

Retroperitoneal Malignant Peripheral Nerve Sheath Tumour without Neurofibromatosis: A Case Report

Veeranathreddy V^b Jagadish B^a, Puranik G^a, Gopinath P^a, Chidananda KV^a, Ahalya reddy^c, Srikanth CH^b, Rahul M Sharma^b.

A, Professor, b, Post graduate, Department of General Surgery, c, post graduate, Department of Pathology
KVG Medical College & Hospital, Sullia-DK, Karnataka, India.

Abstract: MPNST often arises from a major peripheral nerve. MPNST replaced olden terms like malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma, malignant neurilemmoma. They can occur sporadically or in the context of neurofibromatosis type 1 (von Recklinghausen's disease). MPNST is an exception to sarcoma which will not arise from benign precursor. Common sites are major/proximal nerve trunks.

we came across one such patient who presents with mass per abdomen since 2 months, which was insidious in onset, initially 5x5cm, rapidly progressed to 13x12cm. On examination mass was retroperitoneal debulking of the tumour was done, Biopsy report suggestive of MPNST.

Nerve sheath tumours have varied clinical presentations, and a systematic approach, with judicious use of imaging, histopathology needed for diagnosis.

Keywords: MPNST, Retroperitoneal, Von Recklinghausen's disease.

I. Introduction

Peripheral nerve sheath tumours presents as soft tissue neoplasm's and include the spectrum of schwannomas, neurofibromas, neurofibromatosis, and malignant peripheral nerve sheath tumours. The presentation varies from asymptomatic soft tissue swellings to large masses that are recurrent and difficult to excise¹.

II. Case report

A 68 year old male from kodagu admitted with chief complaints of mass per abdomen since 2 months, which was insidious in onset, initially 5x5cm, rapidly progressed to 13x12cm, not associated with pain. No h/o jaundice, hematemesis, melena. No h/o loss of appetite or easy fatigability. No h/o loss of weight, urinary disturbances.

On examination of abdomen, a 15x12cm ovoid mass is present in epigastric and umbilical region extending across the midline up to the left lumbar, all borders were well made out, surface is smooth, firm in consistency, non tender, transmitted pulsations felt, does not move with respiration, non ballotable on bimanual palpation, fingers can be insunated below left costal margins, no renal angle fullness/tenderness, no hepatomegaly and splenomegaly. Mass dull to percuss. Auscultation bowel sounds heard.

On admission patient was haemodynamically stable. Routine blood tests are within normal limits. USG abdomen, Well defined homogenously hyperechoic mass lesion in the left upper quadrant of the abdomen displacing the spleen superiorly and the left kidney medially- Retroperitoneal mass lesion. CT abdomen, Well defined poorly circumscribed large heterogeneous retroperitoneal mass lesion with predominant fat component in the entire left quadrants of the abdomen arising from the left posterior pararenal space displacing the viscera as mentioned above infiltrating the left kidney-? Liposarcoma. Biopsy report suggestive of Malignant peripheral nerve sheath tumour.

Exploratory Laparotomy done. Intra operatively tumour is retroperitoneal in left side pushing the descending colon medially. Debulking of the tumour is done. Patient tolerated the surgery well. Post operative period was uneventful. Patient was discharged. Follow up after 15 days with histopathology, sections are densely cellular, tumour is composed of spindle cells arranged in fascicular pattern. The neoplasm display variable pleomorphism with some areas being highly pleomorphic (moderate to severe) with abnormal nuclear contours like buckle, giant and irregular nuclei while other areas appear relatively less pleomorphic (mild to moderate) and quite monotonous in appearance. Areas of necrosis and dense round cell inflammatory infiltrate are seen focally. Mitosis is a little sparse and variable from field to field. Nuclear palisading similar to that of neural neoplasms is observed in some of the sections. The background shows a fine fibrillary network. Suggestive of MPNST.



Figure 1 Gross specimen

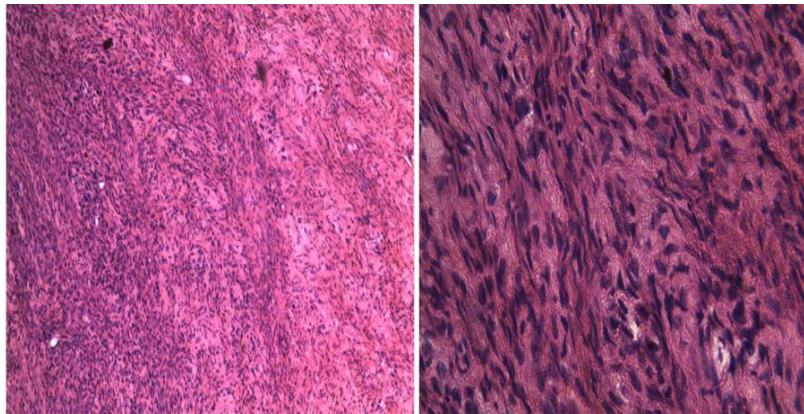


Figure 2 & 3 : Histological features are those of MPNST

III. Discussion

MPNST is defined as soft tissue tumor of putative Schwann cell lineage with anaplastic features in the form of high cellularity, cellular and nuclear pleomorphism, high mitotic rate and necrosis².

Malignant peripheral nerve sheath tumours (MPNSTs) are rare soft tissue sarcomas of ectomesenchymal origin, comprising 5% to 10% of all soft tissue sarcomas, and often arise in anatomically discernible peripheral nerves or from neurofibromas. 50% of MPNST s arises in patients with type 1 neurofibromatosis. This patient under study didn't have any stigmata of neurofibromatosis. MPNST s develops in deep seated large nerves³.

Incidence is 4.6% with neurofibromatosis and .001% in general population. Patients with neurofibromatosis develop sarcomas usually after a latent period of ten to twenty years. The mean age of patients with sporadic MPNST is 35.3 years and mean age of MPNST with neurofibromatosis is 28.7 years⁴.

MPNST are large tumours, usually more than 5 cm in diameter. The tumour grossly appears as a large fusiform mass arising from a major nerve. Cut surface of tumour is solid, fleshy, and tan to whitish with focal areas of hemorrhage and necrosis¹.

MPNST surgery is the treatment of choice. Wider margins should be obtained to limit locoregional failure, which eventually may reach the spinal cord. Chemotherapy is insensitive. Prognosis of patient with malignant MPNST is poor⁵. Five year survival is 54% in patients with tumours less than 5 cms in size as compared to large tumours, which have a survival of 16%⁴.

IV. Conclusion

Nerve sheath tumours have varied clinical presentations, and a systematic approach, with judicious use of imaging, histopathology needed for diagnosis.

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