

Malignant peripheral nerve sheath tumor of proximal third tibia

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Abstract

A 16-year-old man had a swelling over the anterior aspect of the proximal third of the tibia for 1 year, which was peanut size initially and progressively increased to its present size of 10 cm × 8 cm. He underwent fine needle aspiration cytology (FNAC) twice during this period and reported spindle cell sarcoma. Malignant peripheral nerve sheath tumor (MPNST) is a malignancy of the connective tissue surrounding the nerves. Previously, MPNST was also known as neurofibrosarcoma, malignant schwannoma, and neurogenic sarcoma. We are reporting this case for its rarity and peculiar mode of presentation. FNAC/core biopsy can be used as an effective tool to achieve the correct pathological diagnosis.

Key words: Tibial malignant peripheral nerve sheath tumor; Fine needle aspiration cytology; Histopathology

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Core tip: In cases of malignant peripheral nerve sheath tumor of the tibia, fine needle aspiration cytology/core biopsy can be used as an effective tool to achieve the correct pathological diagnosis. In such cases, *en bloc* resection is the treatment of choice. Adjuvant radiotherapy/chemotherapy plays a vital role in achieving a good outcome.

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INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas originating from cells associated with the nerve sheath. The lifetime risk of MPNST is 0.001% in the general population. As MPNSTs arise from different types of cells associated with nerve sheaths, for example, Schwann cells and fibroblasts, the clinical presentation and histopathological features varies from case to case. So, it is a real challenge to diagnose and classify this rare entity. Generally, a sarcoma originating from a peripheral nerve or a neurofibroma is assumed clinically as MPNST^[1,2].

CASE REPORT

A 16-year-old man was admitted to YCR Hospital Latur, with a peanut-size swelling when it was first noticed, which progressively increased to its present size of 10 cm × 8 cm. Pain was intermittent in the right proximal tibia, with tingling sensation in the right leg for the previous year.

Physical examination revealed a swelling over the anterior aspect of the proximal end of the tibia (10 cm × 8 cm; Figure 1), shiny skin, a scab in the center of swelling, dilated veins over the swelling, and local temperature increase with tenderness. The swelling was mobile and not attached to underlying structures. The range of movements of the right knee joint was full and free, with intact neurovascular status. There was no history of exposure to radiation and no evidence of signs and symptoms of neurofibromatosis (NF) (Figure 2).

Management

Anteroposterior and lateral radiography of the right knee and tibia showed an expansile soft-tissue mass destroying the adjacent cortex on lateral view, but it did not extend into the medullary cavity. Congruency of the knee joint was well maintained (Figure 3).

Magnetic resonance imaging showed a lobulated mass lesion (7.5 cm × 3.9 cm × 1.6 cm) along the anterior surface of the tibial shaft, which caused periosteal elevation. There was no extension of the lesion within the medullary space of the tibia and no significant bone marrow edema in the adjacent tibia (Figure 4).

Considering the nature of the growth and high clinical propensity for malignancy, it was treated by *en bloc* resection and immobilization for 2 wk.

In this procedure, through an antero-medial approach, around 20 cm an elliptical incision of around 20

cm was made and radical *en bloc* resection of the tumor was performed.

Care was taken to preserve the neurovascular bundle during resection of the tumor from the surrounding soft tissue. The wound was washed thoroughly with H₂O₂ and the excised mass was sent for histopathological examination. On gross examination, the cut surface was gray-white (Figure 5) and on histopathological examination, the mass was diagnosed as malignant spindle cell sarcoma, *i.e.*, low-grade MPNST (Figure 6). The tumor cells were immunopositive for S-100, thus confirming the final diagnosis of MPNST (Figure 7).

The limb was immobilized in a long/medium knee brace for 2 wk and followed by active knee mobilization. The patient was discharged and advised to attend monthly review. He was also advised to consult an oncologist for chemotherapy/radiotherapy.

DISCUSSION

MPNSTs constitute 5%-10% of all soft-tissue malignancies. They are associated with NF-1, or may occur independently in a spontaneous manner.

The cause is not known, but they are strongly associated with history of exposure to radiation^[3,4]. Fifty percent of the cases occur in patients with NF-1^[5-7], and they usually occur in a pre-existing neurofibroma.

The genesis of MPNSTs is associated with genetic mutations in *p53* and *p16* genes^[8-10]. NF-1 gene activity acts as a predisposing factor.

MPNSTs are commonly seen in adults, aged 20-50 years. In the first two decades of life, the incidence is 10%-20%^[6], with exceptional cases seen in infants^[11].

The plan of treatment for MPNSTs is surgical excision with wide margins. Adjuvant chemotherapy or radiotherapy does not achieve a better outcome^[12,13].

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

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

COMMENTS

Case characteristics

A 16-year-old man presented with a peanut-size swelling, when first noticed, which progressively increased to its present size of 10 cm × 8 cm, and intermittent pain in the right proximal tibia and a tingling sensation in the right leg for the past year.

Clinical diagnosis

The case was diagnosed as soft tissue sarcoma.

Differential diagnosis

Soft tissue sarcomas, that is, fibrosarcoma, malignant fibrous histiocytoma, and

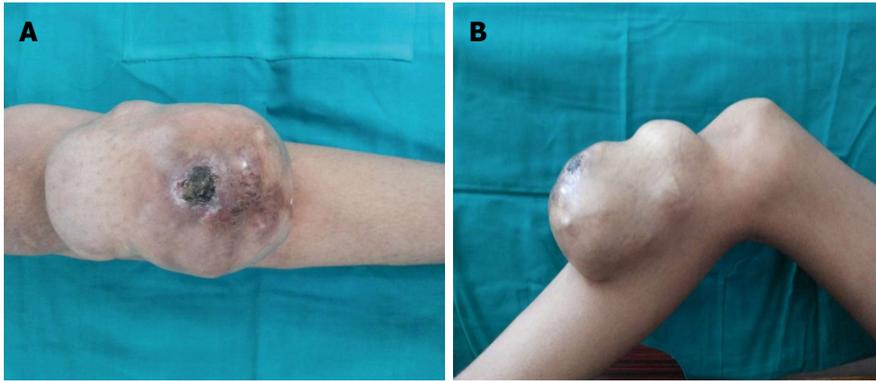


Figure 1 Preoperative clinical photographs (A and B).

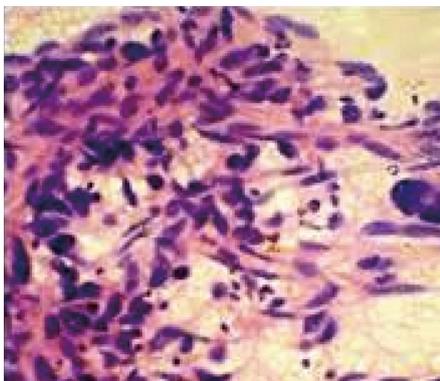


Figure 2 Fine needle aspiration cytology showing loosely scattered malignant spindle cells.



Figure 3 Preoperative X-ray.

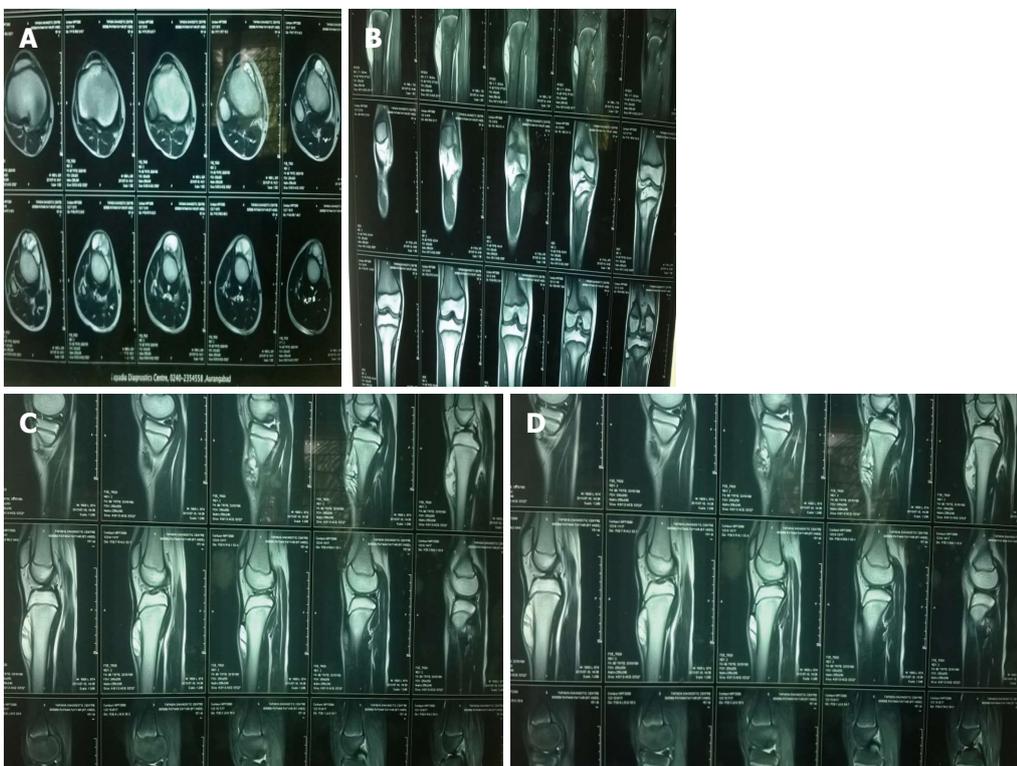


Figure 4 Magnetic resonance imaging transverse (A), coronal (B) and saggital (C, D) section.

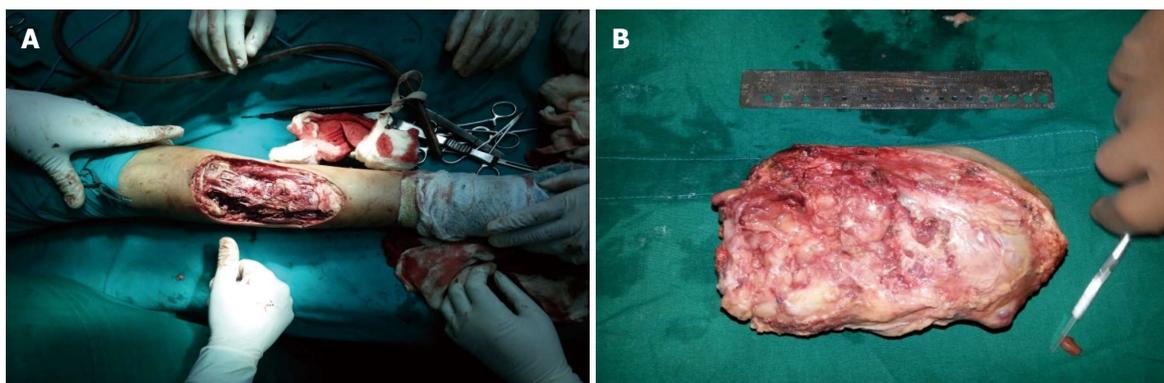


Figure 5 Intraoperative photograph showing excised mass (15 cm × 8 cm × 4 cm) (A and B).

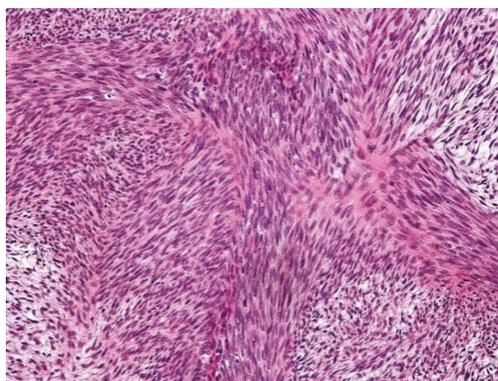


Figure 6 Malignant peripheral nerve sheath tumor on microscopy (LP 10 ×).

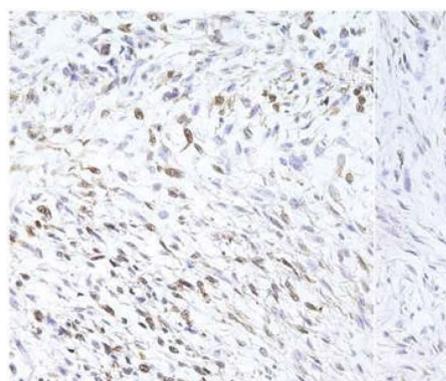


Figure 7 S-100 immunopositive tumor cells.

malignant peripheral nerve sheath tumor (MPNST).

Laboratory diagnosis

On fine needle aspiration cytology (FNAC), the case was diagnosed as spindle cell sarcoma, which was confirmed by histopathology and immunostaining.

Imaging diagnosis

X-ray: Anteroposterior and lateral radiography of the right knee and tibia showed an expansile, soft tissue mass destroying adjacent cortex on lateral view, but it did not extend into the medullary cavity; congruency of the knee joint was well maintained. Magnetic resonance imaging showed a lobulated mass lesion (7.5 cm × 3.9 cm × 1.6 cm) along the anterior surface of the tibial shaft, causing periosteal elevation. There was no extension of the lesion within the medullary space of the tibia and no significant bone marrow edema in the adjacent tibia.

Pathological diagnosis

MPNST was confirmed by immunohistochemistry.

Treatment

En bloc resection followed by chemotherapy/radiotherapy.

Experiences and lessons

FNAC/core biopsy can be used as an effective diagnostic tool to achieve 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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