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Case Report

Malignant Peripheral Nerve Sheath Tumor of chest wall in a patient with Neurofibromatosis: A case report

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ABSTRACT
Malignant peripheral nerve sheath tumor is a relatively rare tumor in general population with an incidence of 0.001%. But in patients of Neurofibromatosis the incidence is 2 -5%. It usually affects the extremities in these patients and rarely occurs in other sites like trunk, head and neck, intra spinal region, scalp and Intra cranial regions. We present a case of malignant peripheral nerve sheath tumor of the chest wall in an elderly patient with Neurofibromatosis with review of literature.
Key words: Malignant Peripheral Nerve Sheath Tumor, Neurofibromatosis -1

INTRODUCTION
Primary tumors of chest wall are rare and MPNSTs constitute 5% of them. MPNST is defined as any sarcoma arising from a nerve or from a preexisting neurofibroma or any malignant spindle cell tumor in a patient with Neurofibromatosis -1(von Recklinghausen disease). [1] It's incidence in general population is 0.001%. About 50% occur in patients with Neurofibromatosis-1, 10% are induced by radiation and 40 % are sporadic. [2] The latent Period for MPNST in Neurofibromatosis is 20 years whereas it is 15.5 years in post irradiation cases.

CASE REPORT
A 60 year old male patient presented with complaints of multiple neurofibromas of varying sizes all over his body since 30 years. On examination there was a large painful nodule on his right chest wall measuring 7 x 4 cms around the nipple. His laboratory parameters were normal .Ultrasound revealed a mixed echoic lesion on the same region. FNAC was reported as spindle cell tumor .Wide local excision of the tumor was done and sent for histopathological examination, which revealed spindle shaped cells arranged in fascicles with mild nuclear pleomorphism, 1-2 mitoses/high power field and giant cells. All the resected margins were negative, and a diagnosis of MPNST –low grade (stage -1 STS staging, AJCC) was given. Patient was sent for adjuvant radiotherapy and followed up.

DISCUSSION
MPNST or Malignant Schwannoma or Neurofibrosarcoma is an aggressive tumor seen mostly in NF-1 or post irradiation patients. It is seen in 50% of patients with NF1, around 2nd to 5th decade of life without any sex predilection as an enlarging painful mass. It is an aggressive tumor affecting the extremities, trunk, head and neck and intraspinal regions. [3] According to the model for pathogenesis Of MPNST advocated by Timothy Beer of Jefferson Medical center, mutation in Neurofibromin gene and dysregulation of RAS, CAMP, Ca+2, lead to plexiform neurofibromas. Upregulation of EGFR, ERB2, C-KIT, and down regulation of p53, p16, p19, and RB are the factors causing MPNST. [4] Histopathologically MPNSTs are characterized by spindle shaped cells with nuclear pleomorphism, frequent mitoses. [5] IHC with S100 shows focal positivity in 50% of cases, differentiating it from plexiformneurofibroma which shows diffuse positivity. 50% of MPNST cases are positive for Leu -7 and Major Basic Protein. [6]
Although Zou et al reported that only positive margins were associated with local recurrence [8], Wong et al stated that adjuvant radiotherapy was required for better disease control [9] Vincent et al reported 57% disease free period and 80% overall survival 2 years after a combination of surgery, radiation and chemotherapy with doxorubicin and ifosfamide in 10 patients with MPNST. According to studies by various authors the favorable prognostic factors are tumor size, complete surgical resection, and histologic grade, tumor location (extremity vs. trunk and head and neck).

CONCLUSION
MPNST of chestwall is a relatively rare tumor. It should be considered in the differential diagnosis of chest wall tumors in patients with Neurofibromatisis -1, and in patients receiving irradiation. Complete resection with adequate surgical margins and adjuvant radiotherapy are the main madalities of treatment of MPNST .In High grade tumors, adjuvant chemotherapy is given to prevent local recurrence.

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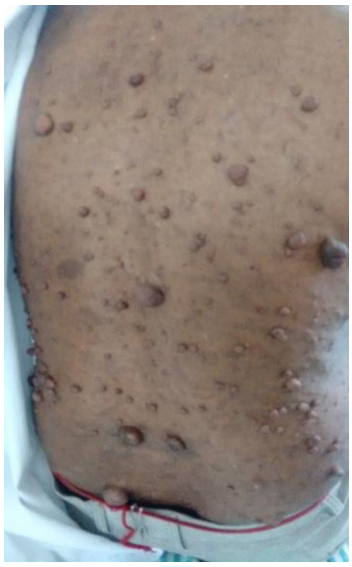
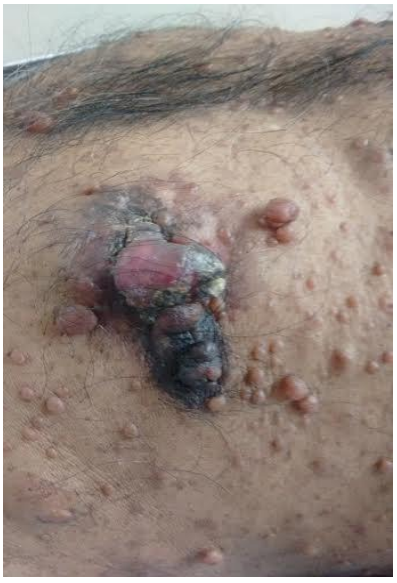
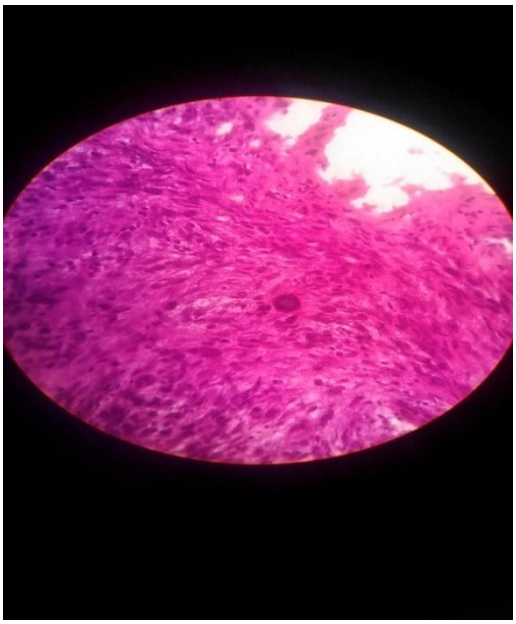
Photograph 1 showing multiple neurofibromas on the back



Photograph 2 showing a large tumor on the right chest wall region around the nipple measuring 7 x 4 cm, grey tan in color



Photograph 3: Microscopic picture of the excised tumor - (Hematoxylin and eosin stain x 400) showing spindle shaped cells arranged in fascicular manner with nuclear pleomorphism and a giant cell.



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