Case Report

A rarity in itself an osseous neurilemmoma – a case study

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Abstract

Schwannomas or neurilemomas are relatively less frequent benign tumors; however, intraosseous schwannomas are even rarer benign tumours of the bones with characteristic radiological and histological features. Though the most common site for intraosseous neurilemmoma is the mandible, we hereby present a case of 18 year old male having femoral diphyseal neurilemmoma managed with wide excision, grafting and internal fixation.

Keywords: Osseous Neurilemmoma, wide excision, grafting and internal fixation.

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INTRODUCTION

Neurilemmoma is a benign soft tissue tumour arising from the nerve sheath. Years may elapse before it gets diagnosed and is finally treated[1]. Furthermore it rarely shows osseous involvement. It accounts for less than 0.1% of all bone neoplasms. Most common site for the osseous neurilemmoma is mandible. Other sites include the vertebra, ulna, humerus, sacrum, tibia, patella, and small bones of the hands[2]. Neurilemmomas are found at all ages but most commonly between age group of 20 to 50. Most lesions are asymptomatic. Most lesions may be observed without surgery. Curettage is effective for symptomatic or large lesions. En bloc excision is the ultimate treatment as this tumour rarely shows recurrence.[3]

CASE REPORT

An 18 year old male patient came to outpatient department with chief complaints of swelling over lower part of right thigh since 4 years with increase in size of the swelling and occasional pain since 4 months. There was no history of any trauma or any systemic or local event prior to development of the swelling. Swelling was painless and fixed in size initially without any functional limitation. Patient had ignored it until 4 months before, when he observed that swelling had gradually increased in size. And there was associated pain specially getting exaggerated after strenuous work and relieving after rest. This was not associated with fever and any other constitutional symptoms. General and systemic examination was found to be normal. On local examination, there was a single oval swelling, longitudinally placed over anterior aspect of the right distal thigh with distal margin of swelling lying approximately 5 cm proximal to superior pole of patella. On palpation there was no local rise of temperature and swelling was tender. It was of the size 12x5 cm approximately, firm to hard in consistency with irregular shape, and was non-mobile. Range of motion at hip and knee joints were complete and without any pain. There were no enlarged inguinal lymph nodes or distal neurovascular deficit. Patient walked with normal gait. Plain radiograph (fig-1) revealed a well defined lytic lesion with sclerotic margins at anterolateral aspect of the diaphysis of the femur.

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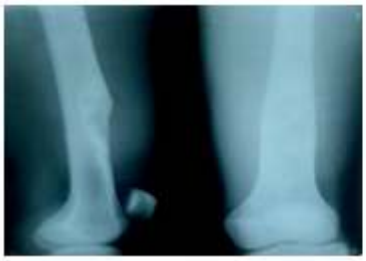

Figure 1:

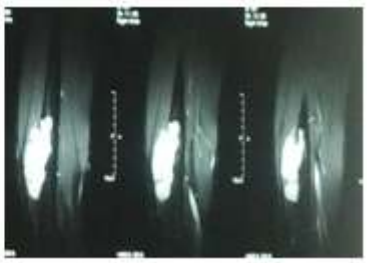

Figure 2:

There was no periosteal reaction and no evidence of fracture. Further MRI scan (fig 2) of the distal right thigh, showed a large (11 x 4 cm) expansile lesion in the distal diaphysis of the femur involving the corticomedullary junction. There was break in the cortex at the anterior aspect. The lesion was associated with a large multiseptaed, lobulated soft tissue in the intraosseous region and in periosteal region. Findings were suggestive of aggressive bony neoplasm, with possible diagnosis of Aneurysmal bone cyst or Osteochondroma. USG guided FNAC of Right Thigh Swelling showed only haemorrhagic aspirate and no other cells seen and thus was inconclusive. After all the routine investigations and fitness for anaesthesia, Patient was operated with 'En - block' excision of the mass along with 1 cm of normal bone surrounding it was done. During the procedure, an encapsulated multisepted mass breaching the anterior cortex was found. The Part of cortex was elevated, the intra osseous mass was curetted and cauterization was done of the medullary cavity. It was followed by fixation of the femur by 10 hole locking distal femur plate (fig 3) because the mass was involving approximately 50% of the total diameter of the weight bearing bone at that site, and filling the defect by graft taken from ipsilateral iliac bone and gel-foam. Gross examination (fig 4) of the specimen showed 10x6x4 cm encapsulated, fusiform, well circumscribed mass with external surface being grey white with part of it eroding the anterior cortex reaching up to the medullary cavity. Diagnosis established by histopathology (fig 5) to be a Neurilemmoma (ancient type), with extraosseus and intraosseus part with erosion of femoral cortex.

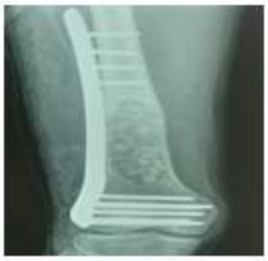

Figure 3:

Figure 4:

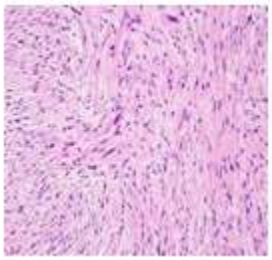

Figure 5:

DISCUSSION

While the term schwannoma has long been used, it has been noted that this tumor actually arises from nerve sheath cells, so neurilemmoma is a preferred term now a days[1]. It is a benign, well encapsulated tumor which forms a single fusiform firm mass on the course of a nerve. These are ectodermal in origin. Osseous neurilemmoma is a rare primary bone tumour.[5] The most common site of involvement is the mandible, with few other as mentioned above.[2] Within a bone, the tumour can be located in the medulla, nutrient canal or can be extra-osseous[6-9]. There are three mechanisms by which schwannomas can involve bone. The tumour can arise within the medullary cavity, from the nerve twigs along with nutrient artery within a nutrient canal[3] or from the periphery with secondary bone erosion.[9] When the tumour arises from the long bones, the most common location appears to be around the nutrient canal in the dia-metaphysis. Looking at the location of tumor in our case, we think the origin of the tumor could probably be from the minute nerve twigs surrounding the neutrient artery or from the nerve supply of the surrounding periosteum. Previously described radiological features include osteolysis with sclerotic borders, trabeculated contours, cortical erosions, and occasional central calcification.[10] Pathological fractures are not reported and neither were the malignant tranformations.[5] Clinical presentation of neurilemomma, particularly those which are deeply situated is often vague and confusing.[4] Patient sometimes has a radiating pain in the course of the nerve.[3] In our

case also, clinical presentation was unyielding. Differential diagnosis was aneurismal bone cyst and osteochondroma. Osseous neurilemmoma was not considered because of its extreme rarity. Even during operative procedure, due to very slow rate of growth of the tumor, it was believed to be benign, and en-block resection was performed instead of a simple biopsy.True nature of the tumor was determined only after histo-pathological examination. It was found to be a Neurilemmoma, with extraosseus and intraosseus part with erosion of femoral cortex. Microscopically, two types of cell arrangements, Antoni A and Antoni B, are found in schwannomas.[4] In our case, predominantly Type B areas were found. They were composed of Schwann cells arranged in a haphazard fashion and separated by a loose myxoid stroma. Type A areas have also been described which were made up of closely packed, spindle cells arranged in bundles and cords. All the intraosseous schwannomas reported have been benign.[9, 12] in our case there was also no signs of recurrence after one year of follow up without any functional limitation.

SUMMARY

In cases of osseous neurilemmoma, confirming the diagnosis of this notorious tumor is the key. If the diagnosis is confirmed, one can opt of watchful negligence if it is asymptomatic. If it becomes symptomatic, excision is recommended treatment modality as we have performed in our case.

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