Case Report

Surgical excision of heterotopic ossification of hip in a rare case of Moyamoya disease with extra articular ankylosis

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Abstract

We report a case of isolated ossification of iliopsoas with ankylosis of the left hip in a 27-year-old female. The patient was diagnosed to have Moyamoya disease, a rare chronic occlusive disorder of cerebrovascular circulation following an acute onset of hemiplegia. The patient presented 9 months later to us with ankylosis of left hip which was successfully treated by surgical excision of the heterotopic bone and there was no recurrence at the end of 5 years. A review of literature failed to reveal a similar case with isolated and complete ossification of iliopsoas muscle associated with Moyamoya disease which required surgical intervention. Surgical excision resulted in dramatic improvement in the quality of life. Surgical excision of neurogenic type of heterotopic ossification is a very successful procedure and timely intervention after maturity of mass is very important to prevent the onset of secondary complications and to avoid recurrence.

Key words: Isolated iliopsoas ossification, Moyamoya disease, neurogenic heterotopic ossification, surgical excision

Introduction

Heterotopic ossification (HO) occurs around joints in many pathological conditions of the nervous system causing spastic paralysis, with the hip being most often involved.[1] We report a case of ankylosis of the left hip due to HO in a 27-year-old female with Moyamoya disease,[2-4] a rare disease causing stenosis of internal carotid arterial system of the brain. This is a rare complication in isolation, but good outcome can be achieved by a well performed surgery. We report the perioperative considerations and the long term followup of neurogenic HO and also provide an insight about this rare condition called Moyamoya disease.

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Case Report

A 27-year-old girl presented to us with ankylosis of the left hip following an acute onset of left-sided hemiplegia 9 months back. She had an angiogram of the cerebral blood vessels and she was diagnosed to have Moyamoya disease [Figure 1a]. She had an indirect bypass procedure called encephalo-duro-arterio-myo-synangiosis which involves transposition of a segment of a scalp artery to supply the surface of the brain to improve collateral blood flow. The patient gradually recovered motor power but had difficulty in walking and sitting with mild spasticity of the left upper and lower limbs. On presentation, her left hip was ankylosed in 20° of external rotation, neutral abduction, and 10° flexion. Ectopic bone was palpable anterior to the left hip. There was no local warmth. Her alkaline phosphatase level was normal and radiograph showed HO in the iliopsoas muscles, extending from the ilium to the lesser trochanter with cortical delineation implying maturity [Figure 1b]. Hip joint appeared normal. 3D CT scan of the hip and pelvis demonstrated that the mass was extra articular, lying anterior to the hip and also its proximal and distal extension [Figure 1c].

Surgical excision was planned to restore hip joint motion as the mass was fully mature and the patient had good neurological recovery. The heterotopic bone was exposed through the anterior iliofemoral approach. The femoral neurovascular bundle was situated just medial to the mass

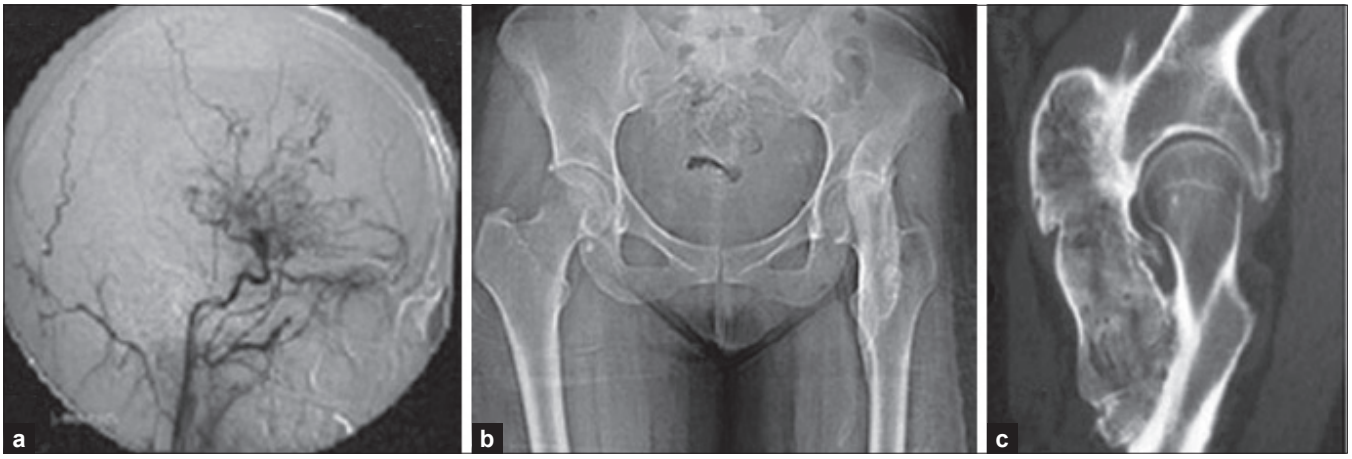

Figure 1: (a) Cerebral angiography showing vascular abnormality in Moyamoya disease with intracranial vascular stenosis of the circle of Willis (b) X-ray showing the heterotopic ossification in the left hip. The cortex is well delineated implying full maturity. The hip joint space is well maintained (c) Sagittal section in CT showing the mass anterior to the hip outside the capsule but contiguous with the anterior wall of acetabulum

and there was a risk of injury to the femoral nerve, femoral artery, or the profunda femoris artery. Dissection was done subperiosteally with careful medial dissection and minimal medial retraction. The mass was exposed from the level of lesser trochanter inferiorly to the level of the anterior inferior iliac spine superiorly. But it was extending into the iliac fossa along the iliopsoas muscle plane [Figure 2a]. Anterior wedge resection was done with an osteotome after making multiple drill holes proximally and distally, and the hip joint capsule could be identified by gently rotating the hip [Figure 2b]. Usually the capsule is uninvolved and there is a layer of tissue separating the hip joint from the mass [Figure 2c]. A part of anterior wall of acetabulum was broken during the excision of the proximal part of the mass and this was fixed with a cancellous screw [Figure 3a]. After excision of the heterotopic bone, the hip moved easily from 0° to 90° of flexion. The wound was closed over large suction drain.

Postoperatively, the patient was mobilized nonweight bearing initially with the help of a walking aid because of the acetabular wall fracture and was on above knee skin traction as there was mild spasticity of the muscles and a detachable derotation boot for 4 weeks to prevent external rotation. She was allowed to sit up and the hip was gently mobilized with continuous passive motion machine to prevent stiffness of the hip. She was prescribed oral indomethacin 25 mg TID for 3 months. At 5 years followup, the patient is now able to drive a two wheeler, sit on the floor, and there is no evidence of recurrence of the HO [Figure 3b]. The flexion in the hip is from 0° to 110° [Figure 1a].

Discussion

Neurogenic HO occurs after neurogenic injuries such as spinal cord and central nervous system injury, burns, head injuries, strokes, and brain tumors.[5] The pathophysiology of HO is unknown. It is due to the inappropriate differentiation of mesenchymal cells into osteoblastic stem cells in response to unidentified inducing factors such as the pool of available calcium in adjacent skeleton, soft tissue edema, vascular stasis, tissue hypoxia, and mesenchymal cells with osteoblastic activity.[6-8] In patients with head injury changes in the levels of circulating catecholamines and sympathetic activity,[9,10] the high levels of calcitonin may well be related to the more rapid healing of fractures seen in this group and HO formation.[11] Bone morphogenetic proteins (BMPs) play a role as a paracrine factor in the differentiation of satellite cells into bone forming cells.[8,11] Conventional histology, histochemistry, immunohistochemistry, electron microscopy, and molecular biology studies of HO reveal that it is a reactive, proliferative, and reversible neoplasia in chronically damaged soft tissue.[12]

The incidence of neurogenic HO varies from 11 to 40%.[1,13,14] HO appears only within the area of neurological deficit.[14] It is found neither below the knees nor above the level of paralysis.[14] Hips are most often involved, while the knees, elbows, and shoulders are less frequently affected.[1,14] Currently, there are no methods to detect HO before mineralization occurs. *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.[15] Bone scan is the most sensitive imaging modality for early detection and assessing the maturity of HO.[16] Nonsurgical treatment with indomethacin and radiation therapy is appropriate for prophylaxis or early treatment of HO.[17] As HO becomes mature, there also is a significant decrease in uptake in the bone scan, often reaching a normal level, in both flow study and blood-pool activity.[16] Anatomically, HO occurs outside the joint capsule without disrupting it. The new bone can be

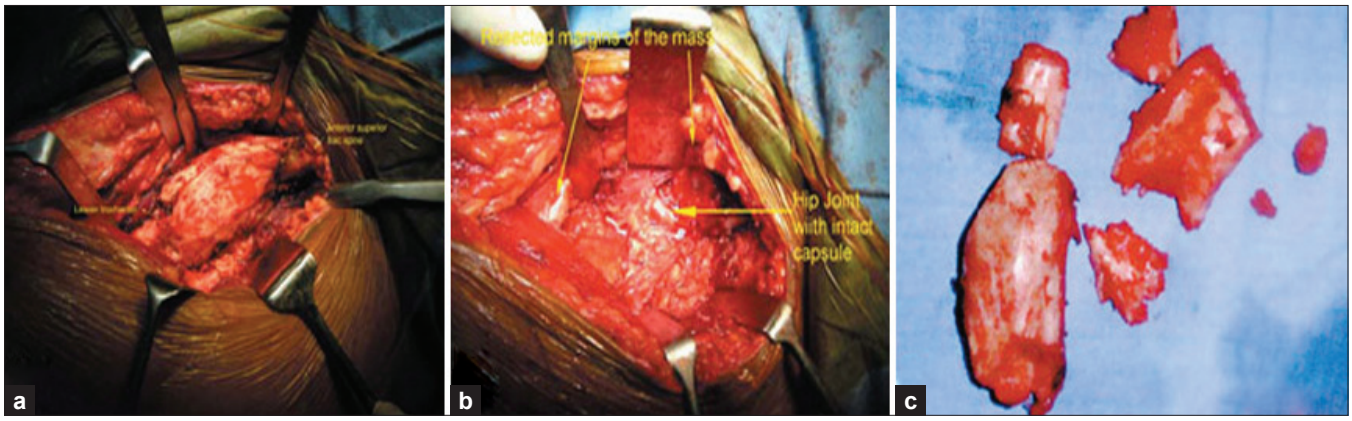

Figure 2: (a) Clinical peroperative photograph showing heterotopic mass (b) hip joint after excision of the mass (c) excised mass of bone

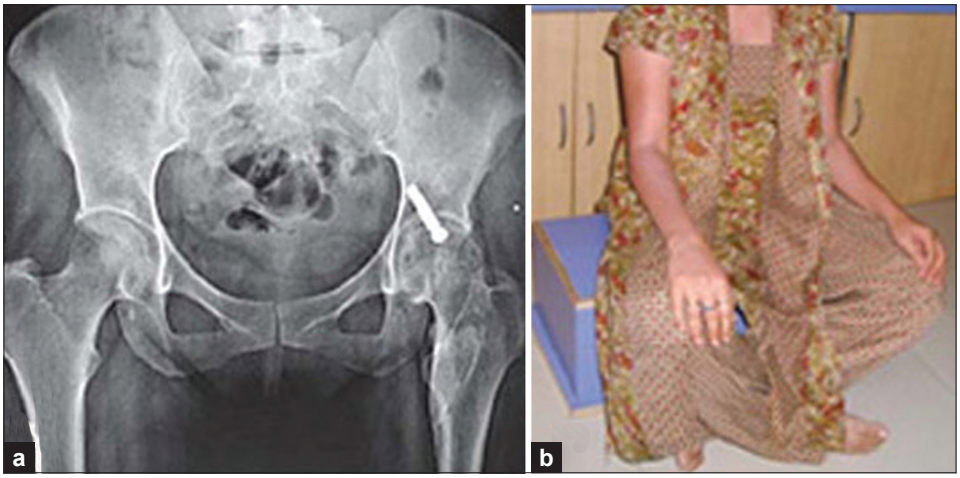

Figure 3: (a) Postoperative X-ray showing no recurrence after 5 years. Small fracture of anterior wall of acetabulum was fixed with a cancellous screw (b) Clinical photograph showing functional outcome

contiguous with the skeleton but generally does not involve the periosteum.[18] Alkaline phosphatase levels have been reported to parallel the activity of ossification.[16] However, alkaline phosphatase levels cannot be used to draw clinical conclusions about maturity or recurrence of HO.[14,16,19,20]

Surgical indications for excision of HO include improvement of function, standing posture, sitting or ambulation, independent dressing, feeding, and hygiene, and prevention of repeated pressure sores from underlying bone mass. The optimal timing of surgery has been suggested to be a delay of 12–18 months[21] until there is radiographic evidence of HO maturation and maximal recovery after neurological injury. The ideal candidate for surgical treatment before 18 months should have no joint pain or swelling, a normal alkaline phosphatase level, and 3-phase bone scan indicating mature HO.[17] In the hip, it is necessary to excise the bony bridge lying between the lesser trochanter and the anterior inferior iliac spine.[1,18] Surgical excision is technically demanding due to close proximity to the neurovascular bundles. Other treatment options include osteotomy, resection of femoral head and neck, or disarticulation of the limb which is indicated in patients with head injury with severe cognitive and physical deficits to improve personal hygiene and posture.[14] In patients with severe cognitive dysfunction with involvement of multiple joints and early surgery in patients with immature bone have a higher chance of recurrence.[19-22]

In cases of prophylaxis against recurrent HO following excision after total hip replacement, a combination of radiotherapy and indomethacin is effective.[23] The efficacy and dose requirement of radiation therapy to prevent HO of nonsurgical origin needs further studies. Seven hundred centigray dose of radiation therapy does not effectively prevent the recurrence of neurogenic HO in high-risk patients.[24] Unnecessary delay in surgery should be avoided as results are poor after intraarticular changes take place and there are increased chances of intraoperative fractures.[25]

HO is common in acquired nervous diseases, but it is one of the rare[3] complications of the neurological deficits

associated with Moyamoya disease, a rare genetic disorder. The progressive intracranial vascular stenosis of the circle of Willis[4] is followed by the development of collateral vessels which are small, weak, and prone to hemorrhage, aneurysm, and thrombosis [Figure 1a]. On carotid angiography, these collateral vessels have the appearance of a "puff of cigarette smoke" (Moyamoya in Japanese). The disease primarily affects children and the first symptom is often stroke or recurrent transient ischemia. Treatment mainly involves a direct or indirect bypass of the external carotid circulation to supply the areas of insufficient blood supply to the brain.

There is only one case of HO reported in a case of Moyamoya disease around the hemiparetic shoulder and hip which was subsequently treated conservatively.[3] A case of bilateral ankylosis of the hip in a 3-year-old child due to isolated ossification following a long period of coma was reported in 1992.[26] In our patient, heterotopic bone affected only the iliopsoas muscle, causing ankylosis. Complications of surgical removal of HO include hemorrhage, wound-healing problems, cellulitis, infection or osteomyelitis, and possible recurrence of HO.

Surgical excision of neurogenic type of HO is a very successful procedure and timely intervention after maturity of mass is very important to prevent the onset of secondary complications and to avoid recurrence. In our patient, the vascular occlusion was corrected by a bypass procedure[4,27] and she had minimal cognitive dysfunction like dysarthria. As she had single hip involvement and good neurological recovery, excision of the mass resulted in marked improvement in function without recurrence in the long term. Our patient was able to walk, sit, and drive within few months after excision. Surgical excision resulted in dramatic improvement in the quality of life.

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