Non-traumatic myositis ossificans circumscripta in the context of vastus medialis obliquus: a case report

Introduction

Myositis ossificans circumscripta (MOC) is an heterotopic proliferation of non-tumoral cells of the bone and cartilage tissues. Clinically and radiologically it is difficult to distinguish this benign disease from soft tissues and bone malignancy. In 1913 Coley (1) reported on three cases of traumatic MO and recognized the difficulty of differentiating it from sarcoma. More recently, Kransdorf et al. (2) defined MO as a benign solitary self-limiting ossifying soft tissue mass, typically occur within skeletal muscles. This is likely the most current accepted understanding of the term. The mass starts from the interstitial connective tissue within the soft parts of the bones. It represents the 0.7% of the pseudotumors in the soft tissues. Most of these cases are attributed to a prior unrecognized trivial trauma (75%) (3). It affects then more frequently the regions of the body who are more prone to trauma, as flexor muscles of the upper limbs, the quadriceps femoris and the abductor muscles of the thigh (4). Male subjects in the second-third decade are mostly affected by this condition (5). When the patient reports no trauma, the clinicians should suspect an underlying systemic pathology, such as hemophilia, causing unrecognized hemorrhages, or other conditions as muscular disorders, infections, tetanus and drug abuse. In the current literature, there are only few case reports about the occurring of myositis ossificans circumscripta without traumas, and there are no case series. That testifies the peculiarity of this condition. We report a non-traumatic case of MOC in the context of the middle-inferior third of vastus medialis obliquus in the right thigh.

Case report

An active 40-year-old woman presented with a 6-month-history of isolated swelling (slowly enlarging) and local pain in the middle-inferior third of the right thigh, on the medial side. The patient reported no history of trauma, fever, infections or weight loss, and reported no comorbidity or familiarity for genetic conditions.

Physical examination showed a palpable mass of approximately 50x40 mm in the context of the vastus medialis. The mass was soft but painful at the palpation, and strongly attached to the deeper muscular layers. Hip and knee joints presented a normal range of motion without mechanic articular pain.

We asked for blood samples examinations, and found that the serum alkaline phosphatase was 155 mU/ml (n.r. 40-150 mU/ml), with a slight elevation of C-reactive protein (1 mg/dL, n.r. <0.5), serum calcium levels of 9 mg/dL (n.r. 8.60-10.3 mg/dL) and a normal white blood cells count. We requested an ultrasound examination, including the color power doppler to exclude vascular pathologies. The exam was negative for vascular anomalies, but showed the presence of an oval intramuscular lesion of 32 x 21 mm, in the context of

Summary

Myositis ossificans circumscripta (MOC) is an heterotopic proliferation of non-tumoral cells of the bone and cartilage tissues. Traumas play an important role in the development of MOC. When the patient reports no trauma, the clinicians should suspect an underlying systemic pathology, such as hemophilia, causing unrecognized hemorrhages, or other conditions as muscular disorders, infections, tetanus and drug abuse. Clinically and radiologically it is difficult to distinguish this benign disease from soft tissues (such as osteomyelitis) and bone malignancy (such as sarcoma). In the current literature there are only few case reports about the occurring of myositis ossificans circumscripta without traumas, and there are no case series. We report a rare non-traumatic case of MOC in an active 40-year-old woman, in the context of the middle-inferior third of vastus medialis obliquus in the right thigh, treated with excisional biopsy and indomethacin prophylaxis. After six months of follow-up, the patient had complete clinical and radiological recovery.

KEY WORDS: myositis ossificans; vastus medialis obliquus; non-traumatic; indomethacin; excisional biopsy.
vastus medialis obliquus, with a calcific surrounding layer (Figure 1). The ultrasound findings were suggesting an isolated MOC, therefore we proceeded with second level imaging and performed an MRI. The MRI was performed with a 1.5 Tesla machine and contrast medium (Dotarem 0.5 nM/ml, 10 ml). The MRI showed a wide region of altered signal, of oval shape and dimensions of 35x24x13 mm (Figure 2). The lesion had an hyperintense signal in the TR sequences and was enhanced by the contrast medium (Figure 3). It was attached to the deep muscular fascia of the poste-

Figure 1 - Longitudinal ultrasonographic view, according to the major axis of the vastus medialis. It shows a peripheral calcific layer with a posterior shadow cone.

Figure 2 - Preoperative MRI: coronal STIR view. It shows an hyperintense oval lesion in the context of the vastus medialis obliquus, with peripheral edema into the muscular fibers.

Figure 3 - Preoperative MRI: axial TSE-T1 view after administration of contrast medium. It shows homogeneous enhancement of the lesion, localized on the posterior deep layers of the vastus medialis obliquus.
rior aspect of the vastus medialis obliquus, and extended to the femoral diaphysis, which showed a local swelling of the periosteum without cortical or cancellous bone involvement. The features were not univocally interpretable, leading to a MOC localization or a mesenchymal malignancy. Therefore a biopsy was mandatory. It was performed three days after the MRI scan. Due the dimensions of the lesion, we decided for excisional biopsy to avoid further surgery with more surgical stress for the patient. The results of the histological examination showed lamellar bone tissue in the context of muscular fragments (Figure 4), with giant multinucleated cells, arranged in small clusters and developing immature osteoid line by regular osteoblastic cells (Figure 5). The sample was richly vascularized with micro blood clots and some inflammatory elements associated with proliferation of spindle cells (Figure 6). There was no evidence of cytological atypical-

We decided then to treat the patient with high doses of indomethacin (a total of 100 mg pd for 3 weeks), to prevent the recurrence of the lesion. The patient was then discharged and we programmed two follow-up MRI, one after 3 months and the second after 6 months. At 3 months of follow-up, the patient had no recurrence of pain and swelling and the physical examination was negative. The control MRI showed only a little area of blood clotting as an outcome of the biopsy (Figure 7), and a slight edema of the femoral periosteum (Figure 8). We decided then to add magnetic therapy to the treatment. The treatment was performed with an 1.5 intensity pulsed magnetic field with a 75 Hz frequency, 6 hours per day for 30 days in total. At 6 months of follow-up, the patient was still in good condition without swelling and pain and the control MRI reported resolution of the periosteal edema and some fibrotic tissue due to the surgical approach.
Non-traumatic myositis ossificans circumscripta is a benign, non-neoplastic generation of bone tissue extra-osseously in the muscle plane. The pathophysiology is incompletely understood (8). Studies have shown that MO has a huge connection with bone morphogenetic protein (BMP) expression (7). Kan et al. (8) demonstrated that the injury to tissues, with the subsequent inflammation, leads to a dysregulation of the stem cells in the muscle, with the switching of them to osteoblastic line and heterotopic bone formation. Normally the clinical, radiological and histological findings progress parallel. The classification of the disease, considering these features, is variable among Authors. In the recent literature (9, 10) three overlapping stages of evolution are commonly described: early, intermediate and mature. The early stage occurs during the first 4 weeks after the injury and the calcifications are not visible at this stage. This makes the diagnosis tricky, because the raise of the inflammatory markers could be related alternatively to infective pathology, malignancy or MO. Between 4-8 weeks (intermediate stage) the calcification becomes radiologically apparent. A marked peripheral bone formation is typical of the mature stage (> 8 weeks) with the lesion clearly visible on X-rays.
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Typically, the clinical presentation shows muscular pain, that differs from a simple muscle strain or contusion because it is normally longer. This uncommon pain is a result of the inflammation of bursae, tendons and joints surrounding the lesion. Symptoms become less evident with the progression of the lesion, and late-presenting patients may have only a mild pain. Paresthesias, weakness, lymphedema and venous thromboembolic disease have been reported with lesions compressing surrounding neuro-vascular structures. It is then useful to perform an ultrasound examination with color Doppler and/or an electroneurographic exam.

In the early phase, the ultrasonography (11) presents three concentric zones: the outer one is hypoechoic, the middle one is hyperechoic and corresponds to the calcific rim, the inner one is hypoechoic due to the central fibroblastic stroma. Despite the advantages of the ultrasonography, the diagnosis must be confirmed with a second-level imaging exam.

The most common second level imaging consists in the execution of a MRI with contrast medium. In the acute phase, the MRI typically shows signs of inflammation with a heterogeneous signal intensity on T1-weighted areas of high signal intensity. T2-weighted images show a heterogeneous localized mass with high signal intensity in the central area. In the later stages, the T2-weighted images show a central hyperintense signal with surrounding hypointense layer, corresponding to the more dense peripheral bone calcifications of the lesion. It is difficult to see the lesion on T1-weighted images, as the signal becomes isointense to the muscle. In that case, the administration of contrast medium is helpful, because it shows an enhancement in that sequence of images.

When the radiological diagnosis of MOC is difficult, the execution of a biopsy is mandatory (12), and anyway the confirmation of the diagnosis is extremely important before proceeding to treatment or interventions (13). The biopsy can be performed with various techniques, like fine-needle aspiration, core biopsy, incisional biopsy or excisional biopsy. The fine-needle aspiration cytology is often confusing and non-conclusive. Therefore, is recommended to perform a core biopsy under CT-scan control. When a minimally-invasive core biopsy is not performable, or when the diagnosis is uncertain, an incisional biopsy should be performed on medium to big lesions. Excisional biopsy is reserved for small, easily accessible lesions, when imaging is consistent to a benign etiology. In the early stages, the histological features are mesenchymal metaplasia, in fact it may be difficult to distinguish MO from sarcoma (10). In the middle stages, the cells differentiate further and the histology shows mixed chondro-osseous differentiation. Mature stages show mature bone (6), with a thin shell of bone covering a central soft red-grey area (9). However, myositis typically exhibits a peculiar picture: a central zone rich in capillary network, surrounded by endochondral and proliferation of fibroblasts. Everything is covered with a more peripheral layer of osseoblastic elements mixed with cartilage. There are no signs of cytologic atypia and no mitotic figures, thus differentiating the diagnosis from that of osteosarcoma.

There are few cases of MOC described in literature. Therefore, the MOC treatment still remains controversial. Ahmed el Bardouni et al. (14) reported a 6x4 cm lesion in the posterior muscular compartment of the thigh, diagnosed as MOC and confirmed with a biopsy. They first treated the lesion with low-dose of radiotherapy (600 cGy in six fractions), as neoadjuvant treatment, and further excise the lesion. In literature, it is demonstrated that radiation therapy can be used to reduce the size and accelerate the maturation of the lesion (12). The timing of surgical excision is debated. Conner et al. (15) suggested that surgery must be performed when symptoms are halted, according to previous literature that affirmed that premature surgery may predispose to recurrence (16). On the other hand, Russo et al. (17) presented a case of non-traumatic MO in the left scapula, with severe pain and lacking of radiologic evidence of calcification. In their experience, early surgical excision resulted in immediate remission of the symptoms. Ogilvie-Harris and Fornasier (18) reported on 26 patients with non-traumatic MO and suggested that early excision has minimal risk of recurrence. Typically, a regression of symptoms is seen in the course of the disease (30%) and a spontaneous resorption or an incomplete regression can occur (14).

The role of COX-2 inhibitors to prevent the recurrence of calcifications is well known in the literature, especially after prosthetic replacement (19). A treatment period of less than 4 weeks is recommended to avoid gastrointestinal injurious effects.

In our case, according to the onset of the symptoms and the radiologic features, we classified the lesion as myositis ossificans circumscripta in the mature stage. Considering the young age of the patient, the dimensions and the easy accessibility to the lesion, according to literature, we decided to proceed for excisional biopsy and avoided neoadjuvant radiotherapic treatments, even because the imaging features were not univocally interpretable. We further decided to add indomethacin to the treatment to prevent the recurrence of the calcification, also according to literature.

Conclusions

MOC is an heterotopic proliferation of non-tumoral cells of the bone and cartilage tissues with an excellent prognosis, but of tricky diagnosis. In fact, despite the conventional radiologic methods, it is always needed to get and histologic diagnosis, especially for differential diagnosis with non-neoplastic pathologies (calciﬁed ﬁbromatosis, local infections) and malignant tumors (lymphoma, osteosarcoma, rhabdomyosarcoma). The optimal timing of excision in not deﬁned yet. A multidisciplinary approach is helpful to accurately diagnose and optimize treatment. Indomethacin is useful to prevent the recurrence of calcification after surgical excision.

References


