



Case Report

Malignant Peripheral Nerve Sheath Tumor of Popliteal Fossa

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ABSTRACT

A 54-year-old female presented with a large mass associated with pain at left popliteal region with fungating surface ulceration on skin. Patient was known case of localized (sporadic) neurofibroma at the same site operated 4 years back. Histopathological examination revealed a Malignant Peripheral Nerve Sheath Tumor (MPNST) of high grade nature. We report this case for its clinical behavior, large size, recurrence and histopathological features.

Key Words: Malignant Peripheral Nerve Sheath Tumor, Soft tissue sarcoma, Popliteal fossa.

INTRODUCTION

Malignant Peripheral Nerve Sheath Tumor (MPNST) comprises approximately 5-10% of all soft tissue sarcomas. ^(1,2) Malignant tumors arising from peripheral nerves or displaying differentiation along the lines of various elements of the nerve sheath e.g. Schwann cell, fibroblast, perineural cells are collectively referred as MPNSTs. The term MPNST replaces previously used names like malignant Schwannoma, neurofibrosarcoma and neurogenic sarcoma. ⁽³⁾ They can occur either spontaneously or in association with neurofibromatosis-1(NF-1). ⁽⁴⁾ In our case it was arising spontaneously in an adult patient, with changing clinical behavior.

CASE REPORT

A 54-year-old female presented to our hospital with a large mass at left popliteal region, associated with pain and weakness in lower extremity. She had history of similar swelling at the same site operated 4 year back, diagnosed as localized neurofibroma. The current swelling recurred 1 year back with gradual increase in size, but with-in last 4 months swelling rapidly enlarged giving surface skin ulceration and fungation. Patient did not have any history of occupation or therapeutic radiation, family history or any other systemic illness. Radiological examination showed soft tissue mass at popliteal region approximately measuring 16x12x8 cm with heterogeneity, ill-defined margins and surrounding soft tissue edema with no evidence of distant metastasis. Mass was excised with wide

surgical margins and sent for histopathological examination.

Gross:

We received a single, large, ovoid mass with ill-defined margins. The mass was covered with skin. Mass measured 15x12x7cm and weighed 370gm with attached thick popliteal nerve. Cut section showed grey-white, bossilated, and firm mass with variegated appearance with focal mucoid and hemorrhagic areas (Figure-1).

Light microscopic examination:

We studied multiple sections from tumor mass which showed spindle cells arranged in diffuse sheets, fascicles and wavy pattern (Figure-2). Individual tumor cells were spindle to ovoid having moderately pleomorphic nuclei with occasional nucleoli and moderate amount of eosinophilic cytoplasm with occasional tumor giant cells (Figure-3). Hypo and hyper dense cellular areas were noted. In areas high pleomorphic cells with increased mitosis (>6/10HPF) was noted. Areas of haemorrhage, necrosis and myxoid changes were noted. Tumor around blood vessel wall was noted (Figure-4).



Figure 1: Gross specimen showing grey-white, bossilated, and firm mass with variegated appearance.

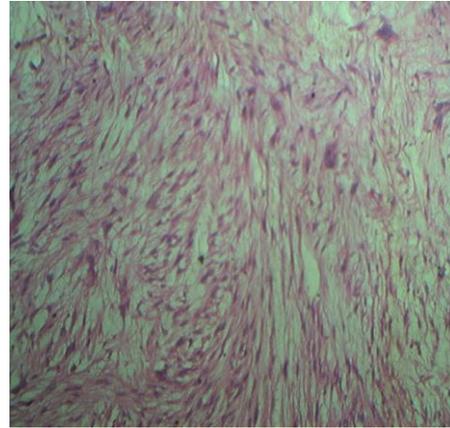


Figure-2: Photomicrograph showing neoplastic spindle cells in sheets, fascicles and wavy pattern (H and E stain, 100x)

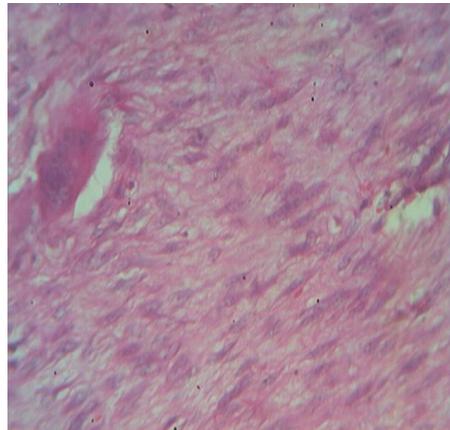


Figure-3: Photomicrograph showing spindle to ovoid tumor cells with pleomorphic nuclei and occasional giant cell (H and E stain, 400x)

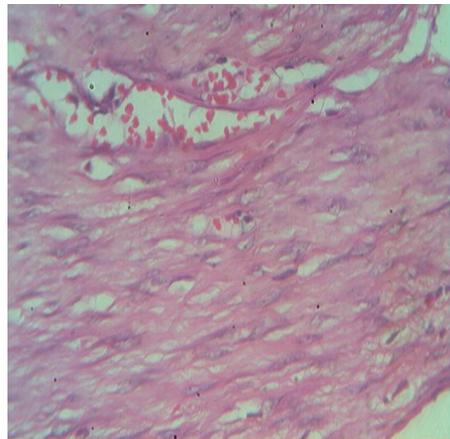


Figure-4: Photomicrograph showing tumor cells around blood vessels (H and E stain, 400x)

DISCUSSION

MPNST is a rare variety of soft tissue sarcoma of ectomesenchymal origin.

As MPNST can arise from multiple cell types the overall diagnosis and classification is somewhat difficult. The diagnostic approach towards soft tissue sarcoma to define as MPNST if one of the following criteria is essential:-

1. The tumor originating from a peripheral nerve
2. It arises from a pre-existing benign nerve sheath tumor (neurofibroma)
3. On histopathologic examination it demonstrates Schwann cell differentiation.

In our case it was arising from peripheral popliteal nerve. Patient was previously diagnosed as a case of sporadic neurofibroma which recurred and showed sudden enlargement in size of pre-existing mass, which favors the criteria.

MPNST can occur either spontaneously or in association with neurofibromatosis-1. The evidence of MPNST is of 1 per 1,00,000 population, however it can increase to 5-42% in patients with neurofibromatosis-1. ^(1,5) MPNST is mostly seen in adult life in an age group of 20-50 years. These lesions are commonly seen in upper and lower extremities and trunk as enlarged fusiform masses. Sudden enlargement in pre-existing mass with pain indicates possibility of malignant transformation, which was observed in our case.

On histopathological examination approximately 80-85% of MPNST are spindle cell tumors with fasciculating pattern. While remaining 15% shows variable differentiation like glandular, rhabdoid, chondroid, myxoid, epitheloid, osteoid etc. ⁽⁶⁾ In our case it began with neurofibroma and over a period of 4 years it recurred and transformed to a high grade MPNST. Other differentiation was not noted in our case on extensive tissue sampling. Areas of haemorrhage and necrosis were

noted and also tumor encroaching around and in the wall of blood vessel was noted.

Most MPNST are histopathologically diagnosed but differential diagnosis like fibrosarcoma, leiomyosarcoma, monophasic synovial sarcoma, pleomorphic sarcoma etc. should be kept in mind.

These cases are managed by surgical resection, chemotherapy, radiotherapy, brachytherapy and combination, but the results have been variable. ⁽⁷⁾

Our patient received treatment of surgical excision of mass with wide surgical margins. There was no evidence of any local, regional or distant metastasis on clinical and radiological evaluation. Patient is disease free and on regular follow-up for last 6 months.

MPNST has a highest recurrence rate of any sarcoma ⁽⁸⁾ and adequate initial treatment gives the better chance to survival.

CONCLUSION

MPNST is a rare variety of soft tissue sarcomas, these are often difficult to diagnose, behave aggressively and despite appropriate treatment recurrence is high.

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