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Case of Malignant Peripheral Nerve Sheath Tumor (MPNST) of Lung

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ABSTRACT

Malignant peripheral nerve sheath tumor (MPNST) is very unusual type of soft tissue sarcoma (STS) and occurs mostly in patients of neurofibromatosis. The occurrence of MPNST in lung is particularly rare. Only very rarely cases have been reported till date in absence of history of neurofibromatosis. We are reporting such case in a 34 year old female who presented with complains of chest pain, breathlessness and nonproductive cough since 3 months. Microscopic examination revealed a malignant spindle cell tumor of low grade histology. Immunohistochemistry (IHC) was performed which was positive for vimentin and actin and negative for desmin and S- 100. Final diagnosis of MPNST was made on the basis of morphological appearance and IHC. Patient was treated with neoadjuvant chemotherapy ifosfamide and adriamycin for four cycles, followed by surgical excision. At present patient is on regular follow up for one year post treatment.

KEYWORDS: lung cancer; Malignant peripheral nerve sheath tumors.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNST) are an uncommon and the incidence of MPNSTs in the common population is 0.001 percent. MPNSTs, which are classified as malignant soft tissue sarcomas, can arise from preexisting plexiform neurofibromas or perineuriomas, or normal nerves. They do not arise from schwannomas. MPNSTs also occur as secondary neoplasm 10 to 20 years after radiation therapy, accounting for up to 10 percent of MPNSTs [1]. From 22 to 50 percent of MPNSTs occur in patients with neurofibromatosis type 1 (NF1), the rest being sporadic. [2] The risk of developing a MPNST in a patient with NF1 is between 8 and 13 percent. MPNSTs tend to present at an earlier age in patients with NF1 (third or fourth decade of life, versus seventh decade in patients who do not have NF1). MPNSTs are most commonly found on the extremities and trunk, and less often on the head and neck [5]. Only a few cases have been documented in the world literature. We report a case of MPNST occurring in lung in a 34 years old female patient without any evidence of NF1 syndrome.

CASE SUMMARY

A 34 year-old nonsmoker female without any previous remarkable medical history presented with complains of chest pain, exertional breathlessness and nonproductive cough since 3 months. On physical examination there was no supraclavicular lymphadenopathy and physical examination did not reveal any signs of neurofibromatosis. The chest x ray revealed homogenous opacity in right upper and middle lobe and moderate right sided pleural effusion [Figure 1]. Pleural fluid cytology was suggestive of mesothelial cells and inflammatory cells without any malignant cells. Contrast enhanced CT Scan of thorax was done which showed a soft tissue density lesion in right upper lobe and middle lobe with internal necrotic area, involving mediastinal and costal pleuras and compressing right main bronchus as well as

trachea, suggesting a picture of malignant mass [Figure 2]. CT guided biopsy of lung mass revealed spindle cell proliferation with small epithelial looking areas having cells with scanty cytoplasm, vesicular nuclei and small inconspicuous nucleoli. Spindle cells showed mild to moderate pleomorphism, occasional mitosis, these features were suggestive of spindle cell tumor. Immunohistochemistry revealed positive staining for vimentin, actin and negative for desmin and S 100 [Figures 3-8]. From all these findings, diagnosis of malignant peripheral nerve sheath tumors was made. CT scan of the abdomen showed no evidence of metastatic disease. Patient was treated with neoadjuvant chemotherapy; ifosfamide and adriamycin for four cycles, as it was the operating surgeon's view that it would make the surgery less extensive and less morbid. The patient underwent surgical excision of mass and the whole specimen was submitted for histopathological examination, which revealed the same histopathological features as in biopsy specimen. Additionally, the surgical margins were free of tumor cells, but the margins were close, being less than 1 cm. The patient was referred for postoperative radiation therapy; patient was treated with postoperative radiotherapy in dose of 50 Gy in 25 fractions in view of close margins by radiotherapist. At present patient is on regular follow up and disease free after 26 months.

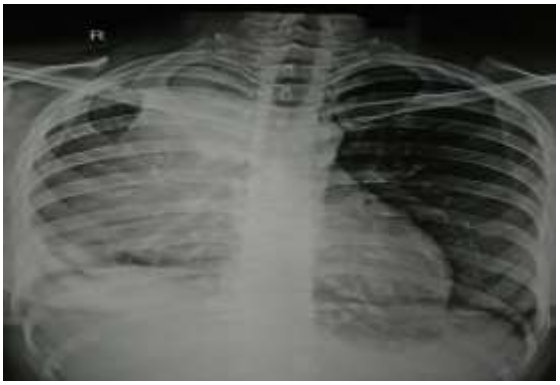

Figure 1: the chest x ray revealed homogenous opacity in right upper and middle lobe and moderate right sided pleural effusion.

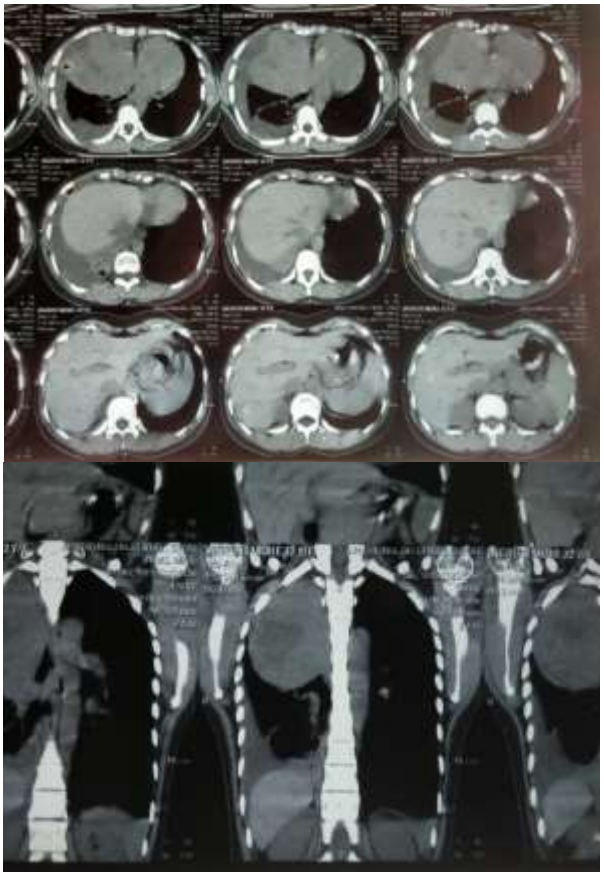

Figure 2: Contrast enhanced CT scan of thorax was done which showed a soft tissue density lesion in right upper lobe and middle lobe with internal necrotic area, involving mediastinal and costal pleuras and compressing right main bronchus as well as trachea, suggesting a picture of malignant mass.

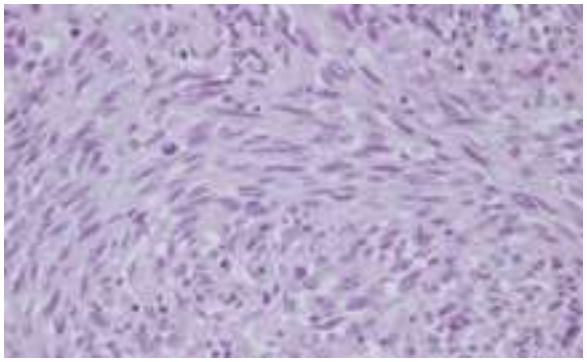

Figure 3: Proliferation of mesenchymal neoplastic spindle cells densely packed, arranged in short bundles (H&E, ×20).

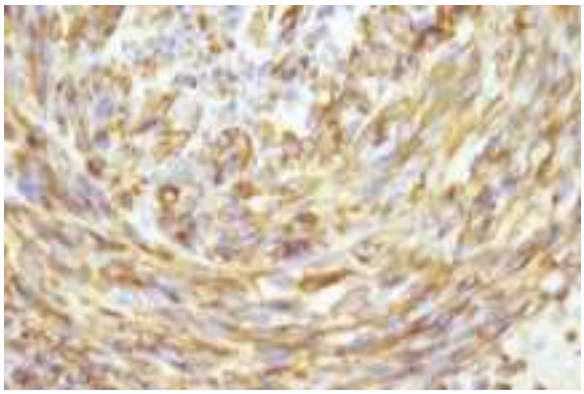

Figure 4: Strong and uniform positivity for vimentin (IHC, ×40).

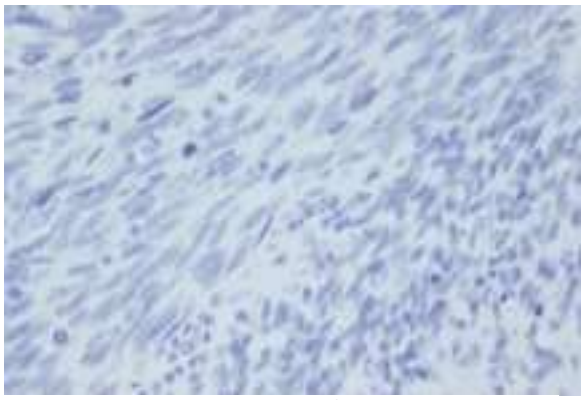

Figure 5: Negative immunostaining for low molecular weight keratin (panCK, ×40).

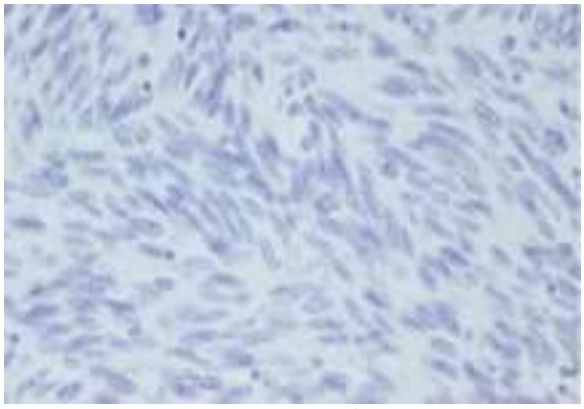

Figure 6: Negative immunostaining for desmin (IHC, ×40).

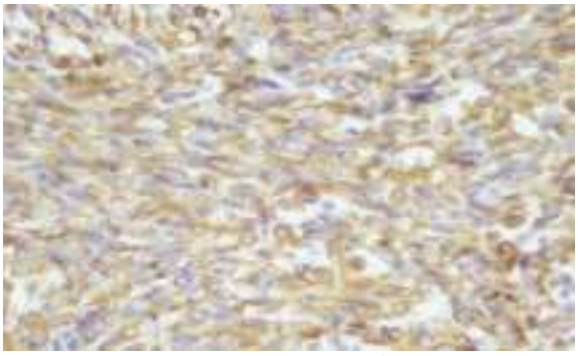

Figure 7: Low positive immunostaining for actin (IHC, ×40).

Figure 8: Negative cytoplasmatic immunostaining for S-100 (IHC, ×40).

DISCUSSION

MPNSTs are rare and highly aggressive soft-tissue sarcomas and they are associated with neurofibromas in NF-1 patients but they can also occur in association with radiation or sporadically. MPNST frequently arises from pre-existing plexiform neurofibromas [7]. In patients with NF1, the presence of an internal plexiform neurofibroma is associated with a 20-fold increased risk of developing a MPNST compared with the risk in those lacking an internal plexiform neurofibroma [7]. MPNST in patient with NF-1 usually present with multiple rapidly enlarging masses or a new onset of pain associated with pre-existing plexiform neurofibromas. Sporadic forms of MPNST present with a new solitary, enlarging and painless mass. [7]

MPNSTs are a tumor of adult age group occurring in patients 20-50 years of age. [3]The most common sites of occurrence of MPNSTs are proximal portions of trunk (46%), upper and lower extremities (34%) and head & neck region (19%). Though these tumors due to their Schwann cell

origin may occur anywhere near a nerve trunk, lung is a rare site of its occurrence.[3] By gross inspection, MPNSTs tend to be large, firm tumors, containing areas of necrosis and hemorrhage. Microscopically, most MPNSTs are highly cellular, comprised of spindle cells reminiscent of Schwann cells. The cells are mitotically active. The diagnosis of MPNST can only be done on the basis of immunohistochemistry otherwise it can be easily mistaken for fibrosarcoma and leiomyosarcoma. The cells are weakly S100 positive, consistent with dedifferentiation from Schwann cells. Variations include malignant triton tumor (malignant peripheral nerve sheath tumor with rhabdomyosarcomatous differentiation), a highly malignant tumor containing embryonic striated muscle components [8]. MPNSTs may also stain positive for neuron-specific enolase; and other proteins, such as actin, cytokeratin (CK), smooth muscle actin (SMA), desmin and vimentin, to differentiate from other spindle cell sarcomas. [9]

Even with aggressive surgical, radiation chemotherapy treatment, the prognosis is not good. Poor prognostic signs include tumors exceeding 5 cm in size, higher tumor grade, association with neurofibromatosis type 1 (NF1), older age, distant metastases at the time of diagnosis, and inability to achieve tumor-free margins.[2,10] Tumor size, grade, location, presence of neurofibromatosis type 1, local recurrence, and adjuvant chemotherapy are all associated with Disease specific survival. [10] Complete surgical resection is the treatment of choice for MPNSTs and provides the only hope for cure. MPNSTs are considered to be chemotherapy- and radiotherapy-resistant tumors. However, postoperative radiotherapy has a definite role in both disease-free and overall survival and is currently recommended by an oncology consensus group.[6] Chemotherapy has role in advanced disease in which combination of doxorubicin and ifosfamide has better response than single agent therapy with either drug. [4] Despite multimodal therapy, including aggressive surgical resection and adjuvant radiotherapy, the prognosis remains poor.

CONCLUSION

MPNST of lung is a very rare tumor and should be considered in differential diagnosis of lung mass. The best possible therapy is surgical excision with clear margins, and neoadjuvant chemotherapy as well as postoperative radiotherapy can be used as per the clinical scenario.

Learning points

1. MPNST in patients without history of neurofibromatosis is very rare.
2. Lung is rarest location for MPNSTs.
3. Surgical excision is the best treatment.
4. Postoperative radiotherapy has role in certain cases when indicated.
5. Neoadjuvant chemotherapy can be used if needed, based on chemotherapy regimens used in soft tissue sarcomas.

Declaration by Authors

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