

# POSTER ID 60: MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR IN A CASE OF NEUROFIBROMATOSIS

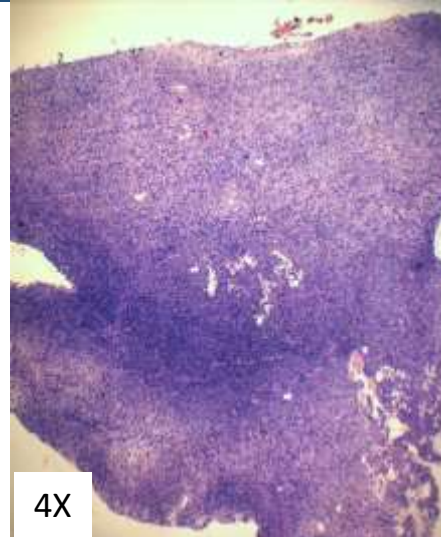
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A 51 year old male with Neurofibromatosis type 1

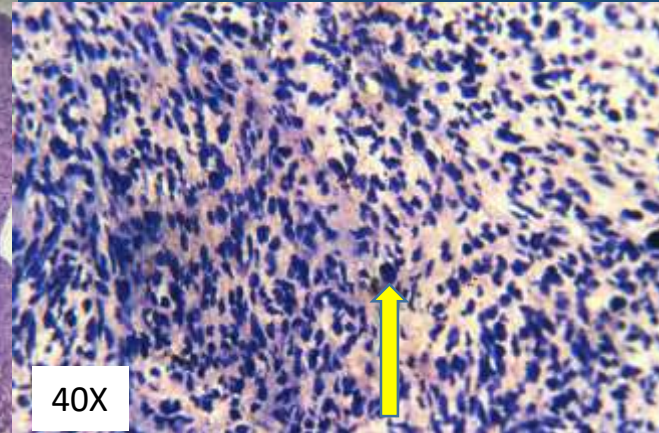
- A single, non-tender, firm, mobile mass, about 8 x 8 cm in size, with normal overlying skin, free from underlying structures, displacing the nipple laterally on left side of chest, rapidly enlarging since 5 months.
- Painless, accompanied only by local discomfort.
- Firm, mobile, non tender left axillary lymph nodes



Bilateral palmar freckling with palmar neurofibromas



4X  
Alternate areas of dense & sparse infiltrate



40X  
Multiple atypical cells with variation in nuclear size. Few Giant Nuclei (arrow)

### CT-SCAN



Large heterogenous lesion in left paravertebral location at the thoracic inlet with underlying bony erosion and sclerosis. (arrow)  
Right axillary lymphadenopathy.

IHC MARKERS	
S-100	+
CD-56	+
EMA	+
CD 34	-
DESMIN	-
MYOGEN	-

**TREATMENT**  
Complete surgical extirpation was performed and the patient was started on chemotherapy with Paclitaxel 100 mg injection every 8 days.

**DISCUSSION**

1. Conversion of a Plexiform neurofibroma into a MPNST- 3-15%.
2. Usually consist of perineural and endoneural fibroblasts rather than schwann cells.
3. Challenging to treat- chances of early metastasis and resistance to chemotherapy.
4. Complete surgical extirpation with clear margins with djuvant radiotherapy advocated.
6. Doxorubicin and Ifosphamide are the preferred chemotherapeutic agents.

## FINAL DIAGNOSIS- MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR

References- 1.Farid M, Demicco EG, Garcia R, et al. Malignant peripheral nerve sheath tumors. *Oncologist*. 2014;19(2):193-201.  
2.Yuan Z, Xu L, Zhao Z, et al. Clinicopathological features and prognosis of malignant peripheral nerve sheath tumor: a retrospective study of 159 cases from 1999 to 2016. *Oncotarget*. 2017;8(62):104785-104795.