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Malignant Peripheral Nerve Sheath Tumor in Mediastinum With Metastases - A Case Report

KEYWORDS: MPNST (Malignant peripheral nerve sheath tumor), Mediastinum, chest wall metastases.

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ABSTRACT *Malignant peripheral nerve sheathtumors(MPNST) are rare soft tissue tumors arising from peripheral nerves or from malignant transformation of pre-existing neurofibroma. The common sites of involvement are head, neck, extremities and trunk.We report a case of MPNST arising from mediastinum in a 45yr old male patient presenting with simultaneous chest wall metastases.*

INTRODUCTION

The group of peripheral nerve sheath tumors include Neurilemmomas, Neurofibromas, Malignant Schwannomas. Malignant schwannomas now renamed as Malignant peripheral nerve sheath tumors(MPNST) occur with equal incidence in male and female patients. They are usually associated with Type-1 Neurofibromatosis [1]. Clinically they are slow growing tumors and become painful on malignant transformation. MPNST constitute 15% of nerve sheath tumors. On CT scan the tumors are large, infiltrating, irregular and inhomogenous in nature[2]. On MR imaging the tumors show signal intensity equal to that of muscle on T1W, hyperintense signal on T2W and with heterogenous enhancement on contrast administration and invasion into adjacent structures.

CASE REPORT

A 45 yr old male was admitted with painful mass in right antero-lateral chest wall of one month duration. There was no previous history of trauma. The mass was measuring 10x8cms, non pulsatile and associated with restriction of shoulder movements. There were no other specific symptoms related to chest. Routine laboratory investigations were within normal limits. Chest radiograph showed a well defined mass in right apical region with erosion of right first and third ribs with associated soft tissue mass in right axilla(figure 1). Ultrasound chest revealed a well defined solid mass in right infraclavicular region and axilla with increased vascularity. On CECT there was a large heterogenously enhancing mass lesion with central necrosis noted in the right apical region. There was associated destruction of right first and third ribs with extrathoracic soft tissue mass(figure2). There was no evidence of mediastinal lymphadenopathy or pleural effusion, heart and lungs were normal. MRI Chest showed a large lobulated mass heterogenously hypointense on T1W and heterogenously hyperintense on T2W, STIR sequences with restriction on DWI, the mass is in close proximity to right subclavian vessels(figures 3a,3b). A provisional diagnosis of mediastinal mass causing rib erosion was made on imaging studies. Right antero-lateral thoracotomy was performed which showed a large multilobulated mass of 10x8x8cms in upper chest which was mainly solid and few cystic areas which were filled with clots and necrotic material. The mass was adhered to the upper lobe of lung, pericardium and debulking of the mass was done.

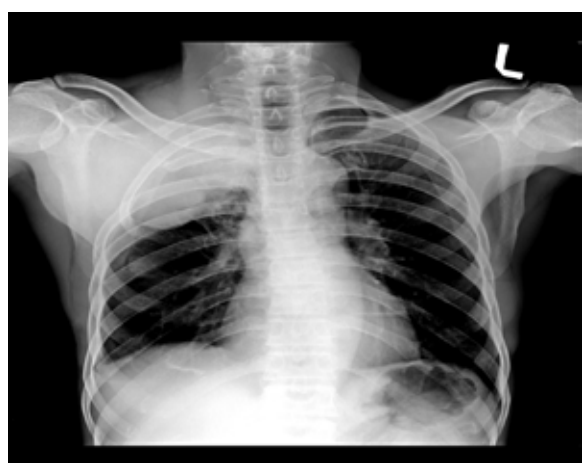

Figure 1: chest radiograph showing soft tissue mass in right upper chest with rib erosions.

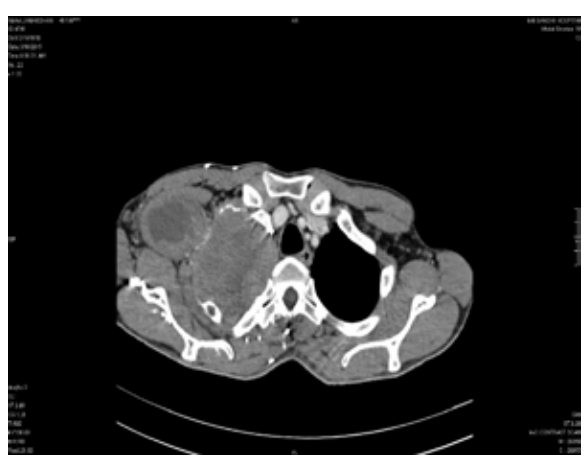

Figure 2: contrast CT chest showing minimally enhancing mass in the right thoracic cavity with erosion of ribs and associated extrathoracic mass.

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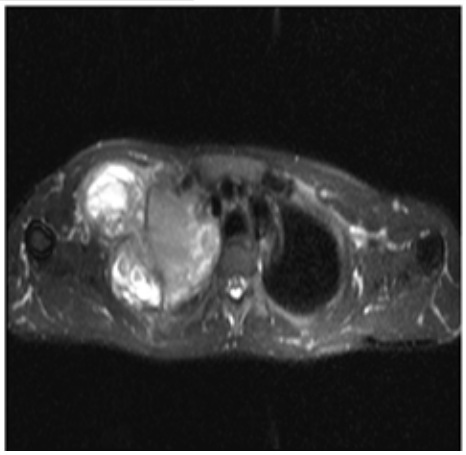

Figure 3a

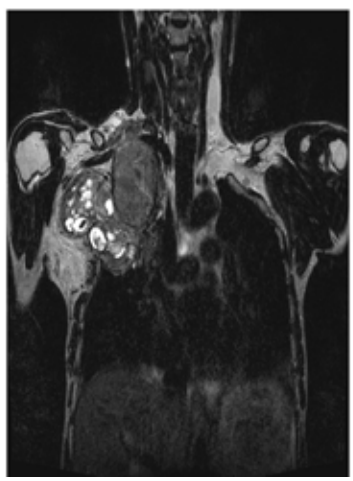

Figure 3b

Figure 3a & 3b: MRI Thorax in axial and coronal planes showing well defined lobulated mass in the right upper thorax with heterogenous signal intensity.

HPE from the mediastinal mass and chest wall mass showed cellular tumor tissue interspersed with haemangiopericytoma like vessels along with hyalinization. The tumor cells are arranged in fascicles, sheets and pseudoglandular pattern. Individual cells are spindle to polygonal in shape with high nuclear/cytoplasmic ratio, some of the cells showed vacuolated cytoplasm. Mast cells along with haemosiderin laden macrophages are also seen(figure 4). The above features are suggestive of malignant spindle cell poorly differentiated mesenchymal neoplasm. IHC showed positive for S100 and negative for CK and Desmin. The final diagnosis was Malignant peripheral nerve sheath tumor.

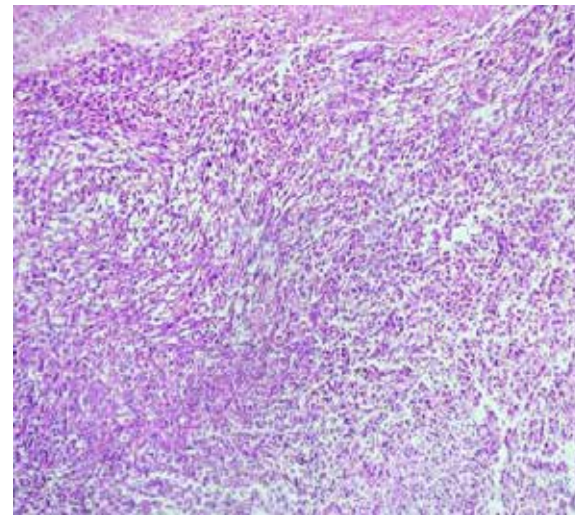

Figure 4: High power microscopic examination showing sheets of spindle to polygonal cells with high nuclear/ cytoplasmic ratio.

DISCUSSION

MPNST are sarcomas arising from a peripheral nerve or a Neurofibroma which are called as Malignant Schwannomas or Neurofibrosarcomas previously. It can occur spontaneously or in association with Neurofibromatosis-1[3] or following radiation therapy. They will occur in adults in the age group of 20-50 years. The clinical features include radiating type of pain, paraesthesia and motor weakness. On CT these tumors appear as large irregular infiltrating masses with bone destruction, with central heterogenous density due to necrosis or haemorrhage. Sudden increase in the size or alteration in the attenuation of a preexisting benign mass suggesting malignant degeneration. On MR they appear as a large invasive mass of variable signal intensity on T1W and hyperintense on T2W and have fusiform shape with the nerve roots entering and exiting the nerve[4]. The tumors show heterogenous enhancement on contrast.

Neurogenic tumors account for about 9% of all the primary mediastinal masses in adults and about 95% of them occur in posterior mediastinum of which malignant tumors are more frequent in children(61.5%) compared to adults(8.5%). MPNST metastasize to lungs mostly, followed by bone and pleura[3]. FDG-PET reliably evaluates the presence of metastases or recurrence of the primary tumor. The American joint committee on cancer staging system for soft tissue sarcomas is planing an important method for the appropriate treatment of these tumors.

Fumihiro shoji et al reported a case of malignant schwannoma arising from vagus nerve presenting as upper mediastinal mass[5]. MPNST developing in association with Neurofibromatosis-1 is well established in literature[6,7] and also cases have been reported without any association with Neurofibromatosis-1[4].

Sophia et al reviewed the imaging findings in 15 pathologically proven cases of MPNST and concluded that the tumors are typically elongated in the direction of nerves and the presence of invasion required neoadjuvant or adjuvant chemoradiation[8].

Muret Oncel et al retrospectively reviewed the clinicoradiological features of neurogenic tumors of the mediastinum

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and found one case of MPNST out of 13 cases which were surgically treated[9].

Isolated case of MPNST of the chest wall associated with Neurofibromatosis-1 has been reported[10,11]. Satoshi kozeuka et al reported a case of MPNST from phrenic nerve which presented as a right anterior mediastinal mass[12].

Takeshi kawaguchi et al foundno significant differences in the clinical features of both primary intrathoracic malignant neurogenic tumors in comparison with benign neurogenic tumors[13].

In the present case the patient presented with a painful chest wall mass and there were no symptoms attributed to mediastinal involvement. Concurrent presentation of mediastinal mass with chest wall metastases is considered quite unusual. About 80-85% of MPNST are spindle cell tumors with features similar to fibrosarcoma [14]. S-100 is identified in 50-90% of MPNST. Leu7 and myelin protein are noted in 50% and 40% of cases respectively[3]. Wide surgical excision along with radiation therapy or chemotherapy has improved patient care and survival rate.

CONCLUSION

MPNST is one of the important neurogenic tumor of mediastinum. It can present as metastases simultaneously along with primary tumor.

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