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Case Report

Non-traumatic myositis ossificans circumscripta: A diagnosis trap

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ABSTRACT

Myositis ossificans circumscripta (MOC) is a benign condition of non-neoplastic heterotopic bone formation in the muscle or soft tissue. Trauma plays a role in the development of MOC, thus, non-traumatic MOC is very rare. Although MOC may occur anywhere in the body, the lesions are localized predominantly in the high-risk sites of injury, such as the thigh, buttock, and elbow. MOC can easily be mistaken for osteomyelitis or a malignant tumor, specifically osteosarcoma or soft-tissue sarcoma. We report a rare case of non-traumatic myositis ossificans circumscripta of thigh which appear clinically and radiologically as a malignant neoplasm. Despite its rarity, MOC should be contemplated in the differential diagnosis of malignant tumors.

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1. Introduction

Non-traumatic MOC is a rarely reported benign heterotopic ossification characterized by the aberrant formation of bone in extraskeletal soft tissues. It is usually confined to a single muscle or muscle group, and is most common in active male within the second and third decade of life.¹ It usually occurs after muscle injury such as repeated microtrauma, but it can also occur without previous trauma. In a small number of cases, possible etiologies include infections, burns, neuromuscular disorders, hemophilia (factor-IX deficiency), tetanus, and drug abuse. Clinically and radiologically, it is difficult to distinguish this disease from soft tissue and bone

malignancy, so a biopsy is necessary to confirm the diagnosis.^{2,3}

2. Case report

A 27-year-old woman presented with a 7 month history of insidious onset of an isolated left thigh mass, located dorsally at the junction of middle and distal third of the thigh. The mass was painful, and slowly enlarging. The patient denied any history of trauma, fever, chills or weight loss. She wasn't involved in professional or recreational sports activities. Physical examination revealed a 7 × 8 cm, soft and painful mass in the distal one-third of the left thigh, little sensitive to

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Fig. 1 – Plain radiographs revealed no specific bone abnormalities and didn't show any calcifications facing the mass (at 2 month).

pressure, firmly attached to deeper tissues. There were no motor deficits or neurologic symptoms. The pain was not severe enough to disturb sleep or to hinder physical activities but worsened after muscular contraction and mobilization of neighboring joints (i.e. hip and knee). There is no stiffness of the knee with normal joint amplitudes. Laboratory tests revealed normal white blood cell count ($6400/\text{mm}^3$), a slight increase of Erythrocyte Sedimentation Rate (31 mm/h) and C-reactive protein (63 mg/dl), alkaline phosphatase levels of 90 U/L, calcium levels of 10 mg/dl, and creatine phosphokinase levels of 200 U/L. Plain radiographs initially revealed no specific bone abnormalities and didn't show any calcifications

facing the mass (Fig. 1). Radiography performed 5 months later showed an oval mass with a regular border containing dense peripheral calcifications with centripetal evolution; these ossifications were distant from adjacent bony structures associated with unilamellar periosteal reaction of the underlying bone (Fig. 2). Magnetic resonance imaging (MRI) objectified tumoral process of posterior muscular compartment of the left thigh with crown aspect (periphery with hyperintense signal on T2 and center with hypointense signal on T2 and hyperintense signal on T1) (Fig. 3A and B). This process measured 6×4 cm with refolement of vessel elements (Fig. 3C). Surgical biopsy was performed in the posterior face

Fig. 2 – Radiography performed 5 months later showed an oval mass with a regular border containing dense peripheral calcifications with centripetal evolution; these ossifications were distant from adjacent bony structures associated with unilamellar periosteal reaction of the underlying bone.

of the left thigh (Fig. 4). Histological examination of the specimen revealed fibrous and adipose tissue altered by a mononuclear inflammatory infiltrate associated with a proliferation of spindle cells. These cells showed a regular nuclei, sometimes vesicular and nucleolus. These elements are arranged in small clusters, developing immature osteoid lined by regular osteoblastic cells. Besides this proliferation, there is bone maturation, made by anastomotic and regular cords. These microscopic features were in favor of a myositis ossificans (Fig. 5A and B).

The patient was received radiotherapy with low-dose (600 cGy) in six fractions. At the 2 months follow-up, the

patient had no recurrence of pain and he had full range of motion with a clinical reduction in the size of the tumor. The surgery by resection of the mass will be programmed after the radiotherapy, when the tumor would be sufficiently mature.

3. Discussion

Myositis ossificans (MO) is an inflammatory pseudotumor of the muscle that may be confused clinically, radiologically and histologically with a malignant soft tissue tumor. It is a pseudo-inflammatory tumor that originates from skeletal muscle and corresponds to a heterotopic, metaplastic, non malignant bone tumor.⁴ The etiology and causative factor of MO remain unclear and not easily identifiable. It might develop secondary to a muscular trauma, generally, 75% of all MO cases are associated with trauma.^{4,5} Men and women are equally affected; no preferential anatomical site can be identified. Non-traumatic MOC is less frequent and seems to have no preference for gender. Adolescents and young adults under the age of 30 years are most commonly affected.^{5,6}

The diagnosis of MO is clinical and mainly radiological. MOC is clinically manifested as a very inflammatory, rapidly-growing, and painful muscular mass,⁴ which may be associated with periosteal reaction in 7–10 days. Flocculent dense lesions signifying the onset of ossification arise in the mass from 11 days to 6 weeks. At 6–8 weeks, a lacy pattern of new bone with a sharp peripheral cortex is formed. From 10 weeks to 6 months, the central zone may enlarge and produce the appearance of an “egg shell” at the end stage of maturation.⁷ Plain radiography doesn't reveal any abnormality in the early stages of MO. Radiographs repeated later may show pathognomonic ossification surrounding a clear central area; typically, these ossifications are distant from adjacent bony structures. However, the ossifications are often missed on radiographs when these are performed 2–3 weeks after MO onset.⁴ Computed tomography is the gold standard in characterizing typical findings of myositis ossificans such as extensive muscle and perilesional edema without bone marrow or cortical abnormalities.² The appearance of MO on MRI is variable and depends on the maturity and the variation

Fig. 3 – A) T1-weighted frontal MRI image showing a hyperintense tumoral process occupying the distal third of the left thigh. B) T2-weighted sagittal MRI image showing tumoral process with a hyperintense signal on the periphery and hypointense signal on the center (crown aspect). C) T2-weighted axial MRI image showing tumoral process with refolement of vessel elements.

Fig. 4 – Scar biopsy on the posterior face of the left thigh.

Fig. 5 – A) Presence of immature osteoid foci within the regular myofibroblastic proliferation. B) Presence of mature osteoid spans.

of the histological pattern within the lesion. In the early stages, T2-weighted images may show an inhomogenous focal mass with high central signal intensity. As the lesion matures and the peripheral ossification becomes denser, the images show a hyperintense center surrounded by a hypointense rim corresponding to peripheral ossification.^{2,7} When the radiological diagnosis of MO remains difficult, the realization of a biopsy is required,⁷ but if premature biopsy is done at the early stage of MO, it may lead to a wrong diagnosis of sarcoma, or if biopsy is delayed, a true sarcoma may be missed.⁴

Histologically, the proliferation is composed of mesenchymal cells secreting a myxoid matrix as well as fibroblasts exhibiting numerous mitoses, which gives it a pseudo-fibrosarcomatous appearance. This is followed by the subacute phase, which continues for about 10 days. The fibroblasts differentiate into osteoblasts and secrete an osteoid matrix at the periphery of the initial myxoid zone, giving it a pseudo-osteosarcomatous appearance. The late phase, also called the maturation phase, usually starts between the second and fifth weeks of the evolution of MO. Bone production can be observed at the periphery of the lesion. At this stage, biopsy will reveal the three characteristic zones of MO and thus allow the correct final diagnosis to be made^{4,8}: a central zone characterized by the presence of an inflammatory infiltrate with macrophages, lymphocytes, polymorphic fibroblasts and angiogenesis phenomena, muscular fibers with atrophic or degenerative appearance; an intermediate zone with a more regular appearance with collagen trabeculae and immature, osteoid cells; and a peripheral zone made by calcified osteoid with areas of cartilaginous metaplasia and lamellar mature bone separated from the surrounding muscle by connective tissue without inflammatory infiltrate.⁹ The differential diagnosis can be represented by non-neoplastic pathologies (calcified fibromatosis, local infections) but especially by malignant tumors (lymphoma, osteosarcoma, rhabdomyosarcoma). The most frequently clinical misdiagnoses are tumor, especially extraskelatal osteosarcoma, which has similar clinical and pathological characteristics.¹⁰ The treatment in most cases is conservative; it consists of rest, ice and anti-inflammatory drugs to relieve pain. Typically, a regression of the symptoms is seen in the course of disease (30%). A spontaneous resorption or an incomplete regression can

occur. Radiation therapy can be used to reduce the size and accelerate the maturation of the lesion.⁷ Surgical intervention is recommended when the heterotopic bone has matured. It is also recommended if it interferes with joint motion, functional limitation or if it generates a significant pain, due to the compression of nerve trunk.¹⁰

4. Conclusion

Myositis ossificans circumscripta is a rare benign lesion with an excellent prognosis but may appear clinically and radiologically as a malignant neoplasm, which can make its diagnosis very difficult and sometimes delayed. A thorough knowledge of the clinical and morphological study is necessary to differentiate this lesion from a malignant soft tissue tumor.

Conflicts of interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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