International Journal of Science and Research (IJSR)
ISSN: 2319-7064
ResearchGate Impact Factor (2018): 0.28 | SJIF (2018): 7.426

A Rare Case Report on Malignant Peripheral Nerve Sheath Tumour in Thorax

Mulimani PN, Kadam SS

Abstract: ***Malignant Peripheral Nerve sheath tumour (MPNST) is defined as a malignant tumour arising from a peripheral nerve or showing nerve sheath differentiation, with the exception of tumours originating from epineurium or the peripheral nerve vasculature.We present an extremely rare case of (MPNST) in a 50-year-old woman without associated neurofibromatosis arising from thorax. Patient had come to the Emergency Department with complaints of shortness of breath since15 days, dry cough since 15 days and with an incidental history of chest trauma 1 month ago.Chest x ray showed a mass in the left hemi thorax,CECT of chest showed a large enhancing solid tumour measuring 11.5cm x 8cm x11cm.USG guided biopsyrevealed low grade spindle cell lesion with possible diagnosis of Myxofibroma, Schwannoma with focal degenerative changes and Reactive Myxofibromatous degenerative lesion.The patient underwent excision of mediastinal mass through left postero lateral thoracotomy under general anaesthesia. Histology of resected mass showed a biphasic neoplasm with a spindle cell component admixed with glandular elements, suggestive of Synovial sarcoma, malignant peripheral nerve sheath tumour in possible pre-existing Schwannoma.IHC report was suggestive of malignant peripheral nerve sheath tumour (MPNST).***

Keywords: MPNST, Solid tumour, Spindle cell, Biphasic Neoplasm, tumour size

1. Introduction

Malignant Peripheral Nerve sheath tumour (MPNST) is defined as a malignant tumour arising from a peripheral nerve or showing nerve sheath differentiation, with the exception of tumours originating from epineurium or the peripheral nerve vasculature [1, 2].These are also called as Neurogenic Sarcomas, Malignant Schwanommas and Neurofibrosarcomas. There is high risk of hematogenous metastasis and local recurrence. The incidence of MPNST arising in neurofibromatosis was 4.6% in the current series and 0.001% in the general clinic population[7].MPNST normally arise from extremities, in about 40-45% of the cases. These more commonly arise from lumbar and brachial plexus [3, 4]. We present a rare case of a 50 year old female presenting with breathlessness and incidental history of chest trauma, which on evaluation revealed mediastinal mass which was diagnosed as a nerve sheath tumour. The thorax is a very uncommon primary site for MPNST.

2. Case Report

A 50 year old female came to the Emergency Department with complaints of shortness of breath since15 days, dry cough since 15 days and with an incidental history of chest trauma1 month ago. Chest x ray showed a mass in the left hemi thorax, CECT of chest showed a large enhancing solid tumour measuring 11.5cm x 8cm x11cm. USG guided biopsy revealed low grade spindle cell lesion with possible diagnosis of Myxofibroma, Schwannoma with focal degenerative changes and Reactive Myxofibromatous degenerative lesion. A detailed Neurofibromatosis gene mutation analysis was not performed in the patient as she did not have any clinical features suggestive of neurofibromatosis and genetic workup is not done usually, as many of NF-1 gene mutations have very few phenotype-genotype correlations. The knowledge of sporadic MPNST with NF-1, doesn't affect treatment decision for an individual [2].

The patient underwent excision of mediastinal mass through left postero lateral thoracotomy under general anaesthesia. The mass was completely resected. Intra operatively a well encapsulated solid mass 11x 7 x 8.5 cm compressing the pulmonary artery and distal arch of aorta was seen.

Microscopically, multiple sections studied show well circumscribed, partially encapsulated lesion composed predominantly of bimodal population of cells distributed in hyper cellular and hypo cellular areas. Cells are bimodal in morphology which consists predominantly of oval to spindle cells which are arranged in hypo cellular sheets interspersed with focal dense lobulated, hyper cellular nests. Focal areas show poorly formed glandular structures surrounded by round to ovoid cells.

Histology of resected mass showed a biphasic neoplasm with a spindle cell component admixed with glandular elements, suggestive of Synovial sarcoma, malignant peripheral nerve sheath tumour in possible pre-existing Schwannoma. IHC showed, positive for S100, focally positive for sma (smooth muscle actin) and negative for Desmond H caldesmon. The report was suggestive of malignant peripheral nerve sheath tumour (MPNST). The patient made an uneventful recovery and was referred to the regional cancer centre for further management.

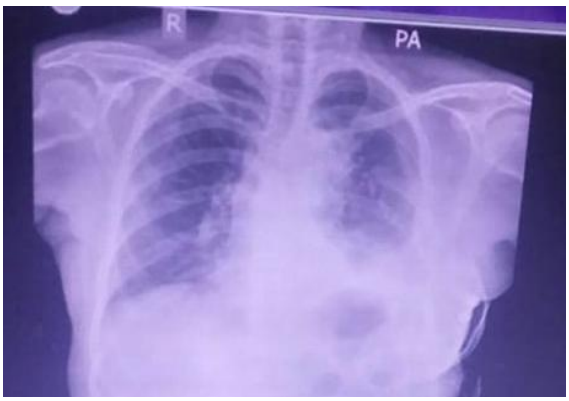

Figure 1

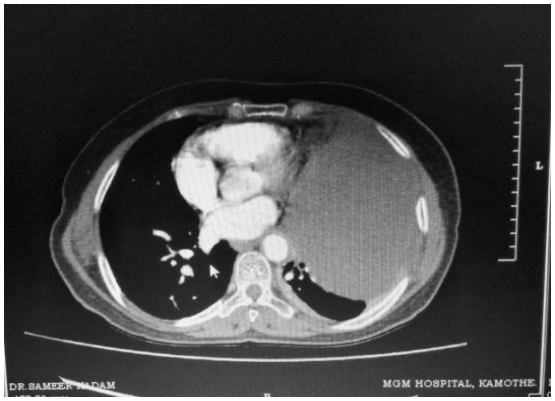

Figure 2
Chest X-ray revealed deviation of trachea with homogenous opacity in the left side chest.

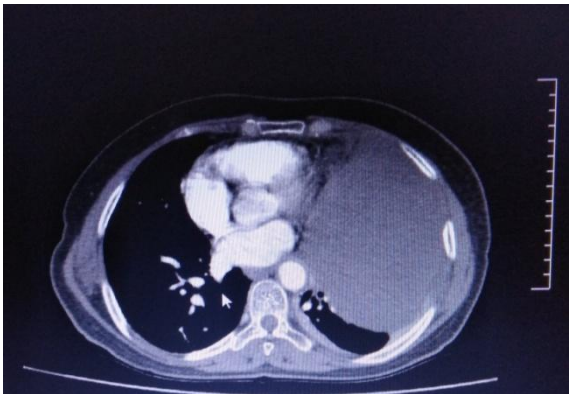

Figure 3

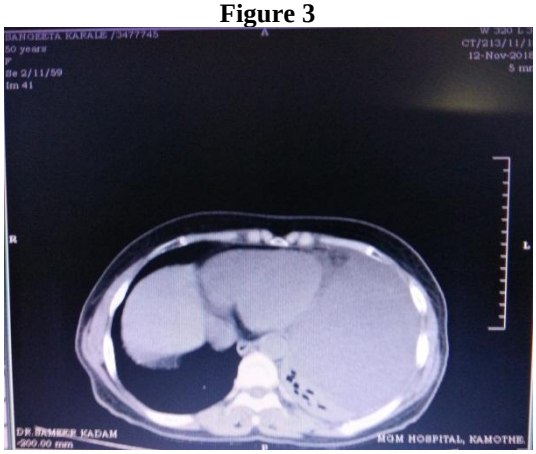

Figure 4
CT (Computed Tomography) of chest revealed enhancing mass in the left hemithorax measuring 11x 7 x 8.5 cm with compression over pulmonary artery and distal arch of aorta.

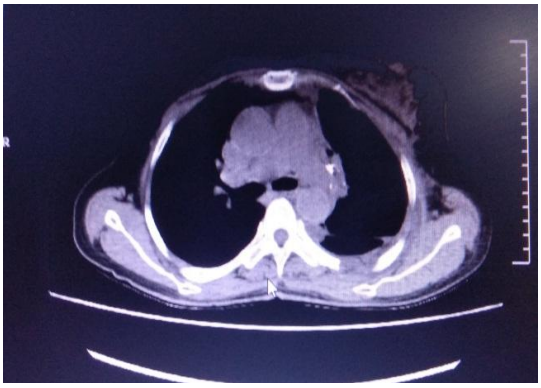

Figure 5

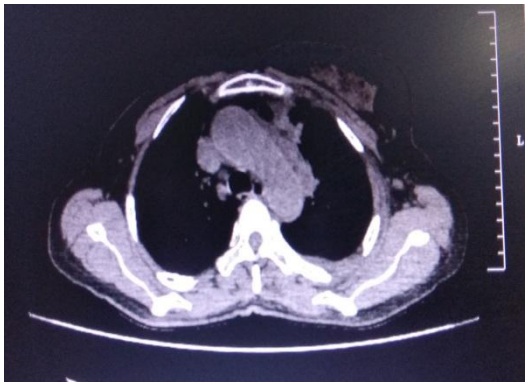

Figure 6
CT chest- Post operative

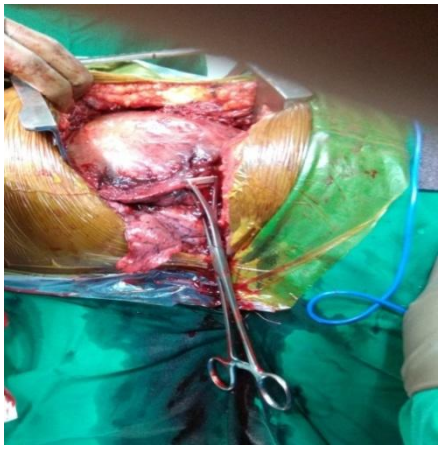

Figure 7

Figure 8

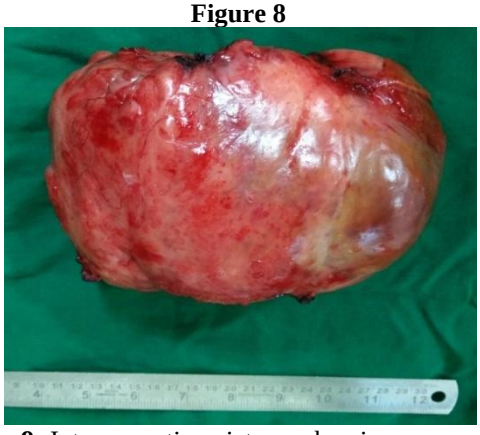

Figure 9: Intra operative pictures showing encapsulated mass in the left side of thoracic cavity with mass excised in toto.

3. Discussion

Malignant Peripheral Nerve sheath tumour (MPNST) incidence increases if associated with risk factors such as Neurofibromatosis (NF) -1, and history of previous radiation exposure. The diagnosis of NF-1 is based on clinical criteria including café-au-lait macules, axillary and inguinal freckling, and presence NF related tumours such as dermal neurofibromas[2]. The length of time required between exposure to radiation and origin of new sarcoma has been modified by most investigators. The origin of new sarcoma in radiation exposed area can be secondary to radiation induced or primary sarcoma originating at the time. The latent period of exposure required for development of radiation induced sarcoma is modified by many investigators. A minimum of 1 month has been suggested by some authors [2].

Like other soft tissue sarcomas, MPNSTs are most commonly staged using the American Joint Committee on Cancer staging system , which includes tumour grade (1–4), tumor size (≤ 5 cm or ≥ 5 cm), location (superficial versus deep), and the presence of distant metastases at the time of diagnosis. Large tumour (> 5 cm), deep location, and the presence of distant metastases are associated with poorOutcome [2].

MPNST can occur sporadically or in association with NF-1. Sporadic MPNST usually present in between 3rd to 6th decade, whereas when associated with NF-1, they present as early as between 1st to 2nd decade. Early diagnosis of MPNST is crucial as it is aggressive and has more chances of regional and distant metastasis[6]. Therefore early diagnosis and complete surgical resection has shown to be curative. MPNST with NF-1 association does not affect the treatment decision and survival difference in sporadic variety.

Patient with NF-1 gene mutation can manifest with superficial and deep seated lesions. Though most of lesions are benign, there might be high chance of having malignant foci within it. PET is useful in differentiating benign from MPNST [6]. S‑100 protein negative status has been associated with lower survival andhigher recurrence rate [5].

Complete resection of the tumour with clear margin is considered to be treatment of choice. Adjuvant radiation therapy can be added if tumour is intermediate to high grade. Chemotherapy can be used in advanced cases. Drugs preferred are doxorubicin, ifosamide. The overall response to chemotherapy is marginal [6].

4. Conclusion

Occurrence of MPNST tumours in the thoracic cavity without neurofibroma association is very less. Thoracic cavity being the primary site of origin is also a rare entity. Awareness and early diagnosis of this rare entity result in complete surgical excisionand better prognosis. A wide range of neoplasms, both primary and metastatic, occur in the mediastinum, which pose considerable diagnostic difficulties.

Malignant peripheral nerve sheath tumour should always be considered in the differential diagnosis. This study highlights the importance of recognizing an unusual presentation of this aggressive neoplasm to aid appropriate clinical management.

5. Conflict of Interest

The authors declare that they have no conflict of interest

References

[1] Kleihues P, Louis DN, Scheithauer BW, Rorke LB, Reifenberger G, Burger PC, *etal.*The WHO classification of tumors of the nervous system. J NeuropatholExpNeurol 2002;61:215-25.
[2] Widemann BC. Current status of sporadic and neurofibromatosis type 1associated malignant peripheral nerve sheath tumors.CurrOncol Rep 2009;11:322-8.
[3] Cha C, Antonescu CR, Quan ML, Maru S, Brennan MF. Long-term results with resection of radiation-induced soft tissue sarcomas. Ann Surg 2004;239:903-9.
[4] Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. Cancer 1986;57:2006-21.
[5] Wang T, Yin H, Han S, Yang X, Wang J, Huang Q, *et al.* Malignant peripheral nerve sheath tumor (MPNST) in the spine: A retrospective analysis of clinical and molecular prognostic factors. J Neurooncol 2015;122:349-55.
[6] Sundaram S S, Sachdeva A K, Penumadu P, Srinivas B. Malignant peripheralnerve sheath tumour of the thorax: Unusual location of a rare tumour. Cancerjournal.net on Sunday, October 20, 2019, IP.103.24.63.186.
[7] Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. Cancer 1986;57:2006-21.