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Case Report

A rare case of extraskeletal Ewing sarcoma ☆,☆☆

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ABSTRACT

Extraskeletal Ewing Sarcoma is a rare tumor of the soft tissues. Primary Ewing Sarcoma of the lung is an exceedingly uncommon variant of Extraskeletal Ewing Sarcoma, with only a small number of cases reported in the literature. We present a case of a 43-year-old woman who presented with a locally invasive mass of the right lung, biopsy-proven to be Ewing Sarcoma.

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Introduction

Ewing Sarcoma is a neuroectodermal tumor that most commonly arises from the bone in the pediatric patient population. Extraskeletal Ewing Sarcoma (EES) defines the subset of tumors occurring in the soft tissues outside of the axial or appendicular skeleton, comprising approximately 20%-30% of all cases of Ewing Sarcoma [1]. Primary pulmonary Ewing Sarcoma, a variant of EES, is exceedingly rare with only a small number of previously reported cases in the literature [2]. As is the case with Extraskeletal Ewing Sarcoma as a whole, Primary Ewing Sarcoma of the lung portends a poor prognosis, particularly in cases of advanced or metastatic disease.

Case presentation

A 43-year-old woman presented to the emergency department with shortness of breath, cough and right-sided chest pain for 2 months. Approximately one month before she presented to our institution, she was seen at an outside facility for similar symptoms. She was prescribed antibiotics at that time; however, her symptoms failed to improve.

On presentation, her blood pressure was 134/75 mmHg. Her heart rate was 95 beats per minute and her respiratory rate was 14 breaths per minute. Her oxygen saturation was 93% on room air. On physical examination, she was alert and oriented to person, place, and time. She had a regular heart rate. She

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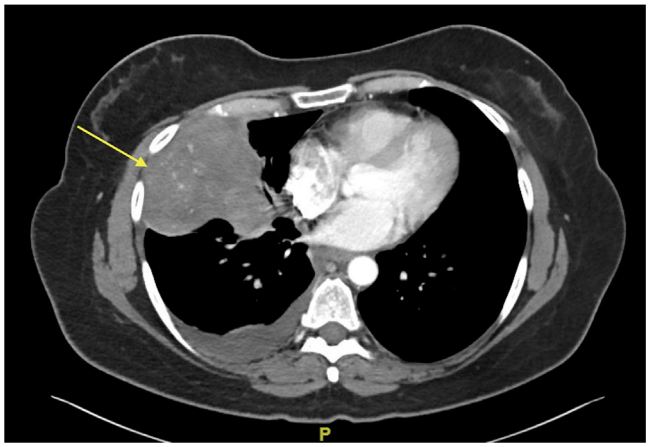

Fig. 1A – Axial CT of the chest on a soft tissue window shows a mass of the right lung (yellow arrow) involving the right middle lobe and portions of the right upper lobe. A right pleural effusion is also present.

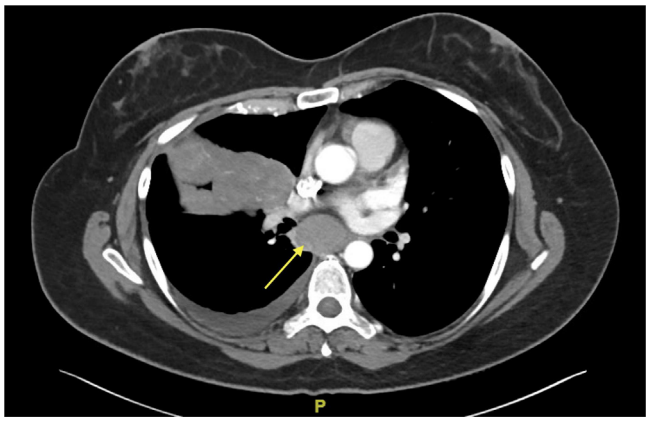

Fig. 1B – Axial CT of the chest on a soft tissue window showing a pathologically enlarged subcarinal lymph node (arrow). The pulmonary mass and pleural effusion are again seen.

had decreased breath sounds and dullness to percussion over the mid to lower right hemithorax.

An anteroposterior chest radiograph was performed. This showed an airspace opacity of the right mid to lower lung with a moderate volume right pleural effusion. A computed tomography (CT) scan of the chest, abdomen and pelvis was then performed (Fig. 1A–D). This demonstrated a mass of the right lung centered within the right middle lobe, measuring up to 16 cm in greatest dimension. The mass invaded the right hemidiaphragm, extending to involve the subdiaphragmatic space of the right upper quadrant. The mass also minimally invaded the right lateral chest wall, without involvement of the adjacent ribs. There were also several pathologically enlarged mediastinal lymph nodes in addition to a moderate volume pleural effusion.

A core needle biopsy was performed. Pathologic analysis demonstrated solid cores of lung tissue with clustered,

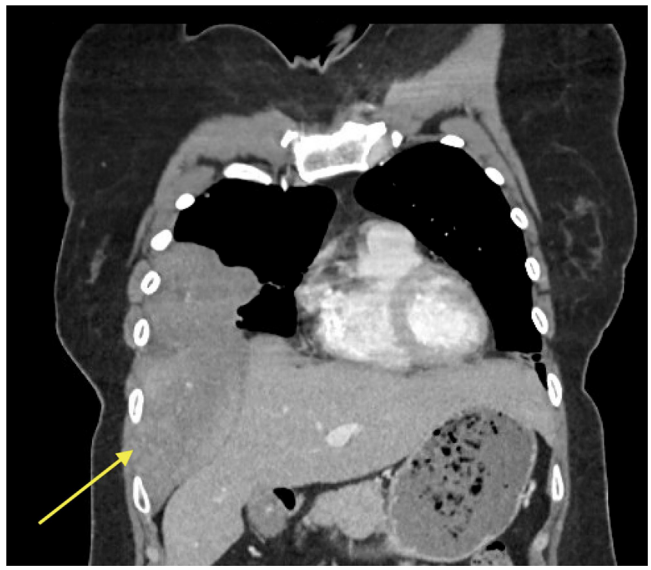

Fig. 1C – Coronal CT of the chest on a soft tissue window showing the mass invading through the right hemidiaphragm into the right subdiaphragmatic space (arrow). There is mass effect upon the adjacent liver.

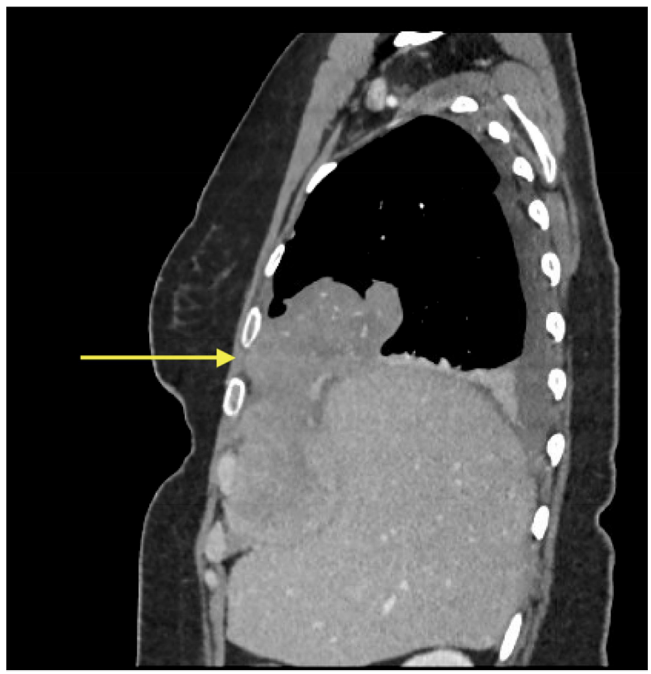

Fig. 1D – Sagittal CT of the chest on a soft tissue window showing the mass invading the abdomen as well as the chest wall (arrow) without substantial involvement of the adjacent ribs.

rounded blue cells with a generally undifferentiated appearance (Fig. 2). Fluorescence in situ hybridization testing was positive for the EWSR1-ERG gene translocation, diagnostic of Ewing Sarcoma.

She was initiated on a regimen of chemotherapy, alternating between vincristine, adriamycin and cyclophosphamide

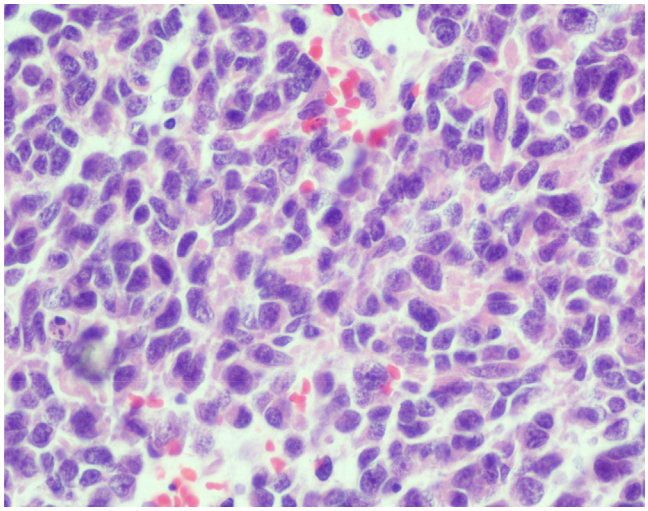

Fig. 2 – Microscopic image of the tumor showing small, rounded blue cells with an undifferentiated appearance. Cells with this appearance are typical of Ewing Sarcoma.

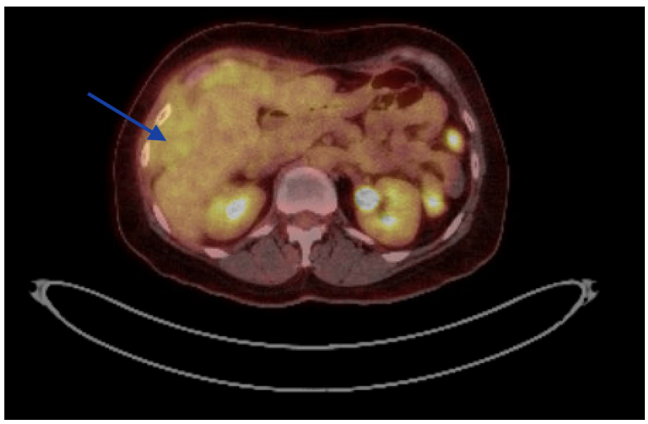

Fig. 3A – Axial FDG-PET CT image showing increasing FDG-avid tumor burden in the right upper quadrant (arrow).

with ifosfamide and etoposide, completing a total of 6 cycles. Follow-up fluorodeoxyglucose-positron emission tomography (FDG-PET) after this treatment demonstrated an overall positive radiographic response with decreased tumor bulk within the lung, however some persistent FDG-avid tumor in the right middle and upper lobes as well as within the right diaphragm and subdiaphragmatic space. The patient was subsequently evaluated by cardiothoracic surgery and underwent a right thoracotomy, right middle lobectomy, right upper lobe wedge resection, and diaphragmatic mass resection with diaphragmatic repair and resuspension.

She continued chemotherapy following surgery with plans for future radiation therapy. However, a follow-up FDG-PET several months after surgery showed worsening disease burden with increasing right upper quadrant tumor bulk and multifocal metastases involving the pleura and abdominal wall musculature (Fig. 3A–C). The patient's clinical status declined over the next several months despite continued treatment and the decision was made to transition to hospice care.

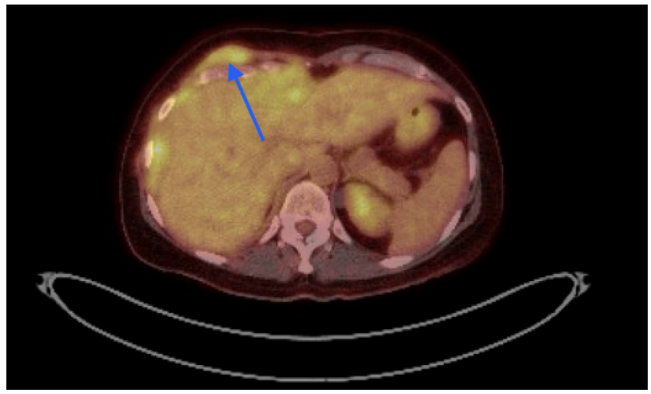

Fig. 3B – Axial FDG-PET CT image showing a right abdominal wall musculature metastasis (arrow).

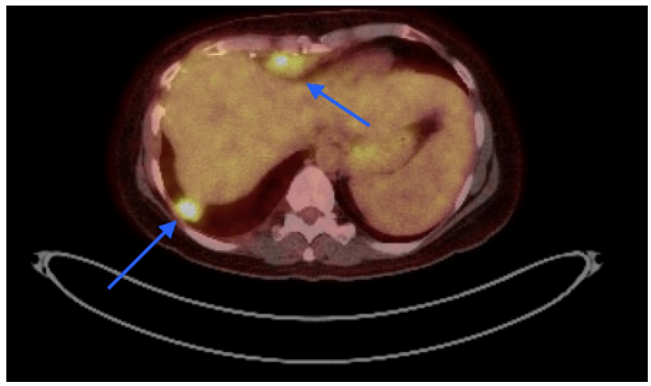

Fig. 3C – Axial FDG-PET CT image showing pleural metastases (arrows).

Discussion

Extraskeletal Ewing Sarcoma can arise from a variety of locations. This includes the paravertebral soft tissues, chest wall (the so-called 'Askin tumor'), pelvis and lower limbs. The lung is an additional, albeit extraordinarily rare, site of origin with only a small number of cases previously reported in the literature [2].

Microscopically, tumor cells of EES appear small, monomorphic and blue, containing large rounded nuclei [3]. Detection of chromosomal translocations involving the EWSR1 gene on chromosome 22 (such as EWSR1-EWS, EWSR1-ERG and EWSR1-FLI-1) within tumor cells is diagnostic of EES [4].

The radiologic appearance of EES is nonspecific. Despite this, imaging plays an essential role in the diagnosis, staging, and preoperative evaluation of EES as well as in the assessment of treatment response [5]. On computed tomography, the tumor often appears as a mass with similar density to that of skeletal muscle with regular-appearing margins. Heterogeneous enhancement with intravenous contrast is typical, in part due to areas of necrotic change within the mass. Additionally, calcifications are seen in approximately 10% of

cases [5]. On magnetic resonance imaging, the tumors are typically hypointense to isointense on T1, hyperintense on T2 and demonstrate variable, heterogeneous enhancement following intravenous Gadolinium infusion [5]. FDG-PET plays an important role in monitoring response to treatment and in the detection of disease recurrence [6].

Surgical excision represents the mainstay of therapy in localized EES [1]. However, surgical therapy alone is often considered insufficient and chemotherapy with or without radiation therapy is also typically administered depending on several factors including tumor location, disease stage, and lesion resectability [7]. In cases where negative resection margins cannot be achieved, postoperative radiotherapy is routinely utilized to achieve greater local disease control [8]. Chemotherapy and radiation are the only treatment options in unresectable disease [9]. The estimated 5-year survival rate in patients with localized disease is 70%-80% [10]. Conversely, in cases of metastatic disease, the 5-year survival is estimated at 32% [10].

Conclusion

Extraskeletal Ewing Sarcoma is an uncommon variant of the Ewing Sarcoma family of tumors, arising from the soft tissues outside of the skeleton. Ewing Sarcoma of the lung is exceptionally rare, with only a small number of cases reported in the literature. Surgery is the mainstay of treatment. Chemotherapy and radiation therapy are additional treatment options used in a variety of situations, including locally advanced, unresectable and metastatic disease.

Patient consent

Written consent for publication was obtained from the patient prior to her death. Approval to publish this manuscript has been given by my institution, its legal representative and an ethics committee. Publication of this document is in accordance with the local legislation.

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