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Case report

Two cases of myositis ossificans in children, after prolonged immobilization

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Abstract

Myossitis ossificans (MO) is a benign disorder characterized by heterotopic bone formation in skeletal muscle. It is divided in three types, fibrodysplasia ossificans progressive (FOP), myositis ossificans circumscripta or traumatica (MOT) and myositis ossificans without a history of trauma (non traumatic or pseudomalignant MO). Myositis ossificans is extremely rare in children younger than 10 years. We present the clinical and radiological findings of two 5-year-old children with pseudomalignant MO due to prolonged immobilization. Plain x-ray films and CT scan with their characteristic findings of mature bone in the periphery of the lesion with smooth contour and well separated from the bone, enabled us to diagnose the lesion. To the best of our knowledge, no such cases have been reported in the literature.

Keywords: Myositis Ossificans (MO), Children, Pseudomalignant Myositis Ossificans, Heterotopic Ossification, Non Traumatic MO

Introduction

Myositis ossificans (MO) is a benign disorder that non-neoplastic heterotopic bone is formed in skeletal muscle[1]. It is a focal self limited condition of unknown origin[2] that can present as a painful soft tissue mass. This well defined entity can be divided in 3 types[1]: Myositis ossificans progressiva (fibrodysplasia ossificans progressive (FOP)[3], myositis ossificans circumscripta or traumatica (MOT) and myositis ossificans without a history of trauma, or pseudomalignant MO (non traumatic MO). This type is seen after prolonged immobilization e.g. burns, haemophilia, paraplegia and poliomyelitis[1,3]. MO usually affects adolescents and young adults, most common in the third decade of life. Myositis ossificans is extremely rare in children younger than 10 years, with few cases reported in the literature.

We present the clinical and radiological findings of two 5-year-old children with MO after prolong immobilization due to respiratory insufficiency. According to our knowledge, this is the first report of myositis ossificans after immobilization in children with no history of trauma and no neurological deficit.

Case I

A previously healthy five year old girl was admitted with severe respiratory distress due to pneumonia of the upper and middle right lobe. Her past medical history was clear. Shortly after admission she went on hypercapnic respiratory failure, required intubation and Paediatric Intensive Care Unit (PICU) transfer. Laboratory testing identified Adenovirus type 3. Supportive treatment with mechanical ventilation was continued for a total of 2 months, with subsequent non-invasive support (BiPAP) for one more month, because of co-existing generalized hypotonia due to adenovirus encephalopathy. As a complication of the prolonged immobilization she developed left femoral vein thrombosis.

After a total of 3 months of PICU stay, she was transferred to the pediatric ward, alert and fully orientated, on BiPAP support for 10 additional days. During her admission her respiratory status gradually improved. Her muscle tone and

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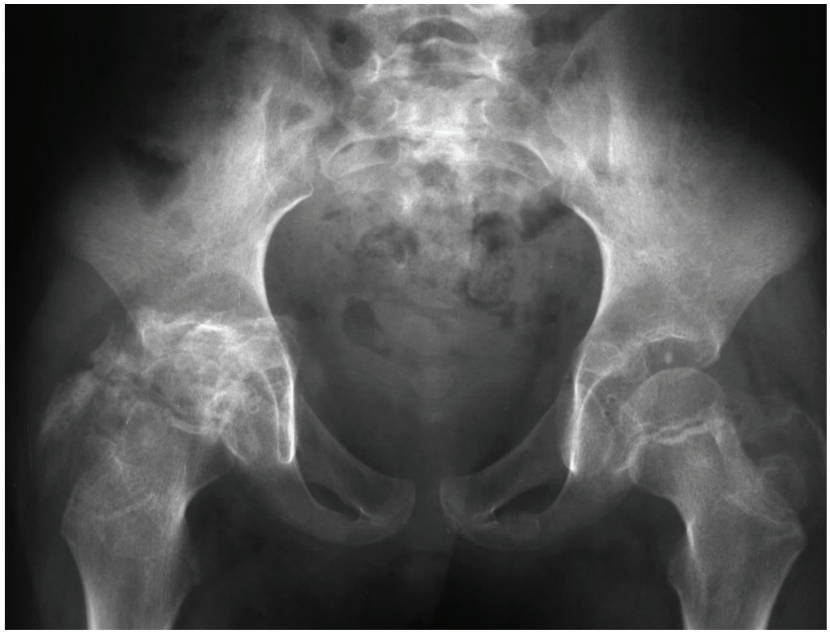

Figure 1. Radiographs of the pelvis show ossifying masses in the soft tissues projecting over the acetabulum, head and neck of both femurs. The lesions are more extensive on the right side. The periphery of the ossifying masses is denser than the center, an indicative finding of myositis ossificans.

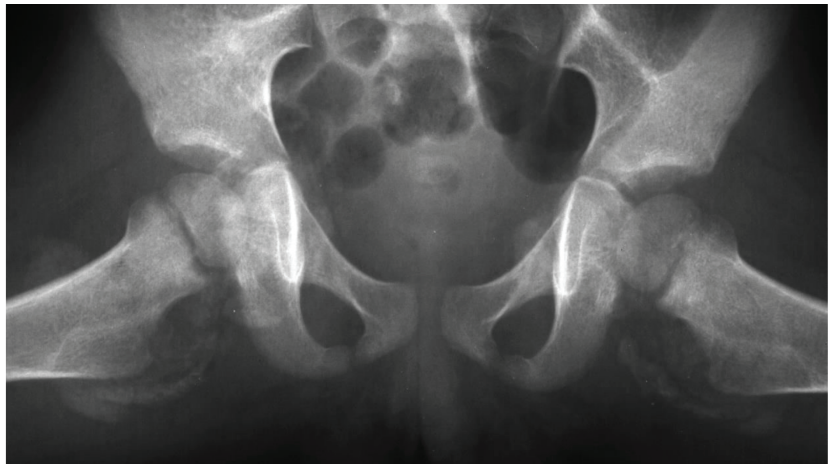

Figure 2. Anteroposterior and frog-lateral projection of both hips reveal extensive heterotopic ossification in the soft tissues adjacent to the medial and lateral cortex of the femoral head and neck on both sides mainly on the right.

power quickly normalized, but when she attempted to walk, she had a limp and complained of a worsening hip pain. On clinical examination both hips had restricted movements. There was marked reduction of internal and external rotation and abduction of the hips. Hip flexion was less affected. There was no previous history of trauma and infection indices were negative. Radiographs of the pelvis revealed ossifying masses in the soft tissues projecting over the acetabulum, head and neck of both femurs. The lesions were more extensive on the right side. They were mainly affecting the gluteus medius with normal bone trabeculae formation and well separated from the neck of the femur on the AP x-ray. The periphery of the ossifying masses was denser than the center. There was absent periosteal reaction of the pelvic bones that appeared normal, (Figure 1). No other joints were clinically found to have restriction of movements. Combining

the symmetrical involvement of heterotopic formation with prolonged immobilization, we proceeded with the diagnosis of non traumatic myositis ossificans. No biopsy was performed.

The patient was started on NSAISs (ibuprofen) and physiotherapy. The patient was regularly followed up, with no signs of worsening. Eleven months later she could walk freely with subsequent radiographical improvement regarding the size of the ossified masses.

Case II

A previously 5 year old boy was transferred to paediatric department after one month stay in PICU, where he was treated for staphylococcal pneumonia of the left lung, complicated by empyema and multi-organ failure. Treatment included *iv* antibiotics, hemodynamic support, mechanical ventilation and pleural drainage. On admission he was clinically stable, with no signs of respiratory distress, but when mobilized, he complained of pain on both hips. Clinical examination revealed restricted range of motion, mainly reduced abduction, extension and internal rotation of hips bilaterally. History of trauma was specifically negative. Clinical examination and blood test showed no evidence of inflammation. We performed hip X-ray with anteroposterior and frog-lateral projection of both hips. Films showed extensive heterotopic ossification in the soft tissues mainly on the posterior capsule, affecting the gluteus muscles. It was clearly separated from the bone surface. Pelvic bones were normal, no signs of periosteal reaction were found (Figure 2). Subsequent axial and coronal CT reconstruction images of the pelvis demonstrated shell-like ossifications in the quadratus femoris, obturator internus and mainly the gluteus muscles on both sides especially on the right (Figure 3). Bone scintigraphy with 99Tc-diphosphonate demonstrated increased uptake in affected muscles. Based on the history and radiographic symmetrical findings, MO was diagnosed and the patient was started on ibuprofen and physiotherapy. No biopsy was performed. Eight months later, repeat CT images showed absorption of the ossified lesions (Figure 4) and 9 months later hip X-ray showed only a small curvilinear ossification in the soft tissues lateral to the RT femoral head. Twelve months later the child was in good condition, with no signs of recurrence.

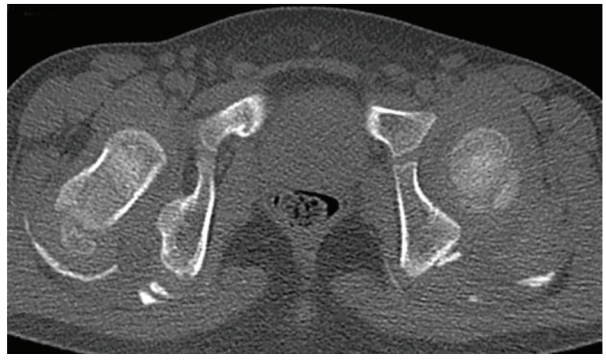

Figure 3. Axial and coronal CT reconstruction images of the pelvis demonstrate shell-like ossifications in the quadratus femoris, obturator internus and gluteus muscles on both sides especially on the right. These appearances are compatible with a diagnosis of myositis ossificans.

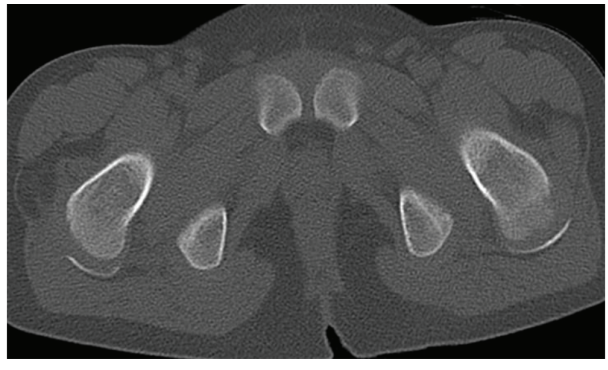

Figure 4. Follow up CT scan after 8 months shows considerable improvement regarding the extent of the heterotopic bone formation and further absorption of the ossified lesions.

Discussion

Myositis ossificans is a benign heterotopic bone formation in soft tissue and skeletal muscle. Non traumatic myositis ossificans or pseudomalignant MO (non traumatic MO) is characterized from bone formation in muscle tissue with absent trauma. In children it is difficult to exclude always the possibility of minor or repeated incidence of trauma[4-6]. In our patients there was prolonged period of immobilization in ICU[1]. No clinical, radiological or histopathological differences exist between traumatic and non traumatic MO[3].

Non-traumatic or pseudomalignant MO can be seen after prolonged immobilization e.g. burns, haemophilia, paraplegia, poliomyelitis, head injuries and prolonged hospitalization in ICU. It is extremely rare in children younger than 10 years. Three cases of traumatic MO have been reported affecting the posterior thigh, the popliteal area and the knee in this age group[3]. All children had unilateral involvement of MO. The most common locations of MO are the large hip muscles. Several other anatomical areas as in the neck, scapula, hand and abdominal muscles have been reported. In infants myositis ossificans circumscripta have been reported in the shoulder and paravertebral muscles[7,8]. They were also affected in one side. Our children were affected mainly in the gluteus muscles and bilaterally. No other cases in preschool children, with no previous generalized illness, were found in the literature with MO after immobilization in the ICU.

The etiologic stimulus for myositis ossificans is not known. In MOT it is hypothesised that an initial trauma to the area

with fibroblastic reaction and subsequent osseous and cartilaginous metaplasia plays a role to a certain degree[9]. Nevertheless, only one third of the cases described in the literature present with a history of trauma[9]. In most of these cases, repetitive minor mechanical injuries, ischemia or inflammation have been implicated as possible causative factors. Our first patient had been complicated with femoral vein thrombosis on the right side when intubated in the ICU, but MO appeared bilaterally. It is difficult to assume that that was the cause of MO but ossification was more extended on the right side.

MO has characteristic clinical features[9]. In the early stages children present with increasing pain and swelling in periarticular location. When affecting the lower limb, difficulty to walk and stand is prominent. Restriction of joint movements is the most prominent clinical sign. It is difficult to be found on clinical examination in children that are intubated in the ICU but become evident when children are improving and return to the hospital ward. Absence of fever or signs of inflammation in the blood tests is in accordance with the diagnosis of MO[3]. One characteristic clinical feature of MO is symptoms' improvement with time: the early inflammatory phase gives the way to maturation phase, with pain resolution usually within 4-6 weeks from symptoms' onset. This clinical feature is also very helpful in differential diagnosis. Ossification due to malignancy or infection is continuing with worsening symptoms and signs.

MO has imaging features that reflect the underlying pathology. Myositis ossificans has three distinct zones: the centre zone that consists of rapidly proliferating fibroblasts with areas of hemorrhage and necrotic muscles, the intermediate or middle zone that is characterized by osteblasts with immature osteoid formation and islands of cartilage due to endochondral ossification and the peripheral zone that is composed of mature bone, usually separated from the surrounding tissue with mixoid fibrous tissue[1-3]. By the 3rd to 4th week calcifications and ossifications appear inside the mass[1]. By the 6th to 8th week a well organized cortical bone with cortex and marrow space formation develops at the periphery. This new bone continues to mature, so that by 6 months a dense ring of compact bone has developed with central core of lamellar bone[1,3]. Our patients on the x-ray and CT scan examination appeared with this smooth well organised bone in the periphery, with the central lycency and intact periosteum, well separated from the femoral bones.

The differential diagnosis must include malignant neoplasm, such as osteosarcoma or Ewing sarcoma and soft tissue sarcoma, rabdomyosarcoma, as well as, various benign conditions such as post-traumatic periostitis, inflammatory arthritis, osteomyelitis, tumoral calcinosis and osteochondroma[1,2]. This is of paramount importance in unilateral cases and require the appropriate investigations. In our children the bilateral symmetrical involvement, with clear radiological signs of MO and absence of suspicious signs, combined with the previous general good health of our children, facilitated the diagnosis of MO.

The combination of different imaging techniques can help for the correct diagnosis[3]. Conventional radiographs should be performed to evaluate the pattern of calcification, and to exclude bone involvement. Extraosseous osteosarcoma or Ewing sarcoma appear with abnormal calcification, asymmetrical, involving different muscle compartments and unilateral. There is more dense irregular ossification in the center of the lesion. Periosteal reaction or cortical erosion is the most suspicious sign in x-ray evaluation.

CT scan is the most accurate examination. There is a smooth pattern of normal ossification of a particular muscle, at the periphery of the joint, with clear separation from the bone. Depending on the stage of ossification, elements of lamellar bone are found in the periphery of the affected muscle, with central radiolucency. CT scan can demonstrate early the typical pattern of ossification when MO is suspected and can confirm the diagnosis. MRI investigations may be confusing in the early stages of MO. There are inhomogenous signal signs because of the different elements of MO. Signs of cellular, cartilage and bony elements are different on MRI, mainly in the early stages of MO. Furthermore, unclear margins of non traumatic MO with surrounding oedema can increase the possibility[3].

Biopsy is performed in doubtful cases to exclude malignancy[1]. It is important to have samples from all the elements of MO. In early stages, if specimen is taken only from the central lesion, the rapidly proliferating mesenchymal cells have increased mitotic activity that may be diagnosed as malignancy. Confirmation of the presence of mature osteoid formation in the periphery and cells with no cellular atypia in the center is essential for diagnosis of MO. In our patients we did not perform biopsy, since we had a confident clinical and radiological diagnosis for a symmetrical lesion of the hip joint. Bone scintigraphy is sensitive in early stages of MO but is not specific for the diagnosis. In our patients it was helpful to exclude other areas of MO.

Because of the self limiting and benign nature of MO, treatment is usually conservative[1]. In most of cases, especially in young children, a spontaneous progressive resorption of the ossified mass can be observed. In cases of a painful mass with nerve compression surgical excision is recommended. If the disease is in the early inflammatory phase, the surgical excision may cause an additional trauma which may favour the recurrence of the myositis. When the diagnosis of MOC is questionable, a reasonable period of observation and complete maturation of the myositis, before considering surgery, is mandatory[1,9]. If the mass does not manifest zoning maturation and shrinkage as would be anticipated for MOC, other diagnosis should be considered and an open biopsy is recommended[9].

Our two cases of MO in preschoolers after prolonged immobilization had typical clinical and radiological findings. The symmetrical bilateral involvement in previously healthy children was important element for the diagnosis. Both were affected in the hip joint that is a common area for MO. Our patients were treated conservatively and had excellent outcome. Pseudomalignant myositis ossificans is a non aggressive condition, compatible with long survival[10].

Conclusions

Myositis ossificans (MO) is a benign disorder of non-neoplastic heterotopic ossification that is usually related to trauma and affects young adults. It is extremely rare in normal preschool children. It can affect muscles in the hip region in children, after prolonged immobilization in ICU for respiratory insufficiency. Symmetrical involvement of the joints, with typical clinical and radiological signs, can confirm the diagnosis. CT scan was the most essential examination. In young children, a spontaneous progressive resorption of the ossified mass can be observed.

Authors' contribution

Victoria Kougias drafted the initial manuscript and approved the final manuscript as submitted. Elpis Hatziagorou followed the cases, conceptualized and designed the study, reviewed and revised the manuscript and approved the final manuscript as submitted. Nikolaos Laliotis diagnosed both of the cases, critically reviewed and revised the manuscript and approved the final manuscript as submitted. Fotis Kyrvasillis followed the cases, reviewed and revised the manuscript and approved the final manuscript as submitted. Vasiliki Georgopoulou reviewed the X-rays and the CT scans, reviewed and revised the manuscript and approved the final manuscript as submitted. John Tsanakas critically reviewed and revised the manuscript and approved the final manuscript as submitted.

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