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Pulmonary artery repair following resection of a primary pulmonary sarcoma

Andrew S. Kaufman, Philip S. Mullenix, John S. Thurber, Daniel G. Nicastri

ABSTRACT

Introduction: Pulmonary sarcomas, which account for less than 0.5% of lung cancers, present a diagnostic and therapeutic challenge. Case Report: A 34-year-old healthy male initially presented with left shoulder pain. Imaging showed a left hilar mass—later found to be primary pulmonary synovial sarcoma on further work-up and biopsy. The patient underwent a left upper lobectomy with pulmonary arterioplasty on femoral access cardiopulmonary bypass. On final pathology, the specimen contained positive margins requiring a challenging re-operative completion pneumonectomy. Conclusion: The authors advocate for aggressive initial resection to avoid the need for reoperation or possible delay of adjuvant radiation and chemotherapy.

Keywords: Lobectomy, Pneumonectomy, Sarcoma

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INTRODUCTION

Pulmonary sarcomas are rare, accounting for less than 0.5% of lung cancers. Initially mentioned in German literature in 1865, synovial sarcomas were thought to arise only in joints and extremities. However, these tumors can be found in the chest wall, pleura, lung, mediastinum or heart [1].

CASE REPORT

An healthy 34-year-old active smoker initially presented with new left shoulder pain, which he attributed to recent exercise, he was otherwise in his normal state of good health, training for a marathon. Significant history includes a 20 pack-year (approximately 1–2 packs per day for 10–15 years) smoking history and prior orthopedic only surgeries. The patient denied fever, chills, sweats, weight loss, dyspnea, cough, or hemoptysis. The patient is a career member of the military.

Workup, including radiographs of the left shoulder, revealed an incidentally found left hilar mass. Computed tomography scan further delineated this

3.7x2.9x3.2 cm left hilar mass involving the superior pulmonary vein and upper lobe segmental pulmonary arteries (Figure 1). Positron emission tomography (PET) scan revealed no systemic involvement or tumor FDG avidity. An endobronchial ultrasound guided biopsy (EBUS) with needle aspirate was performed revealing a spindle cell neoplasm with the following positive markers: BCL-9 +, CD99 +, Vimentin +, CAM 5.2 +, and EMA weakly +. The initial interpretation suggested a finding of World Health Organization Type A thymoma. Repeat EBUS for further clarification, showed t(X; 18), CD99, EMA, synaptophysin, CD57, and vimentin positivity, confirming a monophasic synovial sarcoma. The diagnosis of primary pulmonary synovial sarcoma (PPSS) was made.

Surgery was planned and the patient was consented for left thoracotomy tumor resection including lobectomy, possible pneumonectomy, and possible femoral access cardiopulmonary bypass. The patient advocated for avoiding pneumonectomy, because of his extremely active lifestyle. A fourth interspace, serratus-sparing thoracotomy was performed. Intraoperatively, the mass was found to be encasing the first branch of the left pulmonary artery. The main pulmonary artery was not involved and the lung was mobile. A pericardial patch was created and fixed in glutaraldehyde. The inter-lobar pulmonary artery was dissected to a point where the margins appeared to be disease-free. The upper lobe bronchus and pulmonary vein were then identified and dissected from the mass. The lingular arteries and two other upper lobar branches were ligated and the pulmonary vein explored. The inferior ligament was divided and the lower pulmonary vein identified and preserved. The superior pulmonary vein was dissected and divided as proximal to the atrium as possible. Femoral access cardiopulmonary bypass was initiated to resect the mass. The first branch of the pulmonary artery and the left upper lobe bronchus were ligated with a linear cutting stapler. The specimen was removed and sent for frozen section. Margins of concern included the pulmonary vein and artery as well as the upper lobe bronchus. As cardiopulmonary bypass was being used, an additional proximal margin was obtained on the pulmonary artery which included the staple line. The previously created pericardial patch was then used to repair the pulmonary artery (Figure 2). All margins were negative on intraoperative frozen section. The patient tolerated this procedure well and underwent an uneventful recovery. Final pathology was significant for PPSS, with positive margins at the pulmonary artery and vein and suspicious involvement of the bronchus. Three weeks following the initial resection the patient returned to the operating room for a left pneumonectomy in attempt for an R0 resection. Pneumonectomy final pathology was negative on all margins. The patient tolerated the pneumonectomy well and was discharged from the hospital several days following the operation.

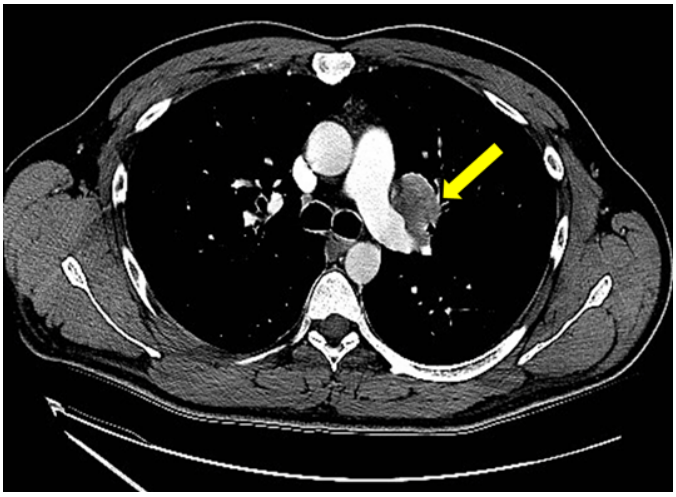

Figure 1: Arrow indicates mass abutting the pulmonary artery.

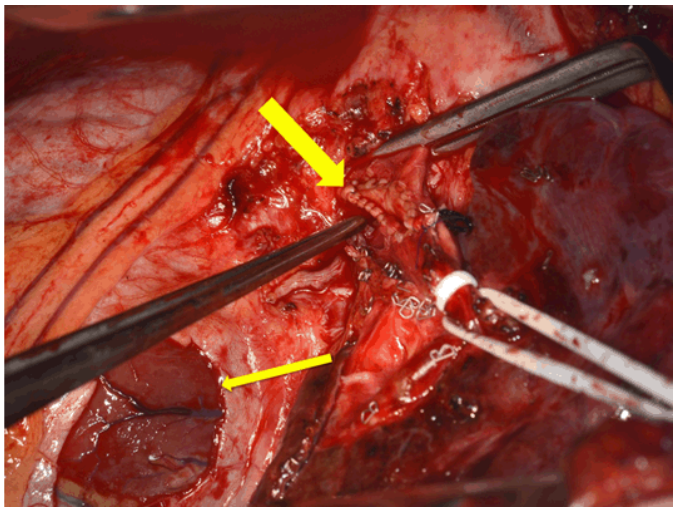

Figure 2: Thick arrow indicates pulmonary artery patch repair and thin arrow indicates patch donor site.

DISCUSSION

Soft tissue sarcomas present a diagnostic and therapeutic challenge. Obtaining the initial accurate tissue diagnosis can be difficult, leading to delay in definitive resection. There are four main subtypes of synovial sarcoma: monophasic fibrous, monophasic epithelial, biphasic, and poorly differentiated monophasic subtype. The prognosis for PPSS is similar to sarcomas of other locations in the body. The tumors do not often metastasize but are locally aggressive and invasive. Overall five-year mortality rate for all sarcomas approaches 50%, which can be worse with tumors of high mitotic rate and size >5 cm [1]. The treatment is primarily resection, regardless of location. Resection must be performed with wide margins, as an inadequate resection can lead to local recurrence. Further important considerations in the evaluation of thoracic sarcoma patients include the determination of the primary tumor location. The favored sites of metastasis for extremity soft tissue sarcomas are the lungs and chest wall. It is important to consider this

scenario before assuming a thoracic sarcoma is primary and not metastatic from an occult extremity lesion. A full body PET scan including extremities may be helpful for PET avid tumors [2].

Several previously published case series involving PPSS show approximately 60 patients with follow-up from 1–20 years. There is a high mortality and recurrence rate seen in these series, suggesting the importance of an adequate primary resection [3–7]. These reports show a slight middle-aged male predominance and that these sarcomas are locally aggressive leading to death from disease in nearly 50% of the patients reported in total. In a patient with a long postoperative life expectancy and good pulmonary function, an early aggressive approach to resection can reduce the need for reoperation to obtain disease-free resection. A confounding factor of this case was the appearance of clear margins and frozen sections with negative margins in the operating room, which led to the decision to stop first at lobectomy. The data for frozen section in primary pulmonary sarcoma is limited. However, a review of 164 patients with extremity sarcomas showed an accurate initial diagnosis only 88% of the time with frozen section. Additionally, frozen section only correctly identified histologic grade and subtype 62% and 47% of the time when compared to permanent fixation [8]. As a result of the reoperation and scar tissue encountered it was technically difficult to obtain a negative margin over the pulmonary artery. However, R0 resection was achieved. If the initial resection were more aggressive with a completion pneumonectomy perhaps a negative margin on the artery would have been more efficiently obtained. Complete R0 resection on initial surgery would have allowed the patient to recover more quickly. The patient continues to be followed and is not planned to receive adjuvant chemotherapy. Per the National Comprehensive Caner Network (NCCN) Clinical Practice guidelines the patient is followed with biannual chest CT scans. The patient's most recent scan, 2.5 year following initial resection, shows no sign of recurrence.

CONCLUSION

Primary pulmonary synovial sarcoma presents a difficult diagnostic and therapeutic challenge. We advocate aggressive resection in a patient with an otherwise long life expectancy because of the rarity of these tumors, leading to lower sensitivity on frozen section.

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Author Contributions

Andrew S. Kaufman – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Philip S. Mullenix – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
John S. Thurber – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Daniel G. Nicastri – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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