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Case report

Pediatric pleuropulmonary synovial sarcoma: A case report in a recurrent spontaneous pneumothorax

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ABSTRACT

Pleuropulmonary Synovial Sarcoma is a rare lung cancer with a prevalence of <1% among all lung cancers. Little is known about the clinical presentation, disease process, and appropriate treatment. Here we present a 9-year-old male who was taken to the operating room for pleurodesis and blebectomy due to a recurrent spontaneous pneumothorax. Final pathology showed a bleb with an associated mass positive for Pleuropulmonary synovial sarcoma.

1. Introduction

Pleuropulmonary Synovial Sarcoma is a rare lung cancer with a prevalence of <1% among all lung cancers [1]. Little is known about the clinical presentation, disease process, and appropriate treatment. This cancer generally occurs in young adults presenting with chest pain, dyspnea, and imaging findings of a lung mass [2]. There are a few case reports of patients in a similar patient demographic that present with recurrent pneumothoraces [5]. Here, we present a case of pleuropulmonary synovial sarcoma in a pediatric recurrent spontaneous pneumothorax, demonstrating the variability in clinical presentation and patient demographics of this rare tumor.

2. Case report

This is a 9-year old male with no significant past medical or family history, who presented with acute right shoulder pain and imaging concerning for a moderate-sized right-sided pneumothorax. Initially, the patient was treated with a right pigtail catheter with resolution of symptoms and removal of catheter prior to discharge. At follow-up appointment, patient chest X-ray showed recurrent moderate to large size pneumothorax (Fig. 1).

A chest thoracostomy tube was placed and chest CT scan showed a bleb in the posterior apical portion of right upper lobe, with an adjacent 1.5 × 1.3 cm indeterminate soft tissue density projecting towards the posterolateral right lung apex (Fig. 2). Patient was taken to the operating room for a right video-assisted thoracoscopic bleb resection, and mechanical pleurodesis. Intra-operative findings were consistent with imaging. (Fig. 3).

Post-operative clinical course was unremarkable, and patient's chest tube was removed on post-operative day 3 and discharged home. On tissue pathology of bleb resection and adjacent lung parenchyma showed abnormal cells with increased mitotic figures concerning for malignancy with negative margins. Immunohistochemical staining pattern was strong for BCL2 and FISH testing showed a X; 18 translocation confirming the diagnosis of pleuropulmonary synovial sarcoma. Tissue was sent for a second review at University of Washington pathology and Boston's Children hospital that confirmed diagnosis. Further workup to rule out metastatic

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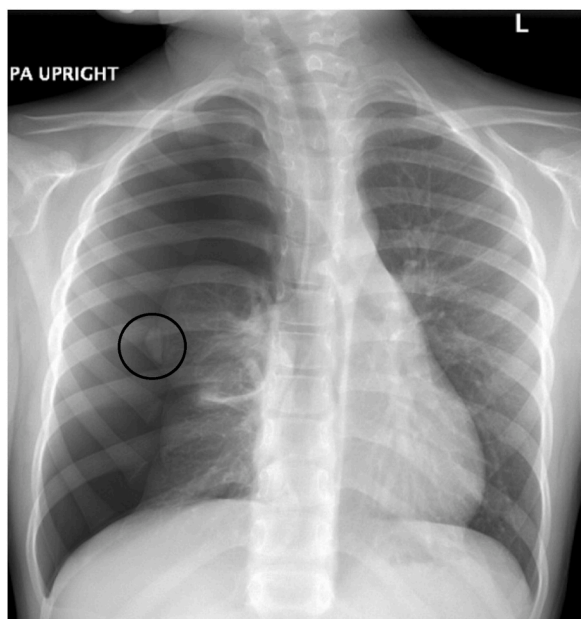


Fig. 1. X-Ray of Recurrent Pneumothorax with adjacent hyperdense lesion.

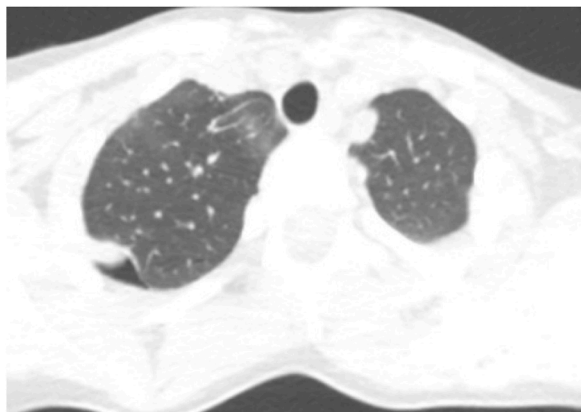


Fig. 2. Non-contrast CT scan of chest.

disease versus extra thoracic primary cancer included a brain MRI and PET CT scan. No evidence of extra thoracic diseases was found.

3. Discussion

Primary pleuropulmonary synovial sarcoma (PPSS) is a rare lung malignancy in the family of sarcomas, derived from immature mesenchymal cells [2]. Clinical presentation of the cancer varies greatly, but typically occurs in young and middle aged adults presenting with cough, chest pain, dyspnea, with a well-defined, uniform appearing mass on radiographic images. Other symptoms that have been associated with disease are hemoptysis and back pain [1,3]. PPSS is often associated with ipsilateral pleural effusions [1]. PPSS masses often lack calcification or associated lymphadenopathy. Recurrent pneumothorax has also been reported in few case reports [4,5]. However, most of these patients were part of the typical demographic of young adults or middle aged adults. The youngest patient described in the literature is a fourteen-year-old female with similar presentation [4]. Our patient is well below the expected age of presentation and exhibits atypical clinical features of the malignancy.

In terms of the histopathology of the disease, there are four subtypes of PPSS: biphasic, monophasic fibrous, monophasic epithelial, and poorly differentiated. This is determined based on the predominance of epithelial or spindle cells [2]. In general, the histology of PPSS is very similar to soft tissue sarcomas extra-thoracic in origin. These tumors are typically positive for pancytokeratin, cytokeratin 7, and epithelial membrane antigen. Synovial sarcomas are characterized genetically by X; 18 (p11.2; q11.2) translocation which fuses *SYT* from chromosome 18 to *SSX1* or *SSX2* on the X chromosome [1].

Our patient showed a monophasic fibrous subtype with predominant spindle cells and high mitotic activity (21 mitoses/10 per

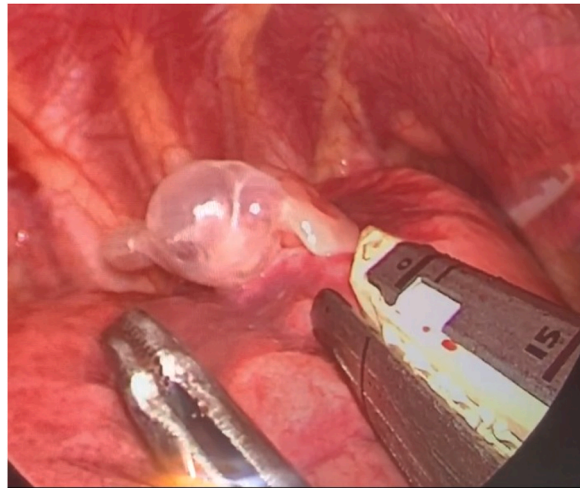


Fig. 3. Right upper lobe bleb visualized and resected during Right Video-Assisted Thoracoscopic Surgery.

units) and FISH studies showed the classic X; 18 chromosomal translocation. Immunohistochemical staining like vimentin and BCL1 has also shown to be beneficial in characterizing these tumors, both of which were strongly positive in our patient [6]. The overall pathology from our patient was highly cellular, without evidence of necrosis, and focal areas of dystrophic calcifications. Unfortunately, outside of the poor differentiation and generic mitotic activity of the tumor, both of which imply poorer prognosis, little is known about the prognostic implications of most of these histopathologic characteristics.

Overall, PPSS is an aggressive tumor, and the few cases with long term follow-up has shown a 50% survival rate at 5 years [2]. However, that can be related to the late presentation showing an advance disease process. Most of these tumors present initially with distant metastasis in the elderly population. Especially in cases when synovial sarcoma is discovered first in the chest, significant care must be taken to rule out an extrathoracic primary sarcoma. Unfortunately due to delayed diagnosis, palliation tends to be a common course of treatment. But the mainstay of treatment for non-metastatic disease surgical resection. In a case report, extensive surgical resection has proven to show some benefits in short term outcomes. Very little is known about the oncological benefit and outcomes comparing anatomical vs non-anatomical resections. Petrosyan et al. reported a pneumonectomy with good results at 16 months for local recurrence and control of the disease [7]. Adjuvant chemotherapy and adjuvant radiation has also been reported being utilized with some benefit, but no studies to determine its efficacy [8]. However, most of the chemotherapy and radiation remains options in palliation not curative treatment.

Given the highly aggressive nature of this cancer, it becomes imperative identifying clinical presentations early. However, the rarity of this entity, the non-specific clinical presentations, and the heterogeneity in patient population make its early diagnosis a challenge.

4. Conclusion

We present a case of a young pediatric patient with recurrent pneumothorax, a seemingly common and benign disease process in pediatric thoracic surgery, sadly turned a more malignant and grim clinical scenario. Primary pulmonary synovial has shown to appear in unpredictable clinical scenarios. The more cases that have occurred, the more variable this disease presentation has shown. This case report highlights some of the novel clinical presentations and patient demographics. Treatment for this highly aggressive malignancy remains resection with adjuvant chemotherapy and radiation as a palliative alternative. Therefore, treatment and timely identification of this cancer remains difficult and further experience must be shared to improve clinical diagnosis and therapeutic measures. As more cases of PPSS emerge, it will be crucial to share experiences regarding clinical course and outcomes to guide future therapy in this disease.

Declaration of competing interest

None.

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