CASE REPORT

HETEROTOPIC OSSIFICATION OF HIP IN A RARE CASE OF MOYAMOYA DISEASE: A RARE CASE REPORT

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HOW TO CITE THIS ARTICLE:
Anita A. M, Dimple Mehrotra, Pratima S, Ravi Palankar. "Heterotopic Ossification of Hip in A Rare Case of Moyamoya Disease: A Rare Case Report". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 17, April 27, 2015; Page: 2620-2623.

ABSTRACT: A case of extensive ossification around the left hip joint involving lesser trochanter of the femur leading to ankylosis of left hip joint in a 60 years male is being reported. The diagnosis of moyamoya disease was made, which is a rare form of occlusive cerebrovascular disorder. Occlusion of an artery may present with Transient Ischemic Attacks, headaches, stroke and seizures. Surgical management is the only option in these cases. This 60 years male had an acute onset hemiplegia 4½ years back and at present came with complaints of pain and swelling over the left hip. Surgical excision was done and histopathological examination revealed extensive ossification of skeletal muscle. A diagnosis of Moyamoya disease complicated with heterotopic ossification was made. There was no recurrence after 8 months of follow up.
KEYWORDS: Heterotopic ossification, Hip, Moyamoya disease.

INTRODUCTION: Moyamoya disease is an uncommon cerebrovascular condition characterized by progressive stenosis of the bilateral internal carotid arteries with compensatory formation of an abnormal network of perforating blood vessels providing collateral circulation. Familial moyamoya disease has been noted in 15% of patients, indicating an AD inheritance pattern with incomplete penetrance.[1] It may cause heterotopic ossification around the joints, most commonly the hip joint. Heterotopic ossification associated with neurologic injury, tends to form at major synovial joints surrounded by spastic muscles. It is commonly associated with traumatic brain or spinal cord injury and with other causes of upper motor neuron lesions. Heterotopic ossification can result in nerve impingement, joint ankylosis, complex regional pain syndrome, osteoporosis and soft-tissue infection. The associated decline in range of motion may greatly limit daily activities.[2]

CASE REPORT: 60 years male presented with pain and swelling in the left hip, since 4 years. It had an insidious onset and had been slowly growing for the past 4 years. Patient noticed the swelling 1 year after he had right sided hemiplegia and seizures. Angiography revealed cerebral vein and dural sinus thrombosis and cerebral vascular thrombi 5 years back. Patient was a known case of hypertension on irregular treatment. Surgery was carried out to clear the thrombus. 6 months later, patient complained of pain over the left hip. He was also diagnosed to have deep vein thrombosis in the left lower limb. Radiotherapy was advised along with physiotherapy. He had been taking irregular treatment for the same. On examination there was tenderness all over the left hip joint particularly over greater trochanter. The swelling was 8x4x3cms in size and bony hard in consistency, (Figure I). The limb was externally rotated with restricted movements of internal rotation and abduction. Gait was antalgic and the patient was unable to sit or walk. CT

Pelvis revealed extensive ossification around the left hip joint with extension leading to ankylosis of left hip joint, Impression- ?Heterotopic ossification. Surgical excision of the mass and correction of the limb was done.

Multiple bits of grey brown tissue largest measuring 4x4x3 cms in size and firm to bony hard in consistency were received for Histopathological Reporting in our department, (Figure II). Microscopy showed extensive ossification, normal skeletal muscle fibers and focal areas of necrosis with normal bony trabeculae, (Figure III). Correlating with clinical data, a diagnosis of Moyamoya disease was considered. Follow up CT at 3 months postoperative visit showed no recurrence. CT Brain revealed gliosis and age related atrophic changes. Follow up visit at 6 months also showed no recurrence and the patient was able to sit, walk and had no pain.

DISCUSSION: Moyamoya disease was first described in 1957 as a "bilateral hypoplasia of internal carotid arteries".[3] The name of the disease comes from Japanese and means 'puff of smoke'. High incidence of the disease is noted in Asia (esp. Korea and Japan), with 3 cases per 100,000 of pediatric population. Moyamoya disease has several unique clinical features, which include two peaks of age distribution at 5 years and at about 40 years.[4]

Currently, the major proteins believed to play an active role in the pathogenesis include vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), hepatocyte growth factor (HGF), transforming growth factor-β_1 (TGFβ_1), and granulocyte colony-stimulating factor (G-CSF). In terms of the genetics, recent literature suggests a low penetrance autosomal dominant or polygenic mode of transmission involving chromosomes 3, 6, 8, 12, and 17 for familial Moyamoya Disease.[5] The most common clinical symptoms of the disease are sudden onset of hemiplegia, with sensation disturbances and aphasia. Other clinical features are headaches, vertigo, seizures, involuntary movements, cognitive dysfunction and psychoorganic syndrome.

According to the classification of Japanese Health Ministry, there are 4 clinical forms of Moyamoya disease: ischaemic, haemorrhagic, epileptic and 'other'. The ischaemic form is most common in children, while the haemorrhagic form is more popular in adults. In children, moyamoya disease manifests as transient ischaemic attacks or lacunar strokes, leading to mental retardation, whereas in adults, as intracranial haemorrhages, including subarachnoid haemorrhages. The risk of repeated ischaemic strokes is from 65% to 82% (for bilateral lesions). In patients from Asian countries, on the other hand, repeated haemorrhagic strokes are quite common. Modern methods of brain vessel imaging like CT angiography, MR angiography or 3D-DSA (rotational digital subtraction angiography) lead to a more frequent diagnosis of asymptomatic cases of this disease.[6]

A rare complication of this rare disorder is heterotopic ossification (HO) which occurs around joints causing spastic paralysis, most commonly involving the hip joint, knees, elbows and shoulders. The incidence of neurogenic HO varies from 11-40%. It appears only within the area of neurological deficit. In vivo molecular imaging and confirmatory ex vivo tissue analyses of an established murine animal model of BMP-induced HO has shown that matrix metalloproteinase-9 (MMP-9) can be detected as an early-stage biomarker before mineralization. Bone scan is the most sensitive imaging modality for early detection and assessing the maturity of HO.[7]

Heterotopic ossification or de novo bone formation involves recruitment and expansion of chondrosseous progenitors capable of undergoing bone formation in nonskeletal tissues. Bone morphogenic protein-2 (BMP-2) initiates inflammation by release of neuroinflammatory factors, substance P and calcitonin gene related peptide (CGRP) from sensory nerves. These factors, in turn, recruit immune cells including mast cells, platelets and neutrophils. Mast cell degranulation releases various proteases and activates matrix metalloproteinases that aid in remodeling the local peripheral nerves. This remodeling process enables not only the expansion and release of progenitors from the nerve, but also other accessory cells essential for patterning and coordinating the bone formation process, thus ensuring the vascularization and innervations of newly formed bone.[8]

CONCLUSION: Moyamoya disease is an uncommon cerebrovascular disease, with a high incidence in East Asia and therefore has been discussed in this review to elaborate its etiology, pathophysiology, manifestations and complications.

FIG. 1: Clinical photograph of the present case showing the swelling

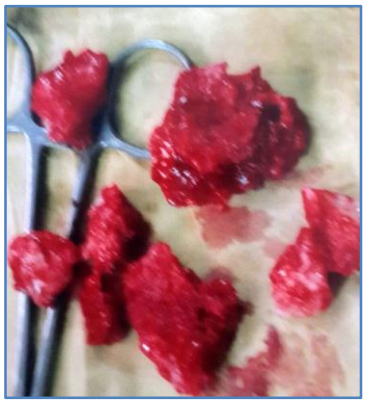

FIG. 2: Postoperative picture showing the excised bony masses which were sent for histopathology

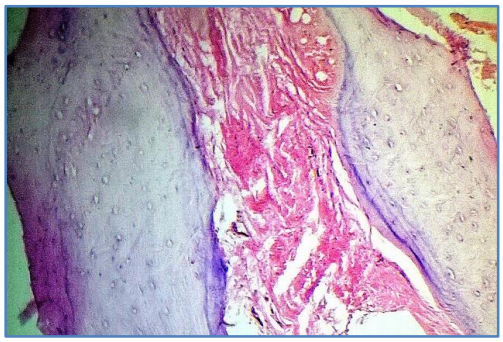

FIG. 3: H & E stained section (400X) shows bony trabeculae and skeletal muscle tissue

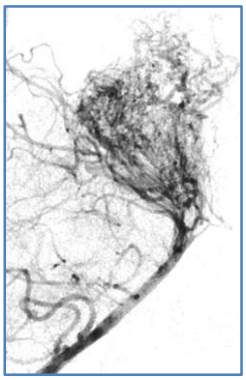

Fig. 4: Coronary Angiography in Moyamoya disease with the characteristic puff of smoke appearance

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Date of Submission: 08/04/2015.
Date of Peer Review: 09/04/2015.
Date of Acceptance: 11/04/2015.
Date of Publishing: 27/04/2015.