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Case Report

Intrathoracic Malignant Peripheral Nerve Sheath Tumors and Management

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Abstract

Malignant peripheral nerve sheath tumors (MPNST) are a rare entity. They account for 10% of all soft tissue sarcomas [1]. These tumors occur most frequently at axial sites and are characterized by local aggressiveness and a propensity to metastasize [2]. We present a case of a 30-year-old male with a history of right chest pain, shortness of breath, and significant weight loss. Imaging demonstrated a massive multiloculated lung mass measuring 12.6 x 19.8 x 14.7 cm eroding into the adjacent posterior 4th and 5th ribs. The tumor was resected successfully via an open thoracotomy incision with rib resection. Pathology identified an MPNST with epithelial differentiation and spindle cell sarcoma features. Currently, the patient has achieved a 7-month disease free survival.

Keywords: *Nerve Sheath Tumors; Sarcoma; Thoracotomy*

Introduction

Malignant peripheral nerve sheath tumors (MPNST) are rare soft tissue sarcomas that are most commonly found in the extremities and very rarely intrathoracic. By definition a MPNST is a malignant tumor arising from a peripheral nerve or showing nerve sheath differentiation. We present a case of a MPNST that is both very large and located intrathoracic, making it especially unusual.

Case Report

Our patient is a 30 year-old-man who presented with a one month complaint of right chest pain, shortness of breath, and weight loss of 15 pounds. On review of systems he had chronic cough with no fever or chills. On physical exam, the patient had decreased breath sounds in the right chest on auscultation. The patient then underwent chest computed tomography (CT) which showed a right pleural effusion, completely occupying the right hemithorax with compressive atelectasis of the right lung and mediastinal shift to the left. The effusion was due to a 12.6 x 19.8 x 14.7 cm multiloculated structure with a soft tissue component eroding into the adjacent posterior 4th and 5th ribs. The patient then underwent thoracentesis, where cytology confirmed the diagnosis of malignant peripheral nerve sheath tumor (MPNST) with epithelial differentiation and spindle cell sarcoma features, pathologic stage T2bN0M0, group stage 3.

After making the decision for surgical resection, the patient underwent a median sternotomy for mediastinal lymph node dissection, then the tumor was approached via a right thoracotomy for a right pneumonectomy and en-bloc resection of the 4th and 5th ribs. The closure was completed using a latissimus dorsi muscle flap transposition to the right bronchial stump. His postoperative course was unremarkable and the patient was discharged from the hospital on postoperative day (5). Pathology of the specimen confirmed the diagnosis of MPNST. Furthermore, FISH staining was negative for SS18 (18q11.2) rearrangement, indicating absence of a synovial sarcoma component. The patient successfully completed an 8-week course of adjuvant radiation therapy. Currently he has achieved a 7-month disease free survival.

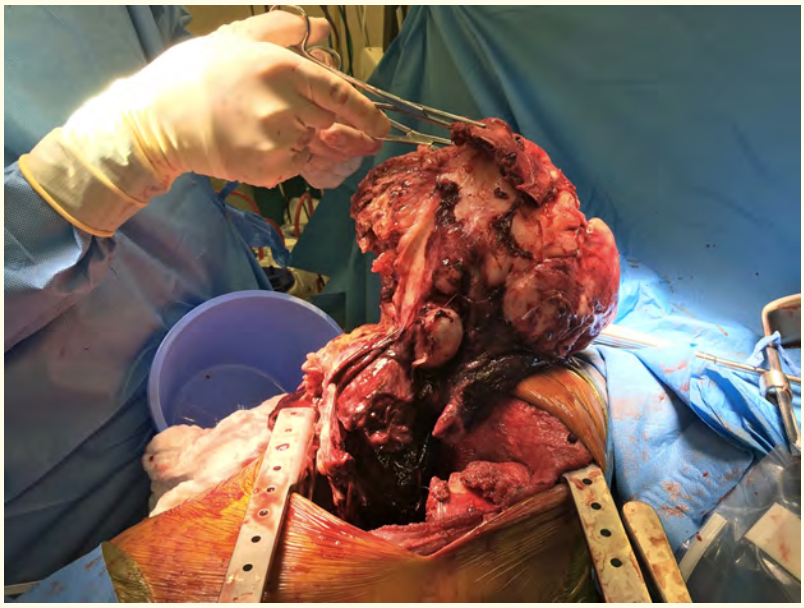

Figure 1: *Gross appearance of intrathoracic malignant peripheral nerve sheath sarcoma.*

Discussion

MPNSTs account for 5 - 10% of all soft tissue sarcomas [3-5]. MPNST is defined as a malignant tumor arising from a peripheral nerve or showing nerve sheath differentiation. MPNSTs are typically seen in patients between 20 and 50 years of age, although occasional cases may be seen in the pediatric population. In symptomatic cases the mass may be palpable, painful or may cause neuropathic symptoms like paraesthesia, motor weakness or radicular pain, or could be discovered incidentally on imaging [5]. MPNSTs are usually sporadic, but 20 - 30% associated with neurofibromatosis-1 (NF1) [6]. Despite the gross similarity between MPNSTs and benign peripheral nerve sheath tumors like schwannomas and neurofibromas, MPNSTs in most parts contain hyperchromatic spindle cells with nuclear atypia, mitotic activity, and areas of tumor necrosis on histology. Most often sites of MPNSTs are the extremities, however, they can appear anywhere in the body. The intrathoracic ones are rare. MPNSTs survival rates are 34 - 60% for 5 years and 22 - 45% for 10 years [6]. Kamran et al. reported their experience with 84 MPNST cases, 15 of which were intrathoracic MPNST. Their analysis showed an average tumor size of 11 cm (range 3 - 32 cm). In the current report, we present a case of the largest resected intrathoracic MPNST with lung and rib invasion. Such cases are usually deemed unresectable. Herein, we demonstrate the feasibility of surgical resection of large intrathoracic MPNST, with concomitant pneumonectomy and rib resection if needed. Naturally, the patient's adequate functional status and absence of other co-morbidities contributed largely to the successful outcome. In conclusion, our group advocates for the consideration of surgical resection of MPNST even when they are detected in large sizes. Aggressive surgical resection, if the patient's condition allows it, could provide an improved survival compared to the conventional non-operative management for such large tumors. Otherwise, neoadjuvant chemoradiation is often used for initially unresectable tumors, with intention to resect it later in treatment.

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Authors Contributions

Manuscript Preparation: Tracy Sambo, MD; Anwar Jebran, MD; Manuscript Editing: Nancy Panko, MD; Francis Podbielski, MD.

Consent for Publication

Patient provided consent for presentation and publication of depersonalized data.

Competing Interests

None.

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