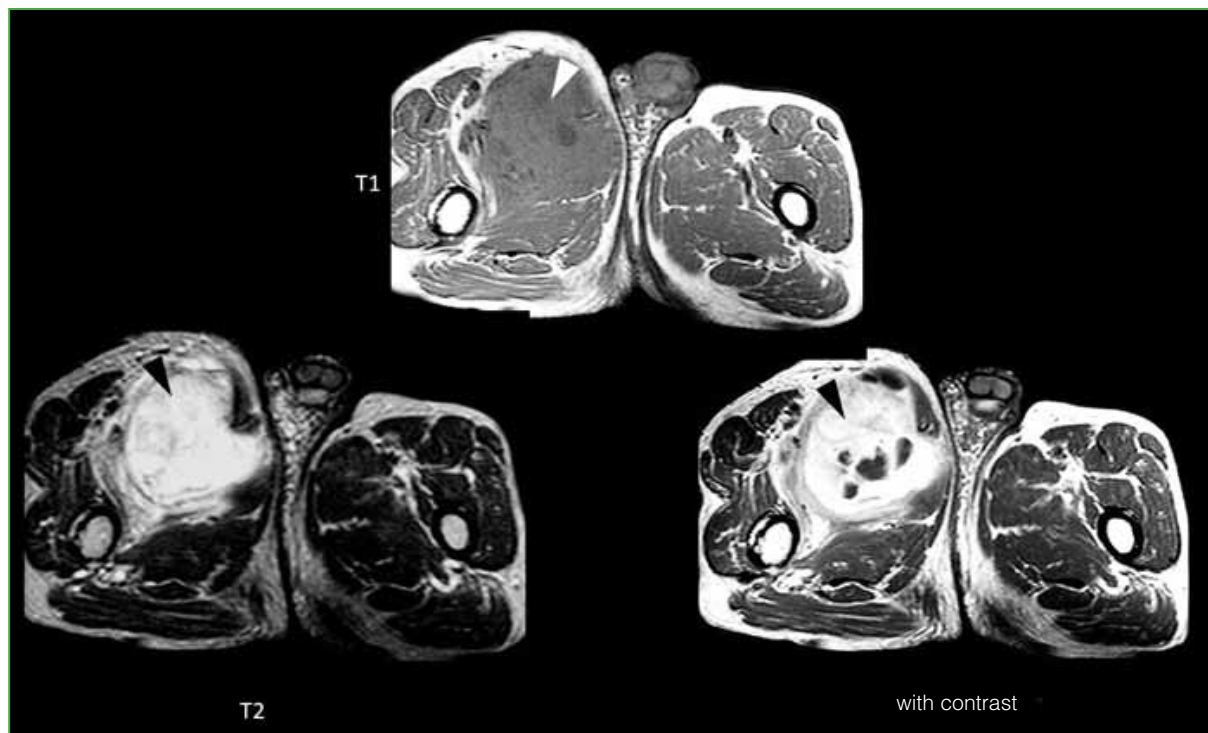


Figure 7. MRI of the pelvis with contrast medium. In the T1-weighted sequence, an expansive lesion with an intermediate signal is observed in the inner region of the proximal third of the right thigh. In the T2-weighted sequence, the lesion is evidently hyperintense. Following the injection of the contrast medium, a marked enhancement is seen with central areas of low signal not taking up the contrast medium.

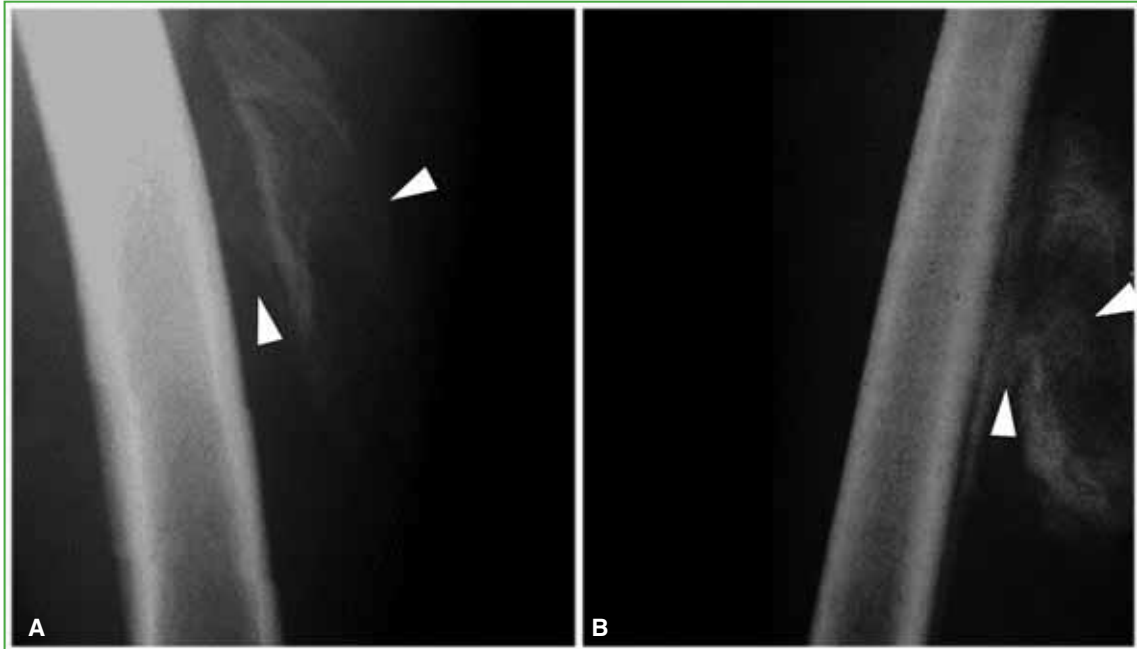


Figure 8. Radiographs of the femur in different patients showing the difference between myositis ossificans (**A**) and paraosteal osteosarcoma (**B**). In myositis, the calcification is peripheral and radiolucency (cleft) is recognized with the adjacent bone. In sarcoma, the calcification is central and there is continuity with the neighboring bone structure.

An important point to clarify is that surgery must be performed 6 months after the appearance of myositis ossificans, that is, it must “mature” before the procedure. The recurrence rate is high when the surgery is carried out earlier. It is important to remember that the surgical procedure must be accompanied by prophylactic radiation therapy and non-steroidal anti-inflammatory drugs.

The patient presented was treated with a single dose of radiotherapy, physiotherapy and indomethacin 25 mg, 3 times a day, for 6 weeks, with a good restoration of quality of life, but the volume of myositis ossificans was not significantly reduced.