Resection of a giant malignant mediastinal peripheral nerve sheath tumour under cardiopulmonary bypass

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ABSTRACT

Malignant peripheral nerve sheath tumour (MPNST) is a rare occurrence in the mediastinum. It is biologically aggressive and is generally resistant to chemoradiation therapy. The mainstay of treatment is complete surgical resection. We report a large MPNST which invaded into the adjacent aortic wall in a 50-year-old man. Extensive resection, which included aortic reconstruction under cardiopulmonary bypass and deep hypothermic arrest, was necessary for a good long-term outcome.

Keywords: aortic reconstruction, cardiopulmonary bypass, malignant mediastinal peripheral nerve sheath tumour, mediastinal nerve sheath tumour, phrenic nerve tumour

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INTRODUCTION

Malignant mediastinal peripheral nerve sheath tumour (MPNST) is a rare mediastinal tumour which requires complete surgical resection for a favourable long-term result. Our case illustrates a large MPNST arising from the left phrenic nerve. Due to the extent of invasion into the surrounding vital structures, complete resection necessitated the institution of cardiopulmonary bypass (CBP) support and aortic reconstruction. This resulted in the freedom of recurrence three years after the surgery. Through this case, we would like to emphasise the need for surgical aggression with the goal of cure. Also, a locally-advanced tumour as such should be resected in a tertiary centre where extended support is feasible.

CASE REPORT

A 50-year-old man with no medical history presented with an incidental large left lung mass demonstrated on a chest radiograph. Subsequent computed tomography (CT) showed a large well-defined 13.1 cm × 9.5 cm × 13.5 cm mass in the left hemithorax (Fig. 1). The mass was heterogeneous in appearance with central necrosis. It had also caused a narrowing of the left main bronchus, as well as of the main pulmonary trunk and left pulmonary artery. In view of the most probable diagnosis of a thymic tumour, the patient was offered surgical resection.

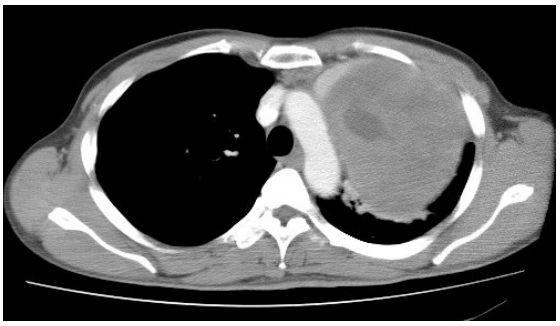

Fig. 1 Axial CT image shows a large mediastinal tumour with central necrosis, occupying most of the left hemithorax.

A median sternotomy was performed to gain access to the mediastinal mass. An intraoperative biopsy of the tumour was obtained, and the frozen section showed an atypical spindle cell tumour. The decision was made to proceed with resection in view of the size of the tumour and the compressive nature to the surrounding vital structures. During mobilisation, the tumour was found to have invaded the pericardium, the left phrenic nerve, and was densely adherent to the wall of distal aortic arch and proximal descending thoracic aorta. The extensive involvement of the tumour required extension of the incision to the left anterolateral thoracotomy. To facilitate the complete resection of the tumour, which had invaded the aortic arch, a CPB was therefore instituted, and the patient was cooled to 18°C via the right atrium and bi-arterial cannulation with the femoral artery and ascending aorta.

Following complete resection of the tumour, the involved aorta was deemed too friable for direct interposition aortic graft without total aortic arch replacement. Hence, the aortic arch just proximal to the origin of the left subclavian artery was oversewn. The left subclavian artery was sacrificed. A size-22 gelseal graft was then anastomosed from the ascending aorta to the proximal descending aorta. In total, 25 min of circulatory arrest was required for the anastomosis. The

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patient was weaned off CPB with a small amount of adrenaline support.

The tumour cells showed strong staining for bcl-2 and focal staining for SMA. There was no expression for CD34, CD99, desmin, caldesmon, S100 and beta-catenin, and all the epithelial markers such as EMA, AE1/3, CK7 and CK19 were also negative. The MIB-1 proliferation fraction was low (≤ 5%) and *in situ* hybridisation for EBER was negative on the tumour mass. Taking into consideration the cytoarchitectural features and anatomical location, the features were most suggestive of a low grade malignant MPNST, although the S100 protein was negative. Postoperatively, the patient's recovery was relatively unremarkable, and he received 60 Gray of external beam radiotherapy to the mediastinum in 30 fractions over 42 days. Three years following resection, the patient was very well clinically with no recurrence on surveillance CT.

DISCUSSION

The majority of the tumours of nerve sheath origin in adults are either benign schwannomas or neurofibromas, and they usually arise from either an intercostal nerve or a sympathetic nerve. MPNST is a rare kind of tumour, usually associated with Neurofibromatosis Type 1, which typically arises from simple or plexiform neurofibroma.[1] Thus far, the best modality of treatment is still complete surgical resection. It has been shown that aggressive surgical resection of this tumour confers long-term benefits in patients who otherwise would have no other viable treatment option.[2] Although the five-year survival rates have been reported to be up to 75%, MPNST often advances locally and can occasionally metastasise to the lung or other organs.[3] Therefore, in addition to a complete surgical resection, adjuvant therapy is usually advocated. Our patient received postoperative adjuvant radiotherapy. In our case, the tumour originated from the left phrenic nerve. Due to its size and the dense adherent nature to the aortic wall, we encountered a great technical challenge during mobilisation. In this case, with the aid of cardiopulmonary support, we had not only avoided a disastrous intraoperative event, but had also managed to achieve complete resection and a favourable long-term outcome. This case highlights the importance of such a complex case being operated on in a tertiary centre where the appropriate experienced team of personnel is available. Any large mediastinal tumour compressing great vessels would need a sternotomy and CBP backup.

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