CASE REPORT

Myositis Ossificans – Two Case Presentations

Shadrick G Lungu

South Hospital, Konkola Copper Mines Plc
Private Bag (c) 2000, Chingola, Zambia

ABSTRACT

Myositis Ossificans [MO], a florid ossification, may occur in muscles and soft tissue. It is also called by many different names. The lesion contains actively proliferating fibroblasts and osteoblasts and early in its development may be confused with a malignant tumor. It commonly affects vigorous young men and more so among athletes. Most often involved are the flexor muscles of the arm and the quadriceps femoris. By time of presentation, ossification is extensive and the benign nature of the lesion is usually evident on radiological studies. This paper presents two cases. While literature shows that MO is common in the flexor muscles of the arm, the hamstrings and quadriceps femoris, it is noted that other muscles of the thigh and around the hip joint can also be affected as demonstrated in the two cases presented here.

Key words: Myositis ossificans [MO], benign lesion, hip area and hip adductus muscles

INTRODUCTION

Florid ossification, often called myositis ossificans, may occur in muscles and other soft tissues[1]; is a pathologic bone formation in soft tissues that do not normally ossify.[2] It is also called heterotopic ossification, ectopic ossification, neurogenic ossifying fibromyopathy, traumatic myositis ossificans.[2,3]

Corresponding Author
Shadrick G Lungu
shadrick.lungu@kcm.co.zm
Tel.: +260-212-350653
Cell: +260 974-999979
Fax: +260-212-351019

Heterotopic ossification (HO) is defined as the process by which trabecular bone forms outside of the skeletal structure, occupying space in soft tissue where it does not normally exist. This misplaced growth occurs between muscle planes and not within the muscle fibers themselves. Furthermore, though the new bone often abuts existing skeletal structure, it does not interfere with the configuration of the periosteum.[4]

It is a rare condition of unknown pathogenesis, with the first reported case dating back to the 1740s.[2] It is a rare non-neoplastic disease causing progressive ossification of soft tissues and a variety of congenital abnormalities of bones.[5] The lesion contains actively proliferating fibroblasts and osteoblasts and early in its development may be confused with a malignant tumor.[1] Fortunately, early lesions are rarely received as surgical specimens.[1]

Myositis ossificans occurs as a result of trauma, either acute or chronic and can also arise near joints in neurological disorders.[6] Young adults and adolescents, predominantly males, are affected most frequently.[5] Myositis ossificans is thought to be transmitted as an autosomal dominant trait with variable expressivity, however most cases are sporadic.[2,5] Vigorous young men who may or may not have had significant trauma are usually affected$_1$. There are both localized form, which is usually posttraumatic, and a widespread syndrome, which occurs in fibrodysplasia ossificans progressive.[2] The former initially presents as a posttraumatic and well circumscribed lesion that frequently complicates hematoma formation of the muscles, particularly of the proximal extremities.[2] It is commonly seen in the

hip musculature of adolescents who are susceptible to sports trauma with contusions.[2] MO is a common condition that occurs among athletes in association with muscle and/or tendon strain or contusion.[7] Causes that have been cited are sports injuries, such as American football, and repetitive occupational trauma, such as in cavalrymen and shoemakers.[6] Connective tissues in voluntary muscles, aponeuroses, tendons, fascia and ligaments are the sites of ectopic bone formation.[5] Most often involved are the flexor muscles of the arm and the quadriceps femoris. Non typically a mass develops rather rapidly[1].

Myositis ossificans includes several different clinicopathologic entities and designations. Although COLEY (1912), NOBLE (1924), GRUCA (1925) and others documented discussions on the classification, the condition can be most relevantly classified as follows according to SAMUELSON and COLEMAN (1976)[7]: 1) myositis ossificans progressive, 2) myositis ossificans traumatic, 3) myositis ossificans associated with neuromuscular and chronic disease, and 4) non traumatic myositis ossificans.[7] The lesions comprise a wide range of histologic features from osteoma-like to osteosarcoma-like appearances.[7] Another event was reported by FINE and STOUT (1956), who described four cases of an osteosarcoma-like benign ossifying tumor of skeletal muscles and para-anal region, employing for the first time the term "pseudomalignant osseous tumor of soft tissues" as an atypical and pseudomalignant form of myositis ossificans. In spite of the literature, the diversity of the histologic features remains.[7]

Any roentgenograms made early may reveal no mineralization, but by the time the lesion is observed clinically, at least some ossification is usually seen[1]. Ultimately, ossification is extensive and the benign nature of the lesion is usually obvious[1]. Sometimes a lesion is located near a bone and on some of the roentgenograms may appear attached to it.[1] The diagnosis of myositis ossificans containing highly cellular areas with islands of osteoid is often difficult.[7]

The paper presents two cases of MO seen at our institution between 2004 and 2009.

Case I

A 33 year-old man presented to our clinic with an eight year history of pain in the right inner thigh. The patient had initially presented to the local clinic where he was put on pain killers and advised to start attending physiotherapy. The physiotherapist noticed that the patient had more than just pain in his thigh.

The pain was present even at rest. Three years after the onset of pain, he noticed that a lump had developed in the painful area. He gave no obvious history of trauma to the area of pain. He also noticed that he was having problems in walking fast or to run.

He worked underground for Konkola Copper Mine Ltd as a driller (using heavy hand held vibratory machinery). His work involved walking long distances both on surface and underground, lifting heavy loads and doing heavy manual work; and he was unable to do the above stated chores effectively because of the said symptoms. He had no other systemic symptoms and no significant or relevant past medical history.

General examination was normal, but the local examination revealed a bony hard lump in the adductus muscles of the right thigh (mid thigh).
Radiographic examination revealed heterotophic bone in the adductus muscle area arising from the right mid-shaft femur (see figure 1).
Blood serum calcium levels were asked for however, no results were made available.
Surgery was done and the ossified muscle was excised.

Histopathology reviewed osteiod and skeletal muscle with no inflammation.

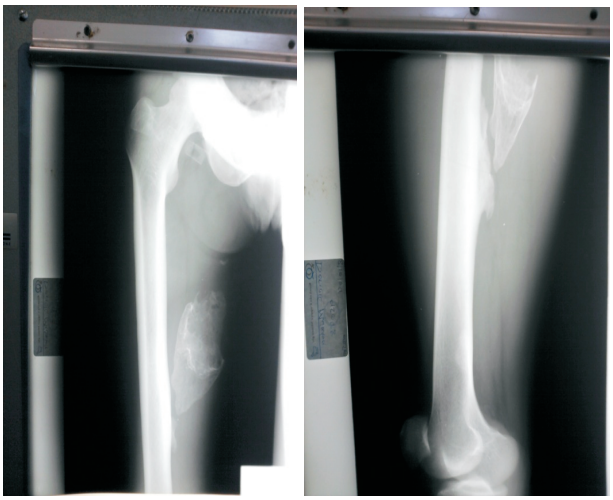

Figure 1. Pre-op AP/Oblique views of the right femur

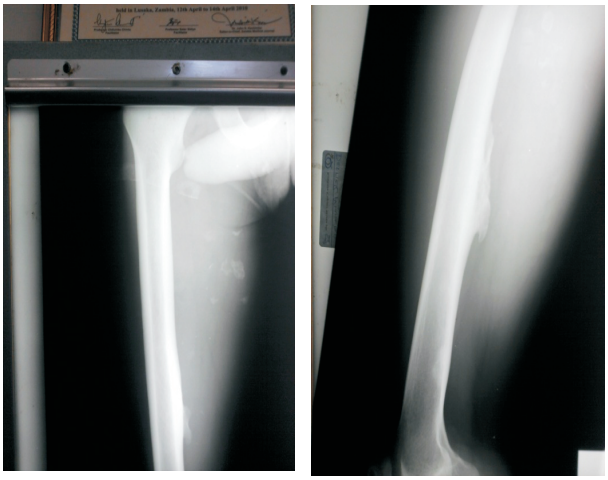

Figure 2. Post op AP / Oblique views with remnants of the ectopic bone

Six years following surgery, the patient has had no recurrence.

Case II

This second patient gave a four months history of pain in the right hip joint if and when he tried to stand and walk. The patient had been referred to the orthopaedic team by the medical team following a protracted treatment of the patient for acute renal failure and the patient had been in coma for over a month in the early part of his medical treatment. The patient did not give any history of trauma. Prior to his medical treatment, however, the patient presented to the hospital in a catatonic state as he was on antipsychotic medication.

Prior to his medical admission, the patient was up and about without the complaints of pain in the hip. The pain in the hip started while he was on the medical ward as he was being rehabilitated (post-coma), to start to stand and walk again.

There were no other systemic symptoms.

On examination, he walked with the torso bent forward, with the help of an elbow crutch on his right side. He was unable to stand erect. General examination did not reveal any abnormality. The musculoskeletal system showed some abnormalities in both the pelvic-femoral areas.

The left hip joint had a jog of movement [internal and external rotation] with the hip joint held in a slightly flexed or near neutral position. Both hips were essentially fixed with no movement at all in the right hip; with a Brooker classification of Class IV[4]. And because of the fixed hips, it was rather difficult to estimate the degree of fixed flexion deformity of either hip. The fixed flexion deformity in the right hip, however, was more severe than the left hip joint. Radiographs were done and a diagnosis of bilateral adductus-muscle myositis ossificans was made. On close examination of the radiographs, it was observed that the myositis ossificans on the right hip involved the muscles just above and below the femoral neck attaching the femur to the pelvis. The hip joints, per se, were essentially radiologically normal (see figure 3).

The patient agreed to surgery [excision on the right side] and, however, was informed that there was a high possibility of recurrence.

Under general anaesthesia, the patient's right hip could not be moved into the neutral position, while the left hip was just able to reach the “neutral” position with an exaggerated lumbar lordosis (see figure 4). This was confirming the fixed flexion

deformity in the right hip. At surgery, in March 2009, accessibility was limited as both the hips could not abduct, flex nor extend. The operation was aborted for the above reason on one hand, and due to large blood vessels in the operation field on the other; and as a consequence of poor accessibility.

The patient continues to mobilize with a walking aid, with the torso bent forward.

Above and below Right Hip Joint

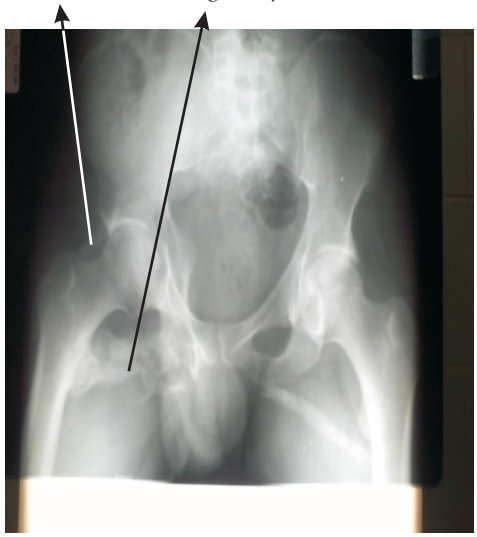

Figure 3. Bilateral "Adductus muscle" MO & right supra trochanteric-pelvic MO

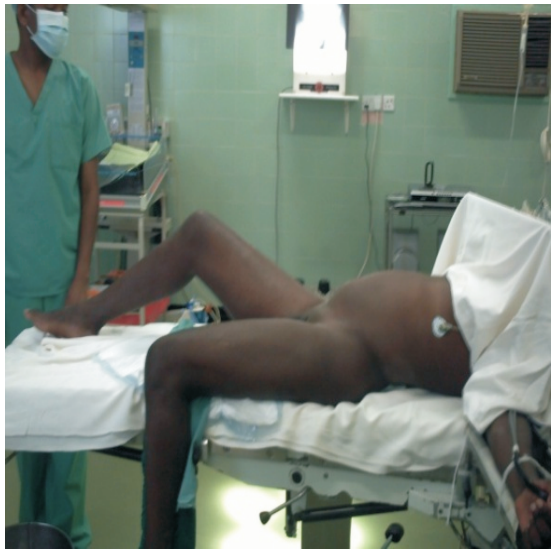

Figure 4. Right hip flexed, with exaggerated lordosis due to left hip "extension"

DISCUSSION

"Myositis ossificans" is a well known benign ossifying process occurring most commonly in the muscle or sometimes in other soft parts.[7] Although myositis ossificans is a relatively rare condition, it is well described with characteristic clinical, radiological and pathological features.[5] The pathogenesis of myositis ossificans is not well understood.[5] This phenomenon of unknown etiology occurs after damage to muscles with subsequent proliferation of connective tissue and differentiation into mature bone.[2] It is assumed that tissue necrosis leads to heterotrophic fibroblastic and vascular proliferation with eventual ossification.[5] Trauma has been associated with the majority of cases, although there are reported cases without an antecedent injury.[5] The most reported risk-factor is re-injury during the early stages of recovery.[2] Causes that have been cited are acute sports injuries, such as in American football, and repetitive occupational trauma, such as in the cavalrymen and shoemakers.[6]

Several clinicopathologic entities have been included in the designation myositis ossificans. They are:

1) myositis ossificans progressive which, occurring in early life, progressively affects all skeletal muscles, and leads to death,
2) myositis ossificans traumatic which follows traumas or surgical operations such as abdominal incision, orthopaedic operation, and dislocation,
3) myositis ossificans associated with neuromuscular and chronic disease such as tetanus, poliomyelitis, paraplegia, and burns, and
4) non traumatic myositis ossificans occurring in those with no definitive causative factor.[7] In the early literature, myositis ossificans traumatic was further divided into two forms; one followed by a single severe trauma and the other representing simple osseous formation appearing in the sites of repeated slight injuries or irritation.[7]

During the major road and railway works in London at the end of the last century, three cemeteries were

excavated and some 3000 bodies exhumed in areas of Farringdon Street, Liverpool Street and Whitechapel. Among the bones from Farringdon Street were three right and two left femurs, all from different individuals, and each having a similar outgrowth of the bone on the shaft.[7] X-rays show an outer layer of dense bone covering a cancellous structure; the typical appearance of myositis ossificans in the femur. [7] The impression is of ossification of the lateral head of the quadratus femoris muscle.[7]

The two patients presented here did not present with any obvious history of trauma. The first case, however, was in a potentially traumatic prone environment and therefore may have had antecedent injury. In a series by Sumiyoshi et al, a history of antecedent trauma was noted in 60 to 70% of the reported cases.[7]

Two types of MO are known, viz-a-viz ossification of organizing hematoma - myositis ossificans circumscripta, myositis ossificans traumatic - and myositis ossificans progressive; also called fibrodysplasia ossificans progressiva (FOP) (International Fibrodysplasia Ossificans Progressiva Association); distinct disease of unknown etiology, usually in young children or young adults, often associated with congenital deformities; relentless progression to entire body (generalized myositis ossificans), no effective treatment.[8]

The organs involved are mostly in skeletal muscle, sometimes subcutaneous fat; most commonly thigh or upper arm and masseter or sternocleidomastoid in head and neck.[3] The most affected are young athletes and the quadriceps and hamstrings are said to be commonly affected in the lower limbs.[3] About 80% of cases of myositis ossificans arise in the large muscles of the extremities, but unusual locations have been described.[5] Ossification of a lesion on the buttock has been reported previously.[5] Andrew et al, however, states that in adolescents and young adults, the thighs and hips are most commonly involved in traumatic myositis ossificans.[2] In 50% of the cases, there is a history of previous injury.[9] The incidence rate is 2% following closed treatment of hip dislocation and increased to 34% when open reduction is required.[2] Other common posttrauma myositis ossificans are the upper arm, calf, and sole of the foot.[2]

In our two patients the neck/head areas were free from this pathology; however the areas of affection were the skeletal muscles around the hip joints, as observed by Oz B et al, and the thigh. The adductor compartment of the thigh was involves in both patients; and the small abductors in the second case presented, as opposed to the quadriceps and the hamstrings as shown in the literature review. These patients were not athletic per se, but young adults.

In the review of literature, the prevalence/incidence is uncommon.[3] Causes and the risk factors are haemorrhage at the injury site and trauma, however, is the precise trigger plus the mechanism which is unknown.[3] While trauma/injury is implicated in many such cases, others present with no history of trauma or injury.[10] The likely risk factors are sports like contact sports (commonly is thighs of rugby players).[3] In the first case presentation there was no history of trauma. Working underground, however, with heavy vibratory machinery/equipment which is sometimes rested on the antero-medial aspect of the thigh in the course of working could be the causal factor despite the patient not giving the actual history of trauma. The second case presentation however did not present any strong evidence to suggest that he could have had suffered any trauma/injury.

In cases of MO secondary to trauma, the pathogenesis is such that the organizing hematoma becomes calcified and then invaded by osteoblasts and in - growth of vascular, ossifying and fibroblastic tissue will eventually produce well circumscribed nodule.[3]

There are various complications associated with this condition such as limited range of motion, pain, contractures, spasticity, and joint impairment with poor rehabilitation results.[3,11]

The patient will normally present with a chief complaint of painful mass in the muscle, pain, and tenderness persisting in the area of large haematoma and usually with a 1-4 week history of trauma.[2] And on general examination, if large, a bony mass is palpable and a differential diagnosis of bone tumour (often misdiagnosed as) osteogenic sarcoma is made.[1,3]

Our two patients did not present with an obvious history of trauma, but pain and a mass in the muscle in the first case. Palpable muscular-bony masses were definite on physical examination. The first

patient had an altered gait, especially when he attempted to walk fast or run, while the second patient was physically unable to walk upright due to the fixed hips. And because of this he was using a walking aid with the support of his spouse. A limitation of the range of joint motion may have serious consequences for the daily functioning of people who are already severely incapacitated because of their original lesion.[11]

Imaging studies are important in these cases. Any roentgenograms made early may reveal no mineralization, but by the time the lesion is observed clinically at least some ossification is usually seen.[1] Ultimately, ossification is extensive and the benign nature of the lesion is obvious.[1] Sometimes a lesion is located near a bone and on some of the roentgenograms may appear attached to it (figure 1 & 3).

Heterotopic ossification is defined as the presence of lamellar bone at locations where bone normally does not exist.[5,11] The condition must be distinguished from metastatic calcifications, which mainly occur in hypercalcaemia, and dystrophic calcifications in tumours.[11] Cytology may offer an accurate preoperative diagnosis of these two entities [proliferative myositis (PM) and MO] ruling out malignancy.[9]

Literature is "silent" about MO affecting the hip joint below and above the joint as well as bilateral affection of the adductus muscles of the same individual patient as the case was in our second patient.

Patients without any reports of pain or decreased mobility maybe better off avoiding the morbidity associated with excision.[2] Excision is only indicated if the lesion is completely ossified because removal of immature bone may cause extensive local recurrences.[2] Some studies have suggested that using prophylactic indomethacin and etidronate [ethane 1 – hydroxyl – 1, 1 – bisphosphoric acid or EHDP] can be beneficial in reducing postsurgical ectopic calcification.[2] The use of biphosphonates has been bolstered by recent case reports that point to its effectiveness.[2] It is, however, hard to assess the precise long-term effects of EHDP because of the variable clinical expression.[5] It is also possible that EHDP is more effective at the initial period of treatment, with the gradual decline in its effectiveness later on. This is supported by the clinical improvement after short-term treatment reported by Bruni et al and Rogers et al.[5]

In spite of their alarming clinical presentation, both PM and MO have excellent prognosis and are completely cured with local excision.[9] The first case presented above underwent surgery as the "tumour" was slowing him down and interfering with his core occupational engagement. The medical treatment was not instituted following surgery and six years following surgery, the first patient remained symptom free. The operation for the second patient was aborted as indicated above.

CONCLUSION

A good history and physical examination are important to arrive at the diagnosis of MO; however, the radiological and histopathological studies will help to confirm the diagnosis and rule out malignant bone disease.

While MO, a benign lesion, is known to affect the flexors of the arm, the hamstrings and quadriceps femoris, it must be noted that other muscles in the thigh, such as the adductors and small abductors muscle of the hip, can also be affected as shown in the above two case presentations.

ACKNOWLEDGEMENT

I wish to thank Dr Duncan D Mugala, Senior lecturer Copperbelt University [CBU] School of Medicine, Head of Department Clinical Sciences; Visiting Consultant KCM Hospitals; Honorary Lecturer University of Zambia – School of Medicine [UNZA SOM] for having accepted not only to proof read this document, but also gave valuable suggestions towards this paper before its submission for publication.

REFERENCE

1. Pitcock, J. A. Tumours and Tumour like lesions of Somatic Tissue Crenshaw A. H, editor: *Campbell's Operative Orthopaedics,* 1971; vol.1, Saint Louis C .V. Mosby Co. Fifth edition pg 1366.

2. Andrew, E. Hendifar, Dorothy, J. Daniel, G. A. Myositis Ossificans: A Case Report Arthritis & Rheumatism. *Arthritis Care & Research* 2005 Oct; vol. 53 No. 5 pp 793 – 795.
3. Myositis Ossificans - *DynaMed [Internet]*. Ipswich (MA): EBSCO Publishing. 1995 - . Record No. 114671, Myositis ossificans; [updated 2010 Jun 09; cited 2010 Oct 11]; [about 4 screens].Available from http://search.ebscohost.com/login.aspx?direct=true&site=dynamed&id=AN+114671. Registration and login required.
4. Baird, O. E. Kang, K. Q. Prophylaxis of heterotropic ossification – an update review. *Journal of Orthopaedic Surgery and Research* 2009; 4:12.
5. Oz, B. B. Boneh, A. Myositis ossificans progressive: a 10-year follow-up on a patient treated with etidronate disodium. *Acta Paediatr* 1994; 83: 1332 – 4.
6. Mann, E. G. Short Report Myositis Ossificans in Medieval London. *International Journal of Osteoarchaeology* 1993; 3: 223 – 226.
7. Sumiyoshi, K. Tsuneyoshi, M. Enjoji, M. MYOSITIS OSSIFICANS A Clinicopathologic Study of 21 Cases. Acta Pathol. Jpn 1985; 35 (5) : 1109 – 1122.
8. Miller, A.E. Davis, B.A. Beckley, O.A. Bilateral and recurrent myositis ossificans in an athlete: a case report and review of treatment options. *Arch Phys Med Rehabil.* 2006; 87(2):286 - 90 (D).
9. Klapsinou, E. Despoina, P. Dimitra, D. Cytologic Findings and Potential Pitfalls in Proliferative Myositis and Myositis Ossificans Diagnosed by Fine Needle Aspiration Cytology: Report of Four Cases and Review of the Literature. *Diagnostic Cytopathology* 2010; 00: 00
10. Meng, S.Q. Sun, X.Q. Gong, L.H. Huang, X.Y. Xi, Y, Myositis ossificans: a clinicopathologic analysis of 15 cases *Zhonghua Bing Li Xue Za Zhi* 2008; 37(10):665 - 9. [Article in Chinese].
11. Vanden, B.L. Vanderstraeten, G. Heterotopic ossification: a review. *J Rehabil Med.* 2005; 37(3):129 - 36.