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Case report

A RARE CASE OF ENDOBRONCHIAL MALIGNANT PERIPHERAL NERVE SHEATH TUMOR

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ABSTRACT

Malignant schwannoma also called Malignant peripheral nerve sheath tumor (MPNST), is a rare and aggressive sarcoma that arises from the nerve sheath, with an incidence of 0.001%. It usually arises from peripheral nerves or somatic soft tissues. We describe a rare case of endobronchial Malignant peripheral nerve sheath tumor in a middle aged female who presented with right middle and lower lobe collapse. The diagnosis was made from the history, clinical examination, radiological investigations and bronchoscopy. The patient was subjected to pneumonectomy and tracheoplasty.

***Keywords**: Malignant schwannoma, Malignant peripheral nerve sheath tumor, Bronchial, Bronchoscopy.*

INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) is a rare variety of soft tissue sarcoma.[1,2] World Health organization (WHO) committee for the classification of soft tissue tumors standardised malignant peripheral nerve sheath tumor (MPNST) as the accepted nomenclature for a spindle cell sarcoma arising from nerve, or neurofibroma, or demonstrating nerve tissue differentiation.[3,4] They are difficult to diagnose because of their cellular origin and histopathological similarities with other spindle cell sarcomas like monophasic synovial sarcoma, leiomyosarcoma and fibrosarcoma.[5] Surgical procedures have been the main stay of treatment in such tumors. We present a case of Malignant peripheral nerve sheath tumor in the right main bronchus. The patient underwent right pneumonectomy with tracheoplasty.

CASE REPORT

A 35 year old female was admitted in the chest ward at Meenakshi Medical College And Research Institute with complaints of breathlessness and cough for 1 year. Breathlessness was present for a period of 1 year with grade 1 MRC (Medical research council) which had progressively increased to grade 2 MRC. Patient complained of cough with scanty, mucoid expectoration with no diurnal or postural variation. .She also had dull aching right sided chest pain. There was no H/O hemoptysis, wheeze, orthopnea or paroxysomal nocturnal dyspnoea. She also had weight loss. There was No H/o of ATT (Anti tuberculous treatment) intake in the past or any significant medical illness. There were no significant abnormal findings on general examination. Chest examination showed tracheal deviation to right side, dull note on percussion and absent breath sounds in the right axillary, interscapular and infrascapular areas. Other systemic examination was normal. **Investigations:** Routine haematological and biochemical parameters were normal. Mantoux test and serology for human immunodeficiency virus were negative. Sputum examination for AFB (Acid Fast Bacilli) smear and culture was negative. Chest radiograph (Figure 1), showed tracheal deviation to right side with collapse of right middle and lower lobes. CT chest with contrast showed a mass measuring 3.5 x 3.5 cm in right main bronchus obstructing the lumen with collapse of the right middle and lower lobes and mediastinal lymph nodes.

Spirometry showed restrictive pattern. A flexible video bronchoscopy was done which showed an endobronchial mass lesion at the distal end of trachea , obstructing the right main bronchus. Bronchoscope could not be passed further beyond. Biopsy was taken from the endobronchial mass (Figure 2) along with bronchial wash. Bronchial wash was negative for malignant cells. Bronchial biopsy showed features of spindle cell neoplasm with myxoid changes. In view of histopathological findings, patient was referred to a cardiothoracic surgeon and she underwent right pneumonectomy with tracheoplasty. Histopathological examination of the surgical specimen revealed a malignant spindle cell neoplasm composed of spindle cells (Figure 3 & 4) with elongated pleomorphic nuclei, numerous mitosis arranged in fascicles and storiform pattern with hypocellular and hypercellular areas. Immunohistochemistry for S 100 showed spindle cells positive (Figure 5).

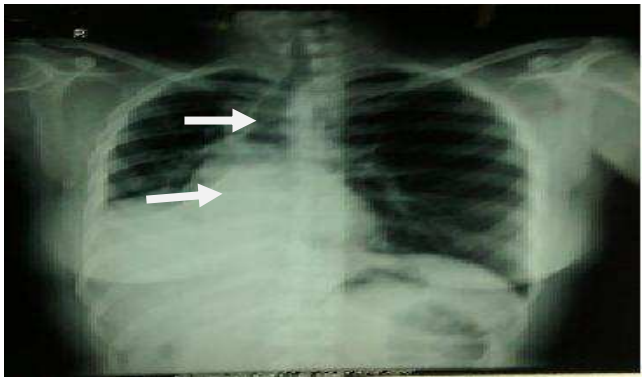

Fig 1: X- ray Chest PA view showing tracheal deviation to right with collapse of the right middle and lower lobe.

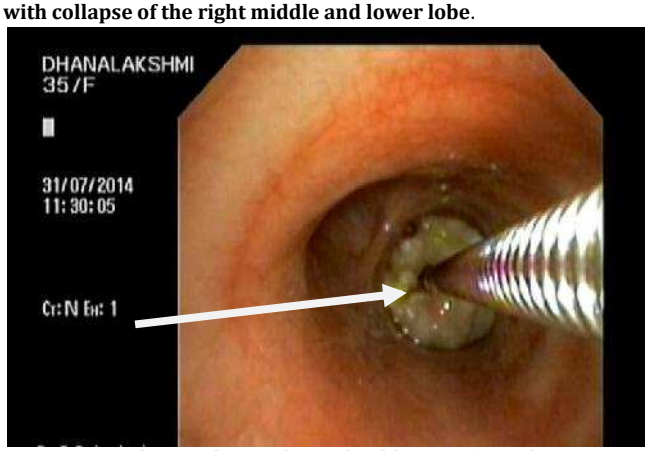

Fig 2: Biopsy being obtained via Flexible FOB from the mass in the Right main bronchus.

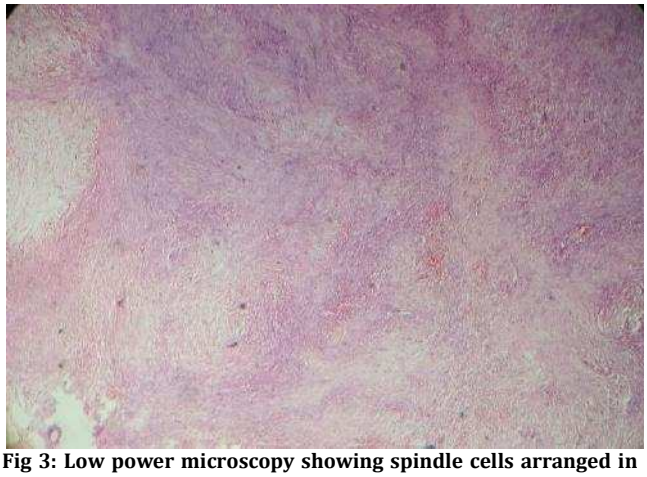

Fig 3: Low power microscopy showing spindle cells arranged in hypo and hyper cellular areas.

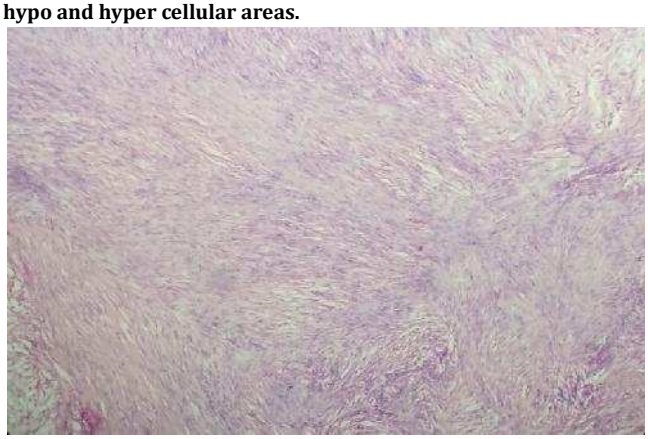

Fig 4: High power microscopy showing spindle cells having wavy nuclei suggestive of neural origin.

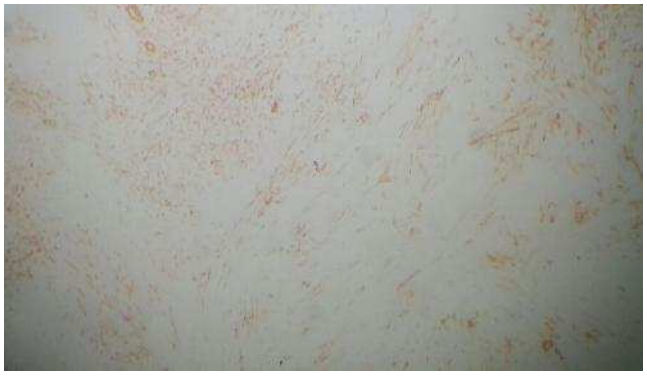

Fig 5: 100X Immunohistochemistry shows spindle cells positive.

DISCUSSION

Malignant peripheral nerve sheath tumors (MPNST) are sarcoma arising from a nerve or benign nerve sheath tumor or showing nerve sheath cellular differentiation. It has an incidence of 0.001% in the general population and 0.16% in patients with neurofibromatosis .[6,7] Most malignant schwannomas are highly invasive and are associated with a low survival rate.[8] MPNST affects adult patients from 20 to 50 years of age with slight female predilection.[9,10] The features of MPNST include 1) dense and hypodense fascicles alternating in a marble-like pattern consisting of 2) asymmetrically tapered spindled cells with irregular buckled nuclei or 3) immunohistochemical or electron microscopic evidence of Schwann cell differentiation. The spindle cell MPNST forms 80%, with features of long fascicles of closely spaced hyperchromatic spindle cells, small round blue cells, pleomorphic cells and extensive necrosis with perivascular preservation. Ancillary test like Immunohistochemistry with S100 protein is positive in about 60% and is usually focal. Successful treatment of MPNST requires complete surgical excision. [11,12] A good three-dimensional clearance is mandatory for a successful outcome. MPNSTs are generally considered chemotherapy and radiotherapy resistant tumors. However, there are reports of routine postoperative radiotherapy and even radiotherapy as a single modality alone for MPNST in literature.[13] In our case, we performed a right pneumonectomy with tracheoplasty. The postoperative duration was uneventful. The patient was followed up periodically after discharge.

CONCLUSION

MPNST constitutes a significant proportion of soft tissue sarcoma. A combination of clinical, radiological, pathological, and immunohistochemistry helps in diagnosing these tumors. The overall treatment approach should be like that of any other high grade sarcomas. Though multimodality therapy, including surgical resection and adjuvant radiotherapy, is available, the prognosis remains dismal. Modern clinical studies and the development of effective targeted chemotherapy are needed to gain control of the disease.

REFERENCES

1. Hruban RH, Shiu MH, Senie RT, Woodruff JM. Malignant peripheral nerve sheath tumors of the buttock and lower extremity A study of 43 cases. Cancer. 1990;66: 1253–65.
2. Angelov L, Guha A. Peripheral Nerve Tumors. In: Berstein M, Berger MS, editor. Neuro oncology Essentials. 1. New York Theme Publishers; 2000;434–44.
3. Enzinger F, Weiss SW. Malignant tumors of peripheral nerves. In: Soft Tissue Tumors. 3rd ed. St. Louis: Mosby; 1995:889-28.
4. Wanebo JE, Malik JM, VandenBerg SR, Wanebo JH, Driesen N, Persing JA. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 28 cases. Cancer. 1993; 71:1247–53.
5. Ducatman SB, Bernd WS, David GP, Herbert MR, Duane MI. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. Cancer. 1986; 57: 2006–21.
6. Ducatman B, Scheithauer B, Piepgras D, et al: Malignant peripheral nerve sheath tumors: A clinicopathologic study of 120 cases. Cancer1986; 57:2006-21,

7. Evans DGR, Baser ME, McGaughran J, et al: Malignant peripheral nerve sheath tumours in neurofibromatosis 1. J Med Genet 2002;39:311-14,
8. Wong WW, Hirose T, Scheithauer BW, et al. Malignant peripheral nerve sheath tumor: analysis of treatment outcome. Int J Radiat Oncol Biol Phys. 1998; 42: 351-60.
9. Weiss S, Goldblum J. Malignant tumors of the peripheral nerves. In: Weiss SW, Goldblum JR, eds. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St. Louis: Mosby; 2001:1209-63.
10. Kransdorf MJ. Malignant soft-tissue tumors in a large referral population: distribution by age, sex, and location. AJR Am Roentgenol. 1995; 164:129-34.
11. Collin C, Godbold J, Hajdu S, et al: Localized extremity soft tissue sarcoma: An analysis of factors affecting survival. J Clin Oncol1987; 5:601-612,.
12. Ferner RE, Gutmann DH: International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis 1. Cancer Res2002; 62:1573-77.
13. Ghosh BC, Ghosh L, Huvos AG, Fortner JG. Malignant schwannoma A clinicopathologic study. Cancer.1973; 31:184–90.