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Intramedullary Osteosclerosis of the Tibial Midshaft in a 38-year-old female: A case report with Review of Literature

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Abstract

Intramedullary osteosclerosis of the tibia is a lesser-known entity with only a few cases reported in literature. We present a case of bilateral diaphyseal intramedullary osteosclerosis in a 38- year old female presenting with vague pain affecting both the legs, aggravated by walking, worse at night. Radiological workup revealed thickening of the cortex at the midshaft region with no periosteal reaction or abnormal soft tissue shadow. Histopathological findings were suggestive of osteosclerosis with normal trabeculae and features of osteonecrosis. The patient was treated with excisional biopsy of the lesion resulting in curing of her symptoms. Intramedullary osteosclerosis is an important differential diagnosis in cases of intramedullary sclerosis of long bone.

Keywords: Intramedullary Osteosclerosis, tibia, excisional biopsy

Background

Intramedullary osteosclerosis of the tibia is a lesser-known entity with only a few cases reported in literature. [1, 2] Although the clinical, radiological and histopathological features are well recognized, the treatment of such patients remains unclear.

We present an uncommon case of osteosclerosis of bilateral tibia presenting as vague pain in both the legs, confirmed by ruling out other causes clinically, radiologically and on biopsy. The patient was treated surgically by excision of the sclerotic bone segment, which relieved the patient off her symptoms.

Case History:

A 38-year-old female presented to our hospital with complains of pain over the left leg of 8 months duration. The pain aggravated on exertion and at night and was not relieved by analgesic use. The patient gave no history of trauma, running or other athletic activities, fever, weight loss, back and groin pain or any other co-morbid conditions.

Local examination revealed tenderness over the shin bilaterally with no palpable swelling or skin changes. Hip and spine examination were within normal limits. Peripheral pulsations were well felt and there was no neural deficit.

Laboratory findings were as follows: Hemoglobin-10.7 g/dl, TLC – 8.2 x 103 / µl (N 64%, L 28.9%, M 6.4%, E 0.5%, B 0.2%), C-reactive protein – 1.36 mg/L, Erythrocyte sedimentation rate – 33 mm/hr, Serum blood Glucose – 102 gm/dl, Serum Calcium, Serum. PO4, Serum Alkaline Phosphate, Serum. Vitamin D3 and Serum Parathyroid were all within normal limits.

Radiographs (Fig. 1) showed cortical thickening at the mid-diaphyseal region of both the legs, left more than right with no periosteal reaction. There was no visible fracture, osteolysis or soft tissue shadow. CT scan (Fig. 2) and MRI (Fig. 3) imaging on a 1.5 T machine revealed cortical thickening and near complete obliteration of marrow cavity at the mid-diaphyseal level of the tibia. An open biopsy was performed on the left leg and approximately 1.5 x1.5 cms bone block removed from medial surface of the tibia and sent for biopsy; which was suggestive of osteonecrosis. The patient had 90% relief of symptoms following biopsy and was started on analgesics.

On 8 months follow-up, the patient continued to have about 90% pain relief in the left leg, however had now developed symptoms on the opposite side. An excisional biopsy was planned on the right tibia. Intra-operatively, the site showed sclerosis of the cortex with complete obliteration of the medullary canal. A cortical window was made and excision of the sclerotic segment was performed. The continuity of the medullary canal was restored by drilling proximally and distally. (Fig.1&4) Histopathological findings revealed normal trabecular pattern with features of osteonecrosis. The patient had no

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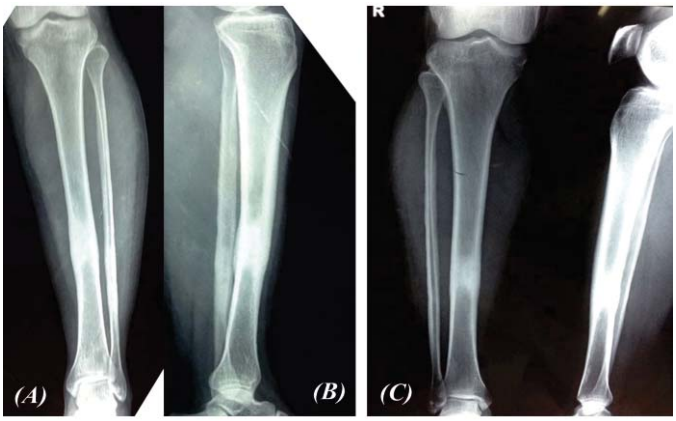

Figure 1: (A) Frontal and (B) lateral radiographs of the left and (C) frontal and lateral radiographs of the right tibia demonstrating bilateral cortical thickening and endosteal sclerosis. No periosteal reaction is seen.

operative complications and the wounds healed well. Complete relief of symptoms was noted in the right leg within 48 hours of surgery. The patient continued to have mild pain in the left leg, however it did not restrict her activities.

Discussion:

Intramedullary sclerosis is an idiopathic uncommon condition associated with new bone formation commonly affecting the shaft of the tibia in middle aged women It presents with vague pain in the legs, which increases on exertion and is not relieved on analgesic use. The pain worsens with disease progression. Examination findings usually reveal soft tissue edema and bony tenderness on deep palpation. [1,2,3]

Radiographs reveal selective endosteal diaphyseal hyperostosis, which varies in length and severity from minimal to complete obliteration of the medullary cavity, and may be associated with expansion of the bone. The sclerotic changes are strictly located in the diaphysis of long bones and when the disease is bilateral there is a tendency for asymmetry. Neither a periosteal reaction nor soft-tissue abnormalities are noticed. High resolution computed tomography confirms the medullary sclerosis and in addition is able to show any cortical irregularities.[1, 3]

MRI typically shows the sclerotic areas with low signal intensity. No or minimal increase in signal intensity is noted in fat suppressed T2 and contrast enhanced T1 images, however mild enhancement may be present in the medullary cavity and adjacent soft tissues respectively, probably as a reaction process.[4, 5]

The differential diagnosis of the condition includes Sclerosing disorders of the bones viz. Tumors (osteoid osteoma, metastasis), infections (chronic osteomyelitis), healing stress fractures, metabolic and endocrinal causes like hypervitaminosis A, Renal Osteodystrophy, pseudohypoparathyroidism. [3, 4]

Other causes include Sclerosing bone dysplasias like Osteopetrosis, Pyknodysostosis, enostosis, osteopoikilosis, osteopathia striata, melorheostosis, metaphyseal dysplasia, hyperostosis corticalis generalisata, Worth Disease (Autosomal Dominant osteosclerosis), Camurati- Engelmann disease, Ribbing Disease, Sclerosteosis, metaphyseal dysplasia.

The lack of periosteal reaction, soft tissue involvement and bilateral affections are features of intramedullary osteosclerosis which are characteristically different from a malignant tumours like an osteogenic sarcoma, lymphoma or osteoblastic metastasis.

Osteoid osteoma may be ruled out by the lack of a classical history and of a radiolucent nidus.

Chronic recurrent multifocal sclerosing osteomyelitis is more common between adolescent and children and commonly affect the metaphyseal region.

The lack of a fracture line and presence of intramedullary sclerosis rule out a stress fracture [5].

Generalised osteosclerosis are features of Renal osteodystrophy and pseudohypoparathyroidism which can be ruled out with the help of laboratory findings. Hypervitaminosis A shows classical features of a periosteal new bone formation with sparing of the medullary canal. Sclerosing dysplasias can be differentiated from Intramedullary osteosclerosis on radiological findings. Paget's diease may mimic Intramedullary Osteosclerosis radiologically, however can be differentiated from it on the basis of normal Alkaline Phosphatase levels. Ribbings disease and Camurati – Engelmann disease are almost identical

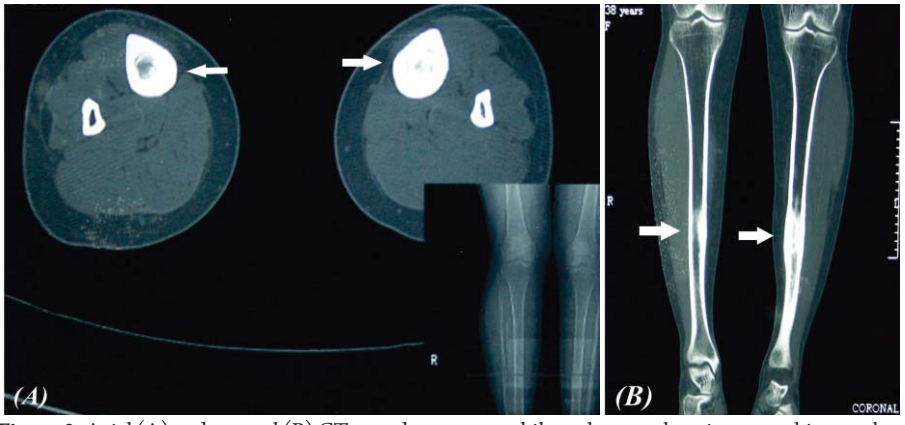

Figure 2: Axial (A) and coronal (B) CT scan demonstrates bilateral osteosclerosis encroaching on the medullary cavity with cortical thickening. However, no periosteal reaction is seen.

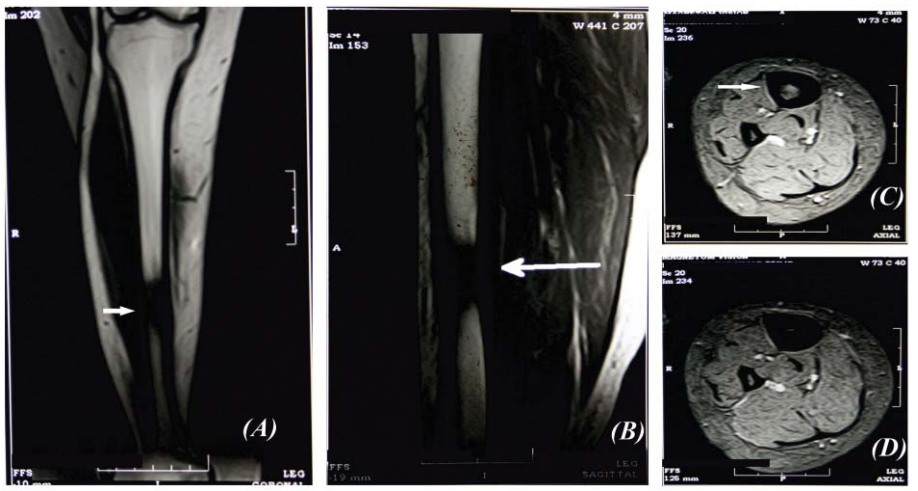

Figure 3: A-D. MRI images of the right leg (A) Coronal section and (B) Sagittal section showing cortical thickening of the diaphysis. An axial T2 weighted sequence (C) and Gradient echo sequence (D) images show diffuse cortical thickening and narrowing of medullary cavity in the diaphyseal region of the tibia.

to intramedullary osteosclerosis on radiology and require clinical, laboratory and histological analysis for differentiation. [1, 6, 7] The differentiating features of Camurati – Engelmann disease include Autosomal Dominance disease presenting in the first decade, manifests as a bilateral disorder causing pain, progressive leg weakness, elongated extremities, and gait disturbances. [8] In contrast, Ribbing's Disease is an autosomal recessive disorder seen after puberty with no sex predominance, and may be either unilateral or bilateral. Intramedullary Osteosclerosis is non-heriditary and commonly affects women. [9]

Radiological features of Camurati – Engelmann disease include bilateral fusiform thickening of the cortex of long tubular bones. In addition, intramembranous bones like skull may be involved resulting in anaemia. On the other hand, Ribbings disease shows similar features however, involves only the long bones. [10] Intramedullary osteosclerosis also presents with similar radiological findings, and is essentially a diagnosis of exclusion. [2]

Histological findings of Camurati – Engelmann disease include osteoblastic and osteclastic activity indicating both bone formation and resorption, whereas Ribbings disease and Intramedullary osteosclerosis demonstrate only osteoblastic activity.[4] The non-hereditary nature along with female predominance helps differentiate Intramedullary Osteosclerosis from the two conditions.

We treated our patient by making a cortical window on the medial cortex of the tibia and restoring the medullary canal by drilling holes through the endosteal surface of the sclerotic segment with the presumption that decompression resulted in pain relief and that obliteration of the medullary canal caused recurrence of the pain as reported previously. When the patient returned to our clinic after a month, her pain had subsided. The 8 – month follow-up radiograph of the left leg showed a persistent cortical thickening in the tibia and an obliterated medullary canal. However, her symptoms had improved remarkably. Many authors [8, 10, 11- 16] have advocated intramedullary decompression by reaming, curettage, or by making a window at the lesion for pain relief based on the belief that bone marrow edema causes pain. Medullary decompression appears to improve symptomatology and can be used to treat patients not responding to the medical line of management. However, it does not correct the primary cause of disease.

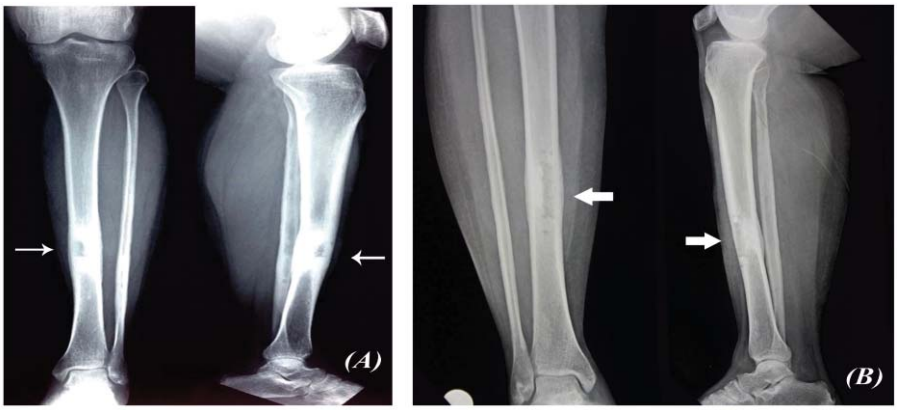

Figure 4: (A) Frontal and lateral radiographs of the left leg obtained 8 months after surgery. (B) Frontal and lateral radiographs of the right leg, immediate post surgery. The tibial medullary canal was decompressed by making a cortical window (thick arrow). The follow-up radiograph (A) revealed a persistent endosteal hyperostosis and obliteration of the medullary canal (thin arrow).

Conclusions

Intramedullary sclerosis is an idiopathic uncommon condition affecting the shaft of the tibia and is an important differential diagnosis for painful diaphyseal sclerotic conditions. Medullary Decompression may offer relief to patients in whom pain does not resolve with conservative treatment.

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