

Malignant Peripheral Nerve Sheath Tumor from Arm and Forearm

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Abstract:

Malignant peripheral nerve sheath tumor (MPNST) comprises 5-10% of all soft tissue sarcomas. It occurs in conjunction with large peripheral nerves such as the sciatic nerve, the brachial plexus, and the sacral plexus. Here under a case of MPNST arising from left arm and elbow region is reported due to its rarity.

Keywords: MPNST; nerve sheath tumor; arm, forearm

Introduction

Malignant peripheral nerve sheath tumor (MPNST) comprises 5-10% of all soft tissue sarcomas. It occurs in conjunction with large peripheral nerves such as the sciatic nerve, the brachial plexus, and the sacral plexus. Here under a case of MPNST arising from left arm and elbow region is reported due to its rarity.

Case Report

A 35 year old female presented with 6 months history of pain in the left arm and forearm and progressive swelling in the left elbow region. There was no history of previous swelling at the site, fever, loss of weight or loss of appetite. Examination revealed a circumferential swelling situated from the middle arm to the proximal forearm without involvement of skin. Swelling was non tender, firm in consistency, margins were ill defined and it was not attached to the bone. Soft tissue neoplasm, possibly neurofibroma was considered as the clinical diagnosis. X ray of the arm and forearm and chest did not show any abnormality. Ultrasonography of the elbow region showed well defined large heterogeneous lesion in the distal arm and proximal forearm adjacent to the humerus and radius without involving the brachial artery and its divisions. There was mild to moderate vascularity in the lesion.

MRI (Figure-1,2,3) showed extensive lobular soft tissue lesion of size 183mm x 47mm x 46mm in the lateral and

anterior compartment of lower arm, elbow and proximal forearm. It was isointense on T1W and hyperintense on T2W and PDF. Findings suggested neoplastic lesion of neural origin from peripheral nerve sheath. FNAC suggested round cell sarcoma, epithelioid variant of MPNST.

The patient was taken up for wide excisional biopsy of the tumor but the muscles were extensively found to be involved on all sides and dissection was not possible. Hence shoulder disarticulation was carried out with proper explanation and consent of the patient's relation. The excised tumor was submitted for histopathology which gave the final diagnosis as malignant peripheral nerve sheath tumor (MPNST). There was evidence of infiltration by malignant mesenchymal cells between muscle bundles. The patient was explained the need of immunohistochemistry, radiotherapy and further regular follow up to detect any recurrences and metastasis. At 6 months follow up the patient was asymptomatic and did not have any evidence of local recurrence or distant metastasis.

Discussion

MPNSTs are sarcomas from peripheral nerve or cells associated with the nerve sheath such as Schwann cells, perineural cells or fibroblasts. MPNST term replaces previously used names including malignant Schwannoma, neurofibrosarcoma and neurogenic sarcoma. The tumor can arise spontaneously or in association with neurofibromatosis. The etiology is unknown but there is a higher incidence in patients with a history of radiation exposure [1, 2]. Up to 50% of MPNSTs occur in patients of neurofibromatosis [3]. These typically occur in adults but rarely reported in 2nd decade and children also. The tumor presents as an enlarging palpable mass which may cause radicular pain, paresthesia and motor weakness. MRI is the imaging modality of choice. Size bigger than 5cm, involvement of fat planes,

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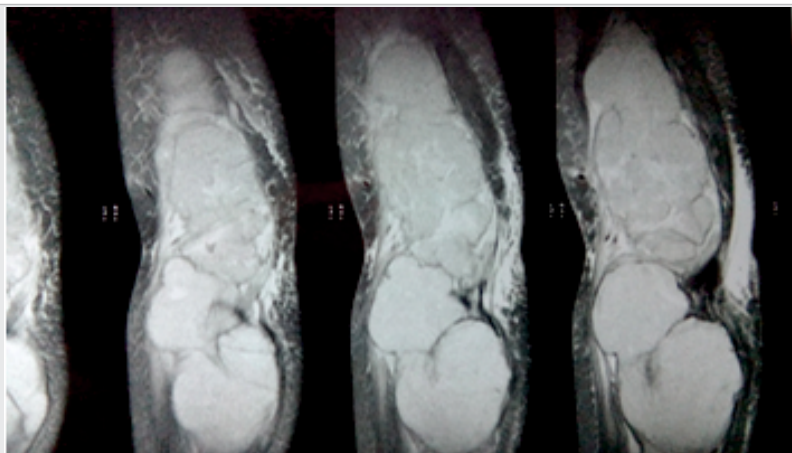


Figure-1, MRI sagittal section of right arm with forearm showing lobulated tumor mass with variable intensity

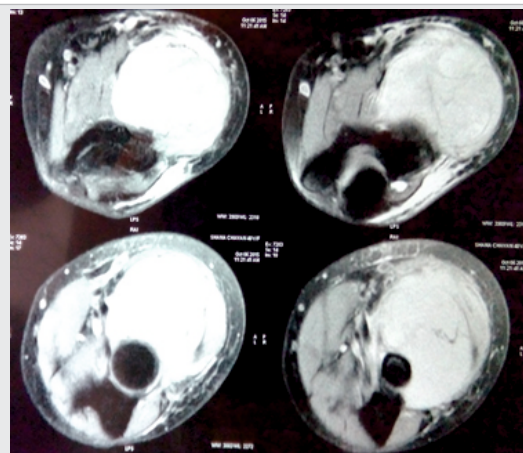


Figure-2, MRI axial sections at the level of arm showing tumor mass encircling the brachial artery. No vascular invasion.

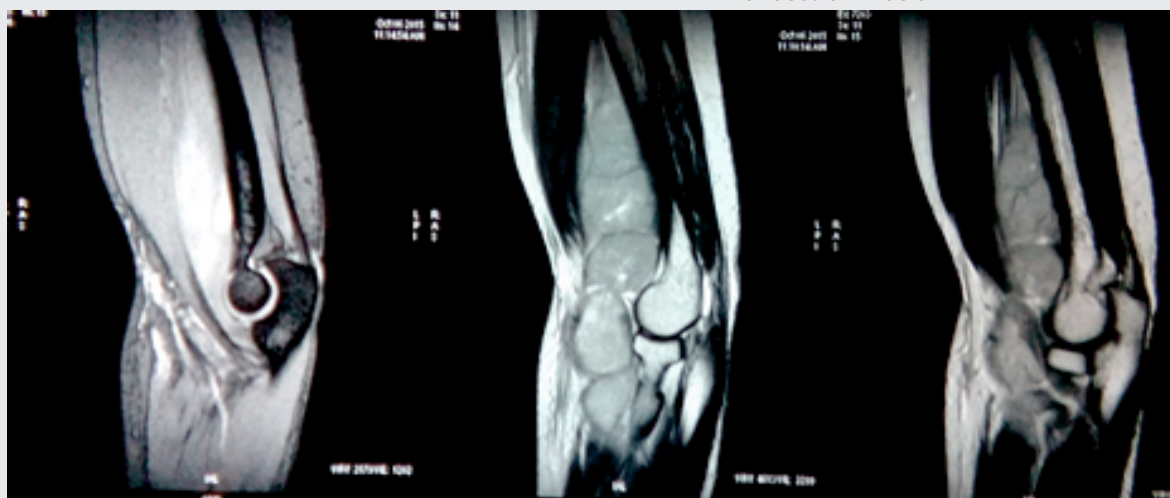


Figure-3, MRI sagittal section showing tumor mass extending from distal third arm lying anterior to distal third humerus and radius

heterogeneity, ill defined margins and edema surrounding the lesion is more suggestive of MPNST. They most commonly metastasize to lungs followed by bones and finally the pleura. Bone scan helps in the detection of bony metastasis. The main stay of treatment is surgical resection. Complete surgical resection with wide margins or amputation offers the best outcome with respect to both local recurrence and distant

metastasis. Radiation therapy and chemotherapy are adjuvant modalities and used for local control and metastasis. 80-90% of MPNSTs are spindle cell tumors with fasciculating pattern that contain histological features similar to fibrosarcoma. Local recurrence and distant metastasis are reported in 40-65% cases [4, 5]. 5 year survival rate is reported to be from 16-52%.

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