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Giant intrapulmonary malignant peripheral nerve sheath tumour

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Abstract

Pulmonary malignant peripheral nerve sheath tumours (MPNSTs) are extremely rare soft tissue sarcomas that develop from the cell constituting the nerve sheaths, approximately 5–10% of all soft tissue sarcomas. We present a rare case of primary lung MPNST in an adult female non-smoker patient, in whom surgical thoracotomy approach has obtained a good control of the disease. Low-grade MPNST was established from excisional biopsy followed by immunohistochemistry.

Introduction

Malignant peripheral nerve sheath tumour (MPNST) is a rare and aggressive sarcoma, which comprises 5–10% of all sarcomas of the soft tissue. In the past, other terminologies such as malignant schwannoma, malignant neurilemoma, or neurogenic sarcoma were used for MPNST [1]. MPNST is derived from Schwann cells or pluripotent cells from neural crest, with the incidence of 0.001% in general population and 0.16% in patients with neurofibromatosis type 1 [2]. Moderate and large nerve tissues are more often affected than smaller ones. MPNST most frequently occurs at the extremities, head, and neck, while intrathoracic MPNST rarely occurs [3]. The risk for MPNST peaks at the age of 20–50 years with a recurrence rate of 40–65% and metastasis rate of 40–68% (depends on the degree of tumour during histological examination). The pulmonary tissue is the most common location for metastasis [1].

Case Report

The case was a 33-year-old non-smoker woman with a giant intrapulmonary MPNST. The patient came with right chest pain and she was first diagnosed with tuberculosis and right pleural effusion. Abdominal ultrasonography and computed tomography (CT) scan with contrast showed a large space-occupying lesion (SOL). Open biopsy and pathology anatomy examination revealed the tip of mass from thorax with low-grade sarcoma/MPNST grade 1. At that time, the patient was suggested to have debulking wedge resection, but the patient refused. A year later, the patient came with severe pain on her right chest. A thoracic CT scan with contrast revealed a giant solid mass (12.8 × 11.1 × 13 cm) at the levels of seventh, eighth, ninth, and 10th right lung segments (Fig. 1). The patient was consulted on gynaecologist, internist, and digestive surgeon. The examination results found no tumours in other organs. Debulking wedge resection was suggested again as the most optimal treatment. The patient refused adjuvant radiotherapy following surgery because of the side effect of radiotherapy. Post-operative histopathological and immunohistochemical examination confirmed that the patient had giant intrapulmonary MPNST with low proliferation rate. A month after surgery, the patient was getting better. Histopathological conclusion (Fig. 2) revealed low-grade sarcoma with MPNST grade 1. Immunohistochemical examination results showed that the tumour stains positive for vimentin, CD56, Epithelial Membrane Antigen (EMA), ki67, S100, and Neuron Specific Enolase (NSE). At present, the patient is still alive and did not show any recurrence.

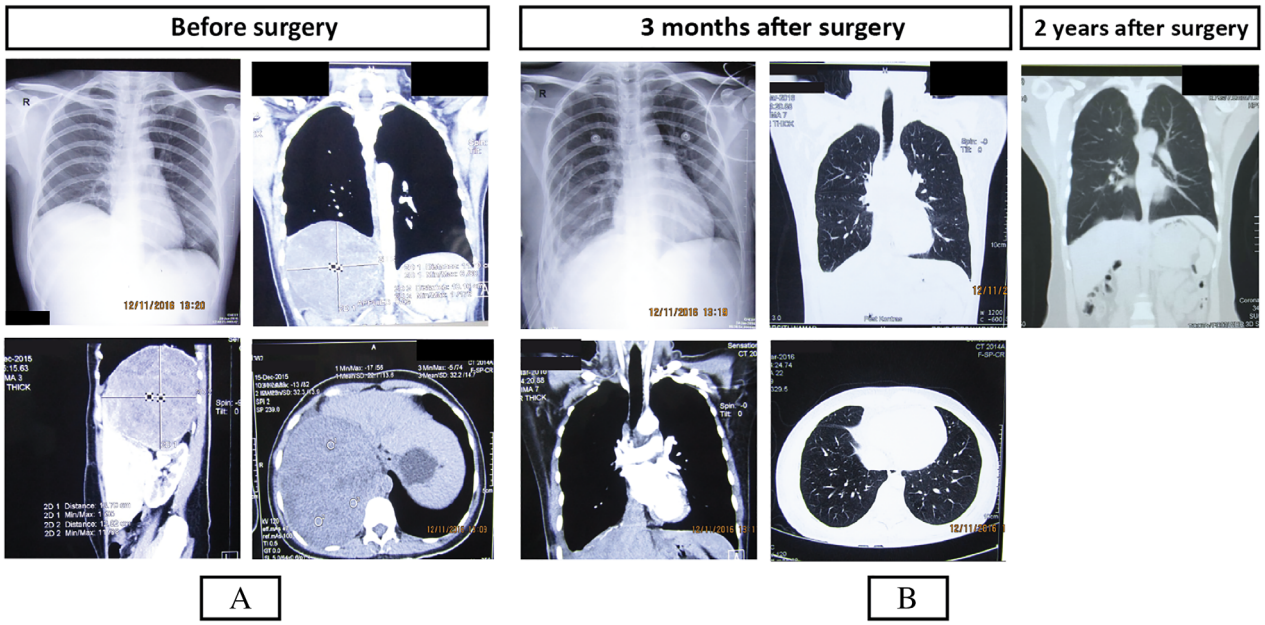

Figure 1. (A) Chest X-ray and thoracic computed tomography (CT) scan with contrast revealed a giant solid mass (12.8 × 11.1 × 13 cm) at the levels of seventh, eighth, ninth, and 10th right lung segments. (B) Chest X-ray and thoracic CT scan three months and two years after surgery.

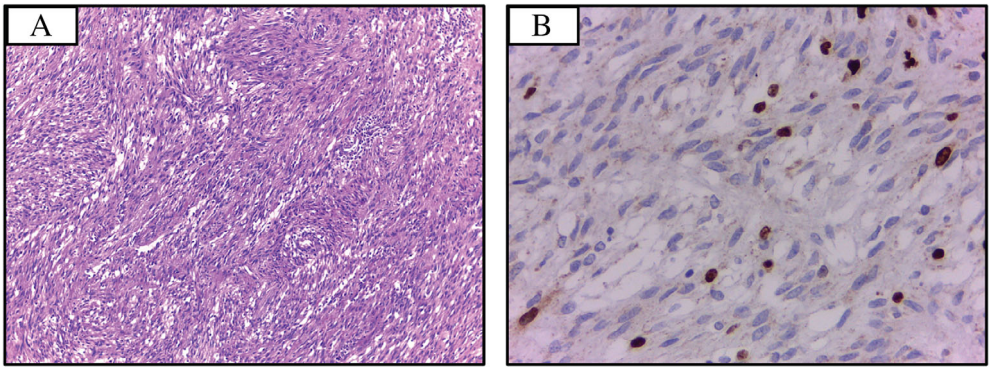

Figure 2. (A) Dark smudged chromatin, nuclear enlargement, and abundant cytoplasm (haematoxylin and eosin (HE) stained). (B) Striking nuclear and cytoplasmic immunoreactivity for S-100.

Discussion

MPNST is a rare neoplasm and comprises 5–10% of incidence rate of all types of soft tissue sarcomas. MPNST is a tumour with delayed growth, appearing asymptomatic for a prolonged period. Therefore, it is often diagnosed during advanced stage and achieved a large size. Pain and sensory-motor disorders appear later when the tumour gives pressure to the nerve tissue [1,4]. Our patient, a 33-year-old and non-smoker woman, complained of pain on her right chest. Pain is the most common symptom [5]. MPNST is often associated with NF1 with varying incidence rate of 20–30% [5]. The contrary was found in our patient. MPNST is usually large, ovoid, with white margin with necrotic and haemorrhagic area [4,5]. MPNST 4,5]. MPNST had high attenuation level, necrosis, and haemorrhagic area on CT. In our patient, a thoracic CT scan with contrast revealed a giant solid mass (12.8 × 11.1 × 13 cm) at seventh, eighth, ninth, and 10th right lung segments (Fig. 1). Microscopically, MPNST showed spindle cells arranged in dense cellular fascicle resembling fibrosarcoma. The tumour cells had slender nucleus with wavy contour and unclear cytoplasm. Cells are arranged in sweeping fascicles pattern [4]. S-100-positive focal staining was seen in 50–90% cases.

Tumour cells showed diffuse positive staining for vimentin. Immunohistochemistry analysis of our patient revealed diffuse positive staining for vimentin and S-100. Dark smudged chromatin, nuclear enlargement, and abundant cytoplasm were observed during haematoxylin and eosin (HE) staining (Fig. 2). The treatment of choice is surgical resection. Neoadjuvant chemotherapy is often used for the early stage of unresectable tumour with the purpose to reduce the level/size of mass before resection. Radiation therapy can be given as an adjuvant to surgical resection to improve local control [1,4,5]. But, other studies reveal that patients who received surgery only or multimodalities had no effect on recurrence rate [5]. However, with the features of MPNST, multimodality treatments are recommended [5]. In our patient, complete resection of tumour was performed and showed good local control.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Acknowledgment

The case report was presented as oral presentation at the APSR conference in 2016 (Ref: APSR6-0517).

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