Myositis Ossificans

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Definition: Myositis ossificans is a self-limiting benign lesion that can arise in any type of soft tissue most frequently in muscle as a solitary lesion. It consists of a process in which soft tissues are interested by mature lamellar bone formation in association with inflammation mostly caused by traumatic or neurological injury surgery burns or other diseases.

Fibrodysplasia ossificans progressiva (also called Munchmeyer's disease) is a hereditary type of myositis configuring an extremely rare genetic disease. Usually sporadic via a gene mutation, but it may also be due to an autosomal dominant hereditary disorder (mutations in a BMP type 1 receptor).

Epidemiology: Classic and fibrodyslpasia ossificans progressiva are very rare. Young active males are the most commonly affected.

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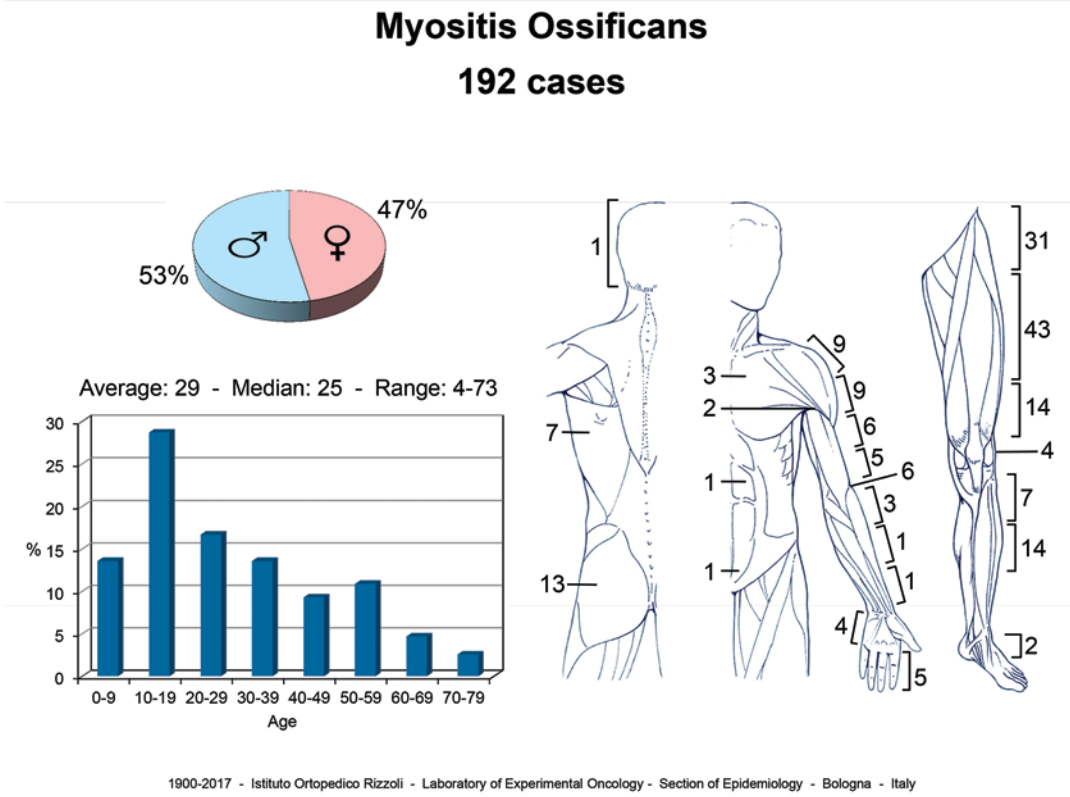


Clinical: The classic MO scenario is pain associated with a growing soft tissue mass usually after a trauma or injury. Even repetitive minor trauma can lead to the development of MO. At the beginning of symptoms, tissues are involved by intense inflammatory response as well as swelling, warming, and rubbishness. Then inflammation and swelling tend to decrease even without any treatment after few days. Pathophysiology is under discussion. During this phlogistic process, a cascade of cytokines activates endothelial mesenchymal cells and induces their differentiation in osteoblasts and chondroblast cells. The final result is bone formation within soft tissue.

The entire process passes through an early, intermediate, and mature stages; each stage has a correspondence between clinical, radiographic, and histopathological aspects (Table 26.1).

So a painful enlarging mass with intense inflammation of the surrounding tissues can be appreciated. Inflammation resolves itself with rest, then the mass slowly regresses. After that a maturation of the mass occurs in several months up to 1 year.

Genetic type: progressive and diffused ossification of tendons, ligaments and connective tissue around muscles, micro-clinodactyly.

Localization: The post-traumatic type may develop in any site. More frequent in the extensor muscles of the thigh, flexor muscles of the arm, then adductor or gluteal muscles. MO of psoas muscle has been reported as well. The genetic type usually starts in the neck, shoulder, axilla, or paraspinal muscles.

Imaging: Standard X-rays in the early stage usually are negative, even if a periosteal reaction can be detected in the first 2 weeks if the process

Table 26.1 Clinical and radiographic findings of myositis ossificans at each stage

Stage	Early	Intermediate	Mature
Clinical aspects	Pain and swelling, functional limitation, rubbishness	Tender soft tissue mass, persistence of functional limitation, decreasing pain	Reduction or absence of symptoms, a hard mass can be appreciated
Imaging	Standard X-ray usually negative	Calcified peripheral border with a radiolucent central area	Radiodense mass with persistent radiolucent central area
Histology	Spindle bland-appearing and mitotic activity	Central immature woven bone associated with peripheral lamellar mature bone formation (zonation)	Maturation of the whole mass into lamellar bone

arises close to the bone. After 3–4 weeks, soft tissue calcifications become evident. Then calcifications tend to mature and organize themselves in a mature ossification at the periphery with a radiolucent central area (6–8 weeks). At the end a rounded calcific mass is clearly shown (6–12 months).

US scan: It is often the first approach to such a lesion. In the early stage three concentric zones can be detected: a peripheral hypoechoic area, an intermediate hyperechoic area with calcification, and an inner hypoechoic area corresponding to immature zone. Despite these characteristics, it remains dependent on the operators' skill, so further examination is the rule.

CT scan can address and lead to the diagnosis, showing the classic pattern of peripheral calcification and radiolucent central area only after few weeks. In the early stage a hypo/isodense soft tissue lesion is visible. So CT scan is very useful and diagnostic in the intermediate stage, while it is neither specific nor helpful in the early stage.

Although MRI is the gold standard examination for soft tissue masses, in MO it can be confounding in particular in the early stages. In the first phase, inhomogeneous T1 and hyperintense T2 and STIR signals are present at the same time corresponding to various phenomena (hematoma, fibroblast proliferation, hemosiderin deposit, edema). These findings may lead to erroneous diagnosis of sarcoma.

After the acute phase (4–6 weeks), the central part of the lesion becomes iso-hypointense in T1 and slightly hyperintense in T2 to surrounding muscles and a low T2 signal at the periphery can be appreciated (peripheral ossification), and in STIR, edema has been resolved.

Then in the mature phase, the lamellar bone pattern and a low signal in all the sequences are the MRI features.

Histopathology: Histology depends on the phase. In the early phase a proliferation of bland-appearing spindle cells with plump nuclei, arranged in a storiform pattern with mitotic activity and scattered inflammatory cells, similar to nodular fasciitis, is seen. In the intermediate phase, the lesions show a central areas with the same spindle cell proliferation, surrounded by immature woven bone (zonation). In the last phase, mature lamellar bone at the periphery is evident.

Course and treatment: Although classic type MO has a benign course, with a self-limiting behavior, a biopsy is often performed in the early stage due to differential diagnosis with more aggressive lesions. The best approach is a US-guided needle biopsy in which the different areas (from periphery to the inner part) must be included.

Treatment is rest, cryotherapy, and anti-inflammatory drugs. Genetic type: progressive severe disability. If respiratory muscles are involved, disease may be fatal.

Treatment: No surgical treatment is usually required. In genetic diseases, good general care and avoidance of trauma (particularly the iatrogenic ones from i.m. injections, biopsies, surgery) are emphasized (Figs. 26.1, 26.2, and 26.3).

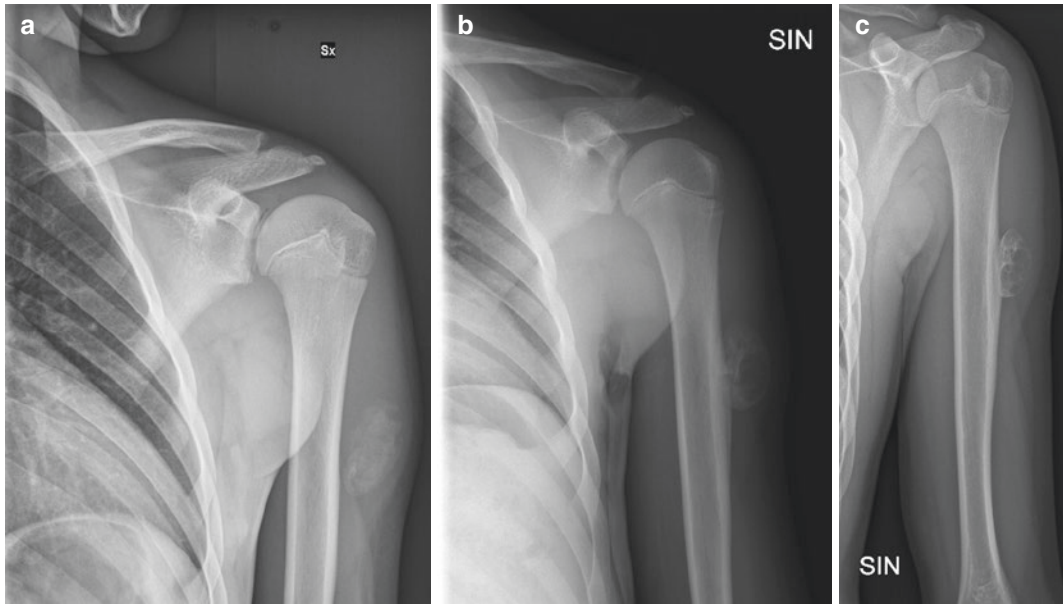


Fig. 26.1 (**a**, **b**, **c**) Evolution of myositis ossificans on standard X-ray. (**a**) X-ray at 3 weeks after clinical onset shows soft tissue calcification in the left arm, (**b**) X-ray at 3 months shows a calcified peripheral border with a radiolucent central area, (**c**) X-ray at 8 months shows a radiodense mass with a persistent radiolucent central area

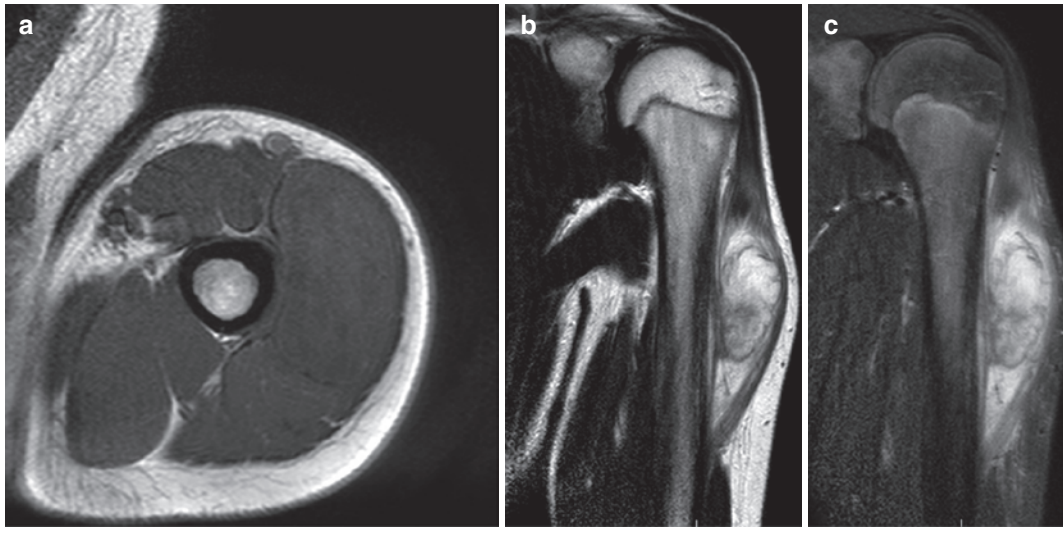


Fig. 26.2 MRI with T1 (**a**), T2 (**b**), and STIR (**c**) signals at 1 month after the onset of symptoms. Inhomogeneous T1 and hyperintense T2 and STIR signals are present at the same time

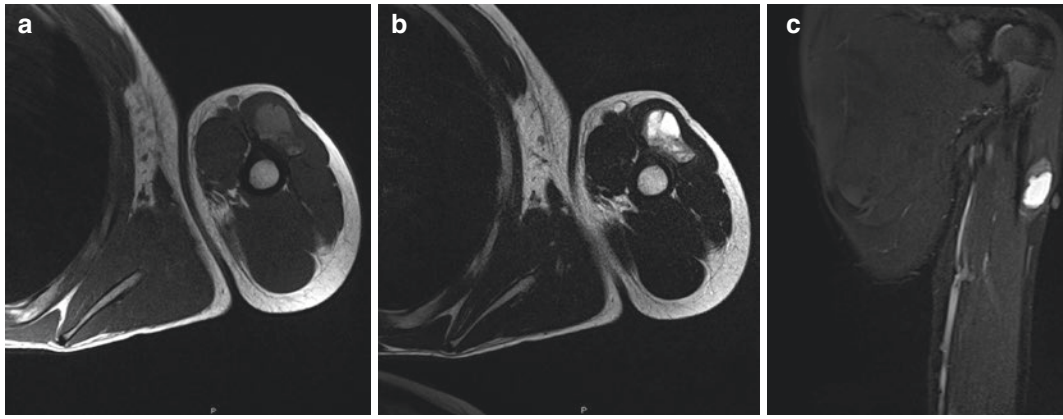
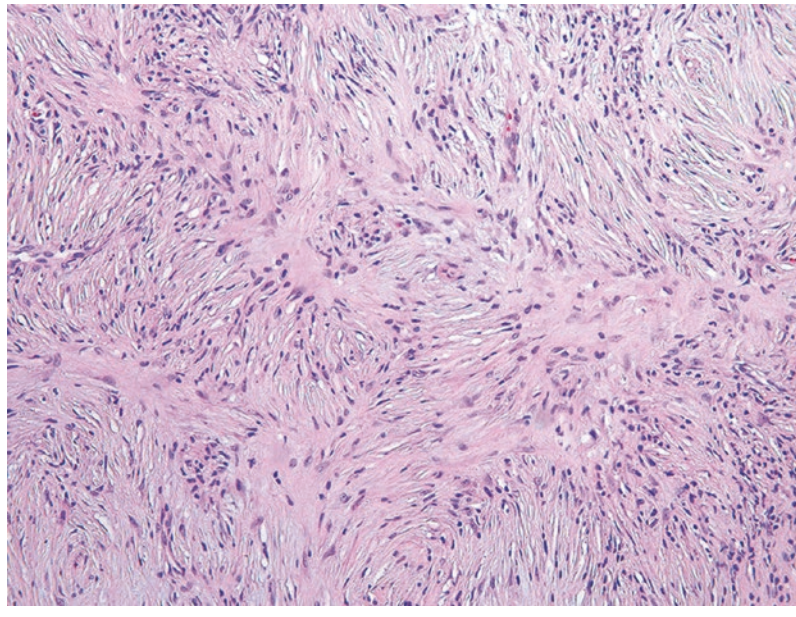
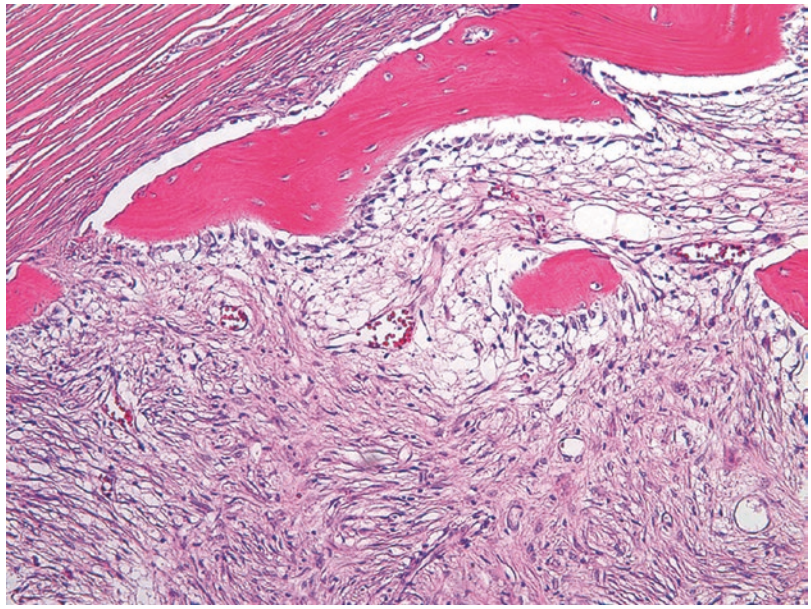


Fig. 26.3 MRI with T1 (**a**), T2 (**b**), and STIR (**c**) signals at 3 months after the onset of symptoms. The central part of the lesion becomes iso-hypointense in T1 and slightly hyperintense in T2 to surrounding muscles, a low T2 signal at the periphery can be appreciated (peripheral ossification) and in STIR edema has been resolved



Storiform pattern of bland-appearing spindle cells and scattered inflammatory cells in the center of the lesion



Ossified rim at the periphery (zoning phenomenon)

Selected Bibliography

Abate M, Salini V, Rimondi E, Errani C, Alberghini M, Mercuri M, Pelotti P. Post traumatic myositis ossificans: sonographic findings. J Clin Ultrasound. 2011;39:135–40.

Bauer AH, Bonham J, Gutierrez L, Hsiao EC, Motamedi D. Fibrodysplasia ossificans progressiva: a current review of imaging findings. Skelet Radiol. 2018;47(8):1043–50.

Martin DA, Senanayake S. Images in clinical medicine. Myositis ossificans. N Engl J Med. 2011;364(8):758.

Parikh J, Hyare H, Saifuddin A. The imaging features of post-traumatic myositis ossificans, with emphasis on MRI. Clin Radiol. 2002;57(12):1058–66. Review.

Sferopoulos NK, Kotakidou R, Petropoulos AS. Myositis ossificans in children: a review. Eur J Orthop Surg Traumatol. 2017;27(4):491–502.

Walczak BE, Johnson CN, Howe BM. Myositis ossificans. J Am Acad Orthop Surg. 2015;23(10):612–22.