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Case Report

A CASE OF MALIGNANT PERIPHERAL NERVE SHEATH TUMORS IN A 40 YEAR OLD FEMALE

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ABSTRACT
A case of malignant peripheral nerve sheath tumor, which is a rare variety of soft tissue sarcoma of ectomesenchymal origin in a 40 year old female. On MRI, there was a fusiform shaped C1-C2 posterior tumor, for which she got operated. On histopathology, there was a brownish white mass with rough external surface and on cut section, whitish homogenous areas with focal hemorrhagic areas are seen. Microscopically, there was perivascular concentration of tumor cells with plumper shaped epitheloid appearance of endothelial cells with palisading tumor cells at the edges of irregularly shaped necrotic areas. Abundant mitosis was seen. Immunohistochemistry showed spindle and glandular tumor component are expressing positivity for S-100.

Keywords: *Soft Tissue Tumor, MPNST*

INTRODUCTION
Malignant peripheral nerve sheath tumors (MPNSTs) are a rare variety of soft tissue sarcoma of ectomesenchymal origin (Hruban *et al.,* 1990).
WHO coined the term MPNST replacing previous heterogenous terminology, such as malignant schwannoma, malignant neurilemmoma and neurofibrosarcoma, for tumors of neurogenic origin and similar biological behavior (Wanebo *et al.,* 1993).
They arise from a major or minor peripheral nerve branches (5) or sheath of peripheral nerve fibres (Hirose *et al.,* 1998).
These tumors may arise spontaneously in adult patients, although 5% to 42% of MPNST have an association with multiple neurofibromatosis Type-I (Brasfield and Das Gupta, 1972). Surgery is the main stay of treatment of this tumor although they are biologically aggressive in nature.

CASES
A 40-year- old female presented with history of painless, slowly growing swelling over back. There is no significant medical or family history. The swelling measured 3x2.5cms and was soft to firm in consistency.

Investigation
MRI shows a fusiform shaped C1-C2 posterior tumor.

Histopathology
Received brownish white soft tissue bits along with one soft tissue mass aggregating 8x6x3.5cm. Single large mass measure 3.5cm in diameter.
External surface of mass is rough. On cut section, whitish homogenous areas with focal hemorrhagic areas are seen.
Microscopically perivascular concentration of tumor cells with plumper shaped epitheloid appearance of endothelial cells was seen.
Tumor cells were seen palisading at the edges of irregularly shaped necrotic areas. There was presence of large gaping vascular spaces resulting in hemangio-pericytoma like appearance. Abundant mitosis was present.

Immunohistochemistry
Immunohistochemical staining showed that the spindle and glandular tumor components were positive for S-100 100x.

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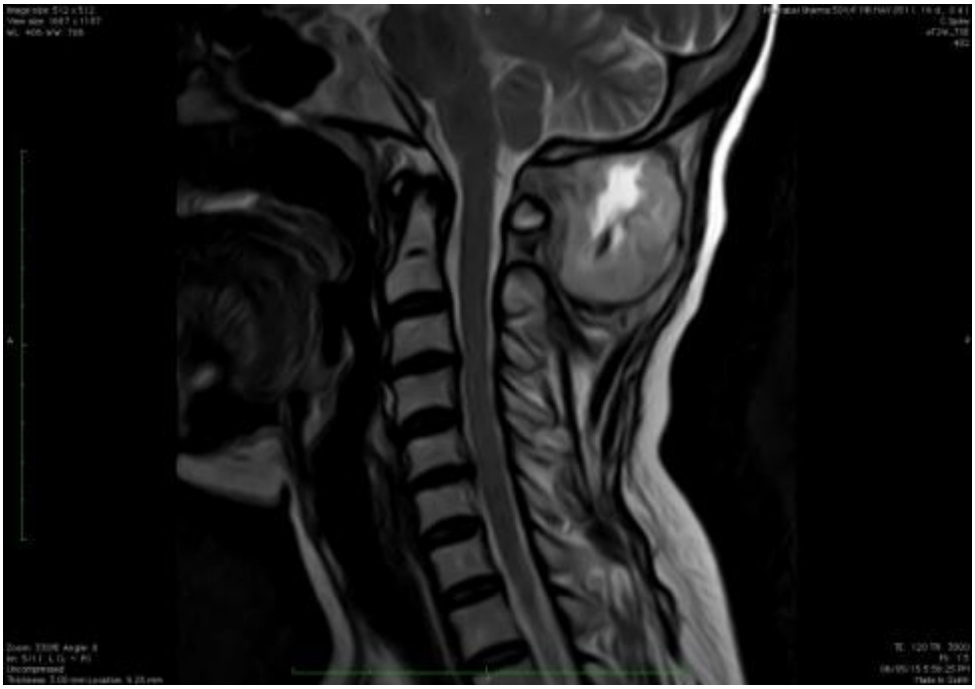


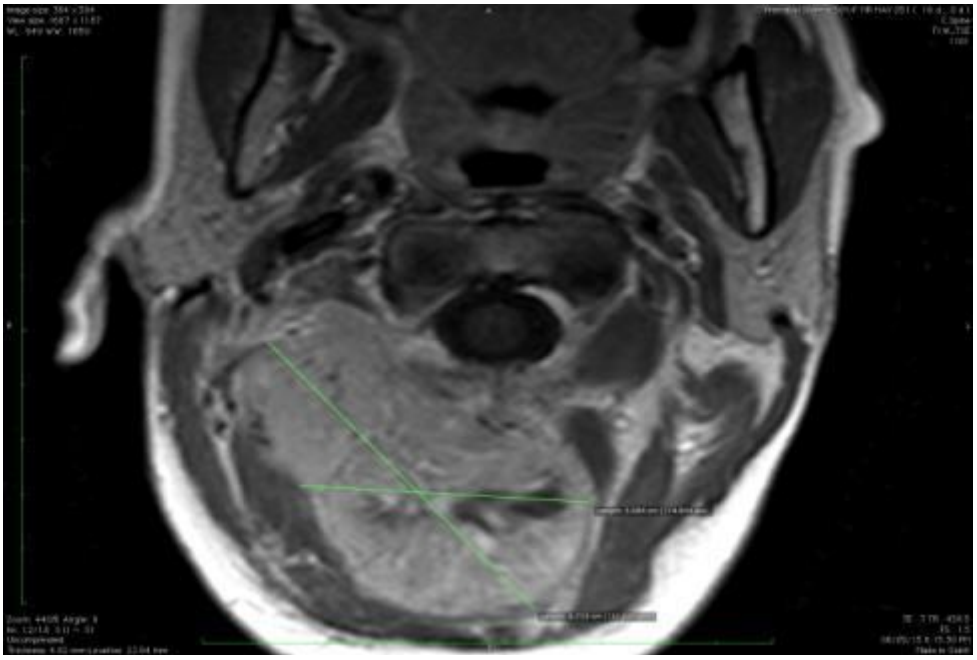


Figure 1 & 2: Showing Fusiform Shaped Growth over C1-C2 Level

DISCUSSION

MPNST is a very rare tumor, with an incidence of 1 per 1,00,000 population and which constitutes between 3 to 10% of all soft tissue sarcomas. Hence, this entity is often managed as a sub-category of soft tissue sarcomas (Hruban *et al.,* 1990). A combination of gross and microscopic findings along with immunohistochemical studies is commonly used to diagnose a case of MPNST (Dasgupta and Choudhuri, 1998). In most instances, the tumor display fascicles of spindle cells woven into herring bone pattern with varying degrees of mitosis and necrosis. However, it is not always possible to demonstrate the origin from a nerve, especially, when it arise from a small peripheral branch (Evans *et al.*, 2002). Still, there are

several other distinct features, such as proliferation of tumor in the sub-endothelial zones of vessels with neoplastic cells herniation into vessel lumen and proliferation of small vessels in the walls of the large vessels, which are very characteristic features of MPNST (Weiss and Goldblum, 2001).

The association of MPNSTs with neurofibromatosis is well known (Hirose *et al.*, 1998). Series reported 5% to 42% neurofibromatosis patient develop sarcomas.

Imaging is routinely performed to assess the extent of the disease and plan surgical resection. A target lesion in T2MR image is an indication of low grade while heterogeneous lesion due to necrosis and hemorrhage and patchy contrast enhancement in MRI is an indication of malignant MPNST (Friedrich *et al.*, 2005). MRI is the investigation of choice because it can reveal the nerve of origin and it's relationship to adjacent structures.

Radical surgical resection is the treatment of choice in MPNST.

MPNSTs are generally considered chemotherapy and radiotherapy resistant tumors.

MPNST has the highest recurrence rate of any sarcomas, and adequate initial treatment gives the best chance of survival.

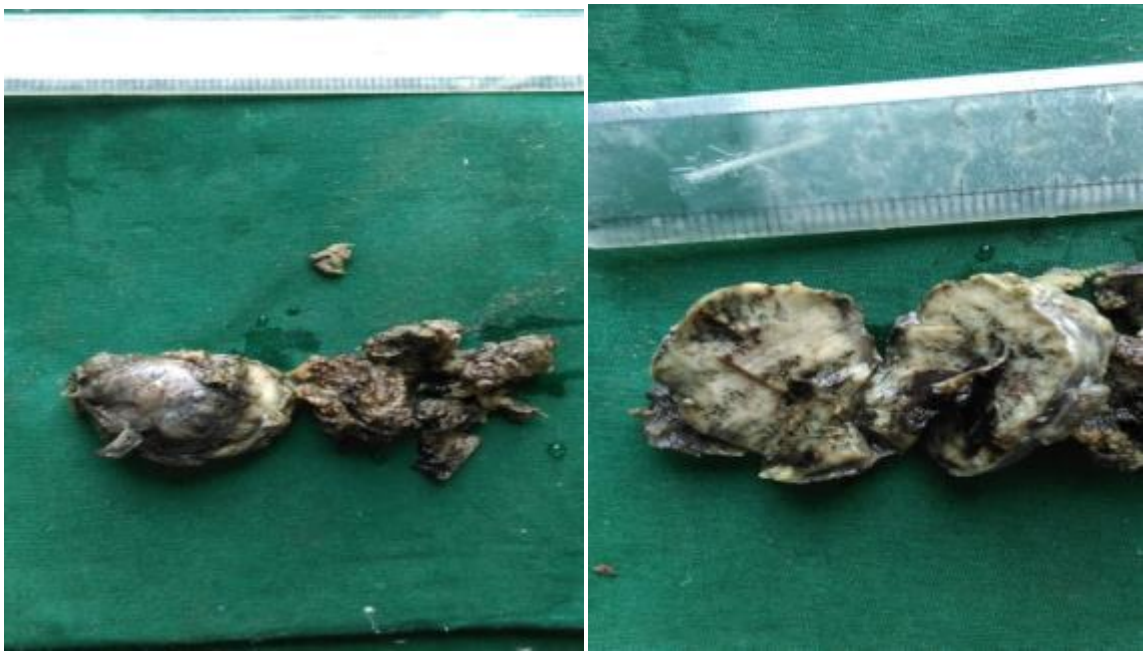
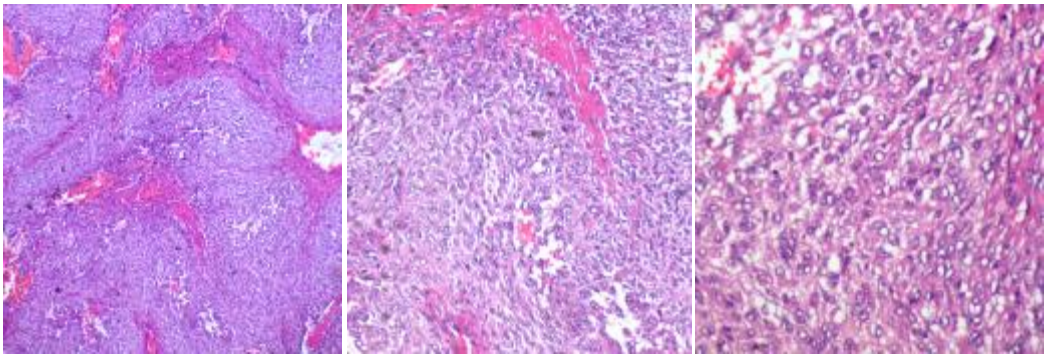
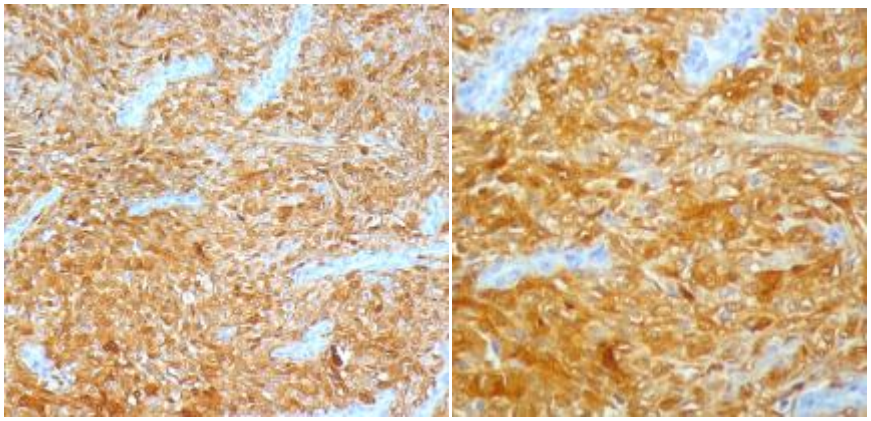


Figure 3 & 4: Showing Gross Appearance of Specimen



4x **10x** **40x**

Figure 5, 6 & 7: Showing Perivascular Concentration of Tumor Cells with Plumper Shaped Epitheloid Appearance of Endothelial Cells with Palisading Tumor Cells at the Edges of Irregularly Shaped Necrotic Areas and Abundant Mitosis



100x **200x**

Figure 8 & 9: Showing Spindle and Glandular Tumor Components were Positive for S-100

Conclusion

MPNST constitute a significant proportion of soft tissue sarcoma. A combination of clinical, pathological and immunohistochemistry helps in diagnosis of these tumors.

REFERENCES

Angelov L and Guha A (2000). Peripheral Nerver Tumors. In: Berstein M, Berger MS, editor, *Neuro Oncology Essentials*, **1**, (New York Theme Publishers, New York, USA) 434-444.

Brasfield RD and Das Gupta TK (1972). Von Recklinghausen's disease: A clinicopathological study. *Annals of Surgery* **175** 86-104.

Cashen DV, Parisien RC, Raskin K, Hornicek FJ, Gebhardt MC and Mankin HJ (2004). Survival data for patients with malignant schwannoma. *Clinical Orthopaedics and Related Research* **426** 69-73.

D'Agostino AN, Soule EH and Miller RH (1963). Sarcoma of the peripheral nerves & somatic soft tissues associated with multiple neurofibromatosis. *Cancer* **16** 1015-1027. doi:10.1002/1097-0124(196308)16:8<1015::AID-CNCR2820160808>3.0.CO;2-I.

Dasgupta TK and Choudhuri PK (1998). Tumors of soft tissue. *Connecticut Appleton & Lange* **2** 127-395.

Evans DG, Baser ME, McGaughran J, Sharif S, Howard E and Moran A (2002). Malignant peripheral nerve sheath tumors in neurofibromatosis 1. *Journal of Medical Genetics* **39** 311-314. doi:10.1136/jmg.39.5.311

Friedrich RE, Kluwe L, Funsterer C and Mautner VF (2005). Malignant peripheral nerve sheath tumors in neurofibromatosis type-1 diagnostic findings on MRI and mutational analysis of NF1 gene. *Anticancer Research* **25** 1699-1702.

Hirose T, Scheithauer BW and Sano T (1998). Perineural malignant peripheral nerve sheath tumors- A clinicopathologic, immunohistochemical and ultrastructural study of seven cases. *The American Journal of Surgical Pathology* **22** 1368-1378. doi:10.1097/00000478-199811000-00007

Hruban RH, Shiu MH, Senie RT and Woodruff JM (1990). Malignant peripheral nerve sheath tumors of buttock and lower extremity A study of 43 cases. *Cancer* **66** 1253-1265.doi:10.1002/1097-0142 (19900915)66:6<1253::AID-CNCR2820660627>3.0.C0;2-R.

Stout AP (1949). *Tumors of Peripheral Nervous System: Atlas of Tumor Pathology, Sect 2, Fasc 6*. (USA, Washington D.C, Armed Forces Institute of Pathogy).

Wanebo JE, Malik JM, VandenBerg SR, Wanebo JH, Driesen N and Persing JA (1993). Malignant peripheral nerve sheath tumors. A clinicopathologic study of 28 cases. *Cancer* **71** 1247-1253. Doi: 10.1002/1097-0142 (19930215)71:4<1247::AID-CNCR2820710413>3.0.C0;2-S.

Weiss SW and Goldblum JR (2001). *Soft Tissue Tumors*, **4**, (USA, St. Louis, Mosby Company) 1209-1263.