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**Columns: Case Report**

**Unusual case of malignant peripheral nerve sheath tumor of proximal third tibia**

Rao A *et al.* MPNST of tibia

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**Abstract**

In this case study 16 year/male having swelling over anterior aspect of proximal 1/3 tibia since one year which was peanut in size initially and progressively increased to present size. Patient had undergone Fine Needle Aspiration Cytology (FNAC) twice during this period and reported as spindle cell sarcoma. Malignant peripheral nerve sheath tumour (MPNST) is a malignancy of the [connective tissue](http://en.wikipedia.org/wiki/Connective_tissue) surrounding the [nerves](http://en.wikipedia.org/wiki/Nerve). Previously MPNST is also called as neurofibrosarcoma, malignant schwannoma; neurogenic sarcoma. We are reporting this case for its rarity and peculiar mode of presentation. FNAC/Core biopsy can be used as an effective diagnostic tool to hit the correct pathological diagnosis.

**Key words:** Tibial malignant peripheral nerve sheath tumour; Fine Needle Aspiration Cytology; Histopathology; Treatment

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**Core tip:** In cases of malignant peripheral nerve sheath tumour of tibia Fine Needle Aspiration Cytology/core biopsy can be used as an effective diagnostic tool to hit the correct pathological diagnosis. In such cases enblock resection is the treatment of choice. Adjuvant radiotherapy/chemotherapy plays vital role for the better outcome of the patients.

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**INTRODUCTION**

Malignant peripheral nerve sheath tumours (MPNSTs) are sarcomas originating from cells associated with the nerve sheath. The lifetime risk of MPNST has a 0.001% in the general population. As MPNSTs arise from different types of cells associated with nerve sheath, *i.e*., Schwann cells, fibroblasts *etc*., the clinical presentation and histopathological features varies from case to case. So it’s a real challenge to diagnose and classify this rare entity. Generally, a sarcoma originating from a peripheral nerve or a [neurofibroma](http://en.wikipedia.org/wiki/Neurofibroma) is assumed clinically as MPNST[1,2].

**CASE REPORT**

A 16yrs year old boy admitted in YCR hospital Latur,with swelling which was peanut size when he first noticed and progressively increased to present size (10 cm × 8 cm) and pain which was intermittent in nature in right Proximal tibia with tingling sensation of right leg for the last one year.

On examination revealed a swelling over the anterior aspect of proximal end of tibia around 10 cm × 8 cm in size(Figure 1), shiny skin, scab in the center of swelling, dilated veins seen over swelling, local temperature raised with tenderness, it was mobile and not attached to underlying structures and range of movements of right knee joint were full and free with intact neurovascular status. No history of exposure to radiation. No evidence of signs and symptoms of Neurofibromatosis (Figure 2).

***Management***

Anteroposterior and lateral radiograph of the Right Knee with tibia shows an expansile soft tissue mass destroying adjacent cortex on lateral view but does not extent in to medullary cavity, congruency of knee joint is well maintained (Figure 3).

MRI showed a lobulated mass lesion (7.5 cm × 3.9 cm × 1.6 cm) along anterior surface of shaft of tibia causing periosteal elevation. There is no extension of lesion within the medullary space of tibia and no significant marrow edema in adjacent tibia (Figures 4-7).

Considering the nature of growth and high clinical propensity for malignancy, treated by *en-bloc* resection and immobilization for two weeks.

In this procedure through antero-medial approach around 20 cm “Elliptical “shaped incision from margin taken and radical en-bloc resection of tumor (measuring 15 cm × 8 cm × 4 cm) removed out. On Gross examination, cut section was grayish whitish mass with yellowish cut surface(Figure 8).

Care was taken to preserve the neurovascular bundle while resection of the tumor mass from the surrounding soft tissue. Thorough wound wash was given with H2O2 and wound closed over drain.

*En-bloc* section of excised mass sent for histo-pathological examination, report was malignant spindle cell sarcoma, *i.e*., Low grade MPNST (Figure 9). The tumor cells were immunopositive for S-100, thus the final diagnosis of MPNST was confirmed (Figure 10).

The limb was immobilized in a longmedium knee brace for 2 wk and followed by active knee mobilization. Patient discharged and advised monthly review. Patient also advised to consult oncologist for chemotherapy/radiotherapy. In our case considerable delay in the treatment was observed by looking at the size of swelling. Near total excision was done. Stitch gaping of the wound of about 1.5 cm noted which healed eventually.

**DISCUSSION**

Malignant Peripheral Nerve Sheath Tumors constitutes 5%-10% of all soft tissue malignancies. They are associated with neurofibromatosis-1 (NF1) or may occur independently in a spontaneous manner.

The cause is not known but strongly associated with history of exposure to radiation[3,4]. Fifty percent of the cases occur in patients with NF1[5-7]. They usually occur in the preexisting neurofibroma.

The genesis of MPNST has been associated with genetic mutations in p53 and p16 genes[8-10]. While NF1 gene activity acts as a predisposing factor.

They are commonly seen in adultshood, age group varies between 20-50 years. In first 2 decades of life the incidence is 10%-20%[6], with exceptionally seen in infants also[11].

The plan of treatment for MPNSTs is surgical excision with wide margins. Adjuvant chemotherapy or radiotherapy has not been shown to be effective for better outcome of patients[12,13].

It has been clearly stated that these tumors have tendency to spread for considerable distances along nerves. In such scenario, frozen section can be advised to ensure clear margins[14].

In a 10 year institutional review for chemotherapy, it did not seem to reduce mortality, so its effectiveness is questionable. Although with recent approaches with the molecular biology of MPNSTs, new therapies and prognostic factors are being examined[15].

**COMMENTS**

***Case characteristics***

A 16 years old boy presented with swelling which was peanut size when he first noticed and progressively increased to present size (10 cm × 8 cm) and pain which was intermittent in nature in Right Proximal tibia with tingling sensation of right leg for the last one year.

***Clinical diagnosis***

Clinically the case was diagnosed as soft tissue sarcoma.

***Differential diagnosis***

Soft tissue sarcomas, *i.e*., fibrosarcoma, malignant fibrous histiocytoma and malignant peripheral nerve sheath tumour (MPNST).

***Laboratory diagnosis***

On Fine Needle Aspiration Cytology (FNAC) case was diagnosed as spindle cell sarcoma which was confirmed on histopathology and immunostaining.

***Imaging diagnosis***

X-ray: Anteroposterior and lateral radiograph of the Right Knee with tibia shows an expansile soft tissue mass destroying adjacent cortex on lateral view but does not extent in to medullary cavity, congruency of knee joint is well maintained. MRI showed a lobulated mass lesion (7.5 cm × 3.9 cm × 1.6 cm) along anterior surface of shaft of tibia causing periosteal elevation. There is no extention of lesion within the medullary space of tibia and no significant marrow edema in adjacent tibia.

***Pathological diagnosis***

Malignant peripheral nerve sheath tumor confirmed on immunohistochemistry.

***Treatment***

Enblock resection followed by chemotherapy/radiotherapy.

***Experiences and lessons***

FNAC/Core biopsy can be used as an effective diagnostic tool to arrive at early diagnosis.

***Peer-review***

It is a well written paper describing an interesting case report of MPNST of proximal third tibia treated by *en-bloc* resection.

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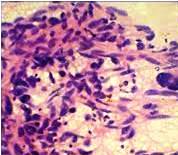
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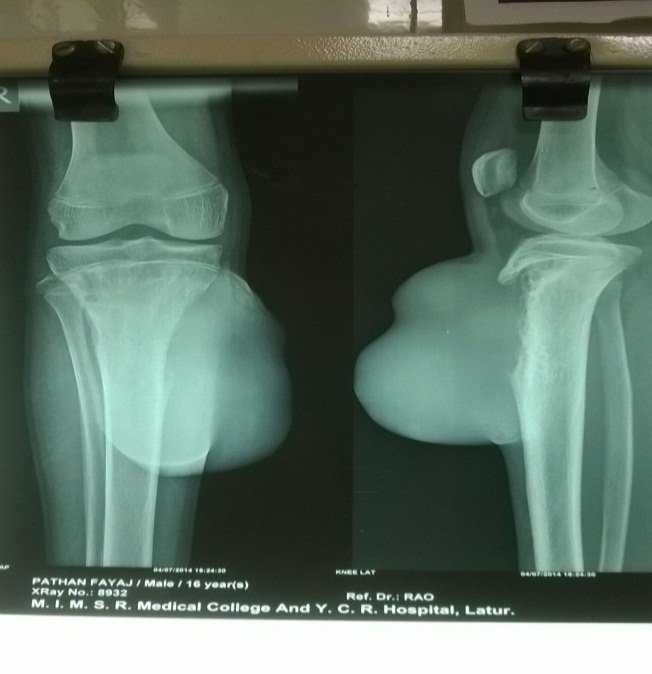
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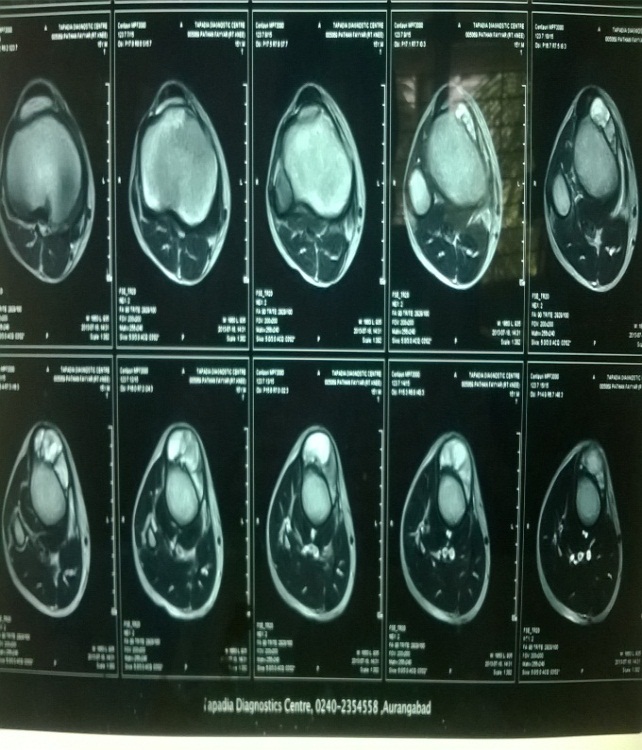
**Figure 1 Pre-operative clinical photographs.**

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**Figure 2 Fine Needle Aspiration Cytology Showingloosely scattered malignant spindle cells.**



**Figure 3 Pre-operative X-ray.**



**Figure 4 MRI transverse section.**



**Figure 5 MRI coronal section**



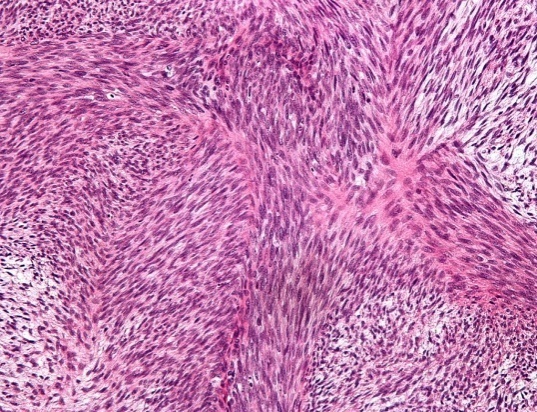
**Figure 6 MRI saggital section.**



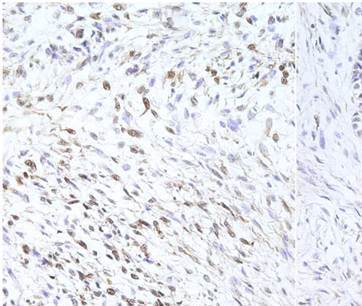
**Figure 7 MRI saggital section.**



**Figure 8 Intra operative photograph showing excised mass (Measuring 15 cm × 8 cm × 4 cm).**



**Figure 9 Showing MPNST on Microscopy (LP 10 ×).**



**Figure 10 Showing S-100 immunopositive tumor cells.**