Pulmonary Synovial Sarcoma: A Rare Primary Lung Tumor

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ABSTRACT

We report a 32-year-old female patient who presented with complaints of cough, chest pain and dyspnea for the past one and a half month and hemoptysis for one week. Work up for tuberculosis was negative. Chest X-ray demonstrated a homogenous opacity in the left middle and lower zones. CT scan chest described a heterogeneously enhancing mass in the left hemithorax with well-circumscribed smooth margins. Patient was transferred to the surgical ward where a left-sided thoracotomy and left lower lobectomy was performed. Histopathology confirmed the diagnosis of pulmonary synovial sarcoma. Post-operatively, she received adjuvant external beam radiotherapy and remains well at 6 months follow-up.

Keywords: Pulmonary synovial sarcoma; Hemoptysis; Thoracotomy; Radiotherapy

CASE REPORT

A 32-year-old female presented with complaints of cough, chest pain and dyspnea for the past one and a half month and hemoptysis for one week. The cough was initially productive but later became dry. Chest pain was a constant mild, dull ache, radiating to the back. On physical examination, her blood pressure was 110/80 mmHg, pulse 82 beats per minute and respiratory rate 14 beats per minute. She was afebrile. Chest examination revealed decreased chest movements and increased vocal fremitus on the left side. Percussion could not be performed due to pain. Decreased air entry was noted in the middle and lower lung zones.

Chest X-ray revealed a well-circumscribed opacity in the left middle and lower chest zones. Ultrasound of her chest showed a well-defined rounded, hypoechoic cystic lesion in the left lower chest with internal echoes. CT scan of her chest with contrast demonstrated a subsegmental left basal collapse due to a well demarcated heterogeneously enhancing mass in left hemithorax along the posterior chest wall, inferior to the oblique fissure. It was abutting the descending aorta medially and left main bronchus anteriorly. No enlarged hilar or mediastinal lymph nodes were noted. Mild left-sided pleural effusion was seen. CT-guided biopsy of the mass was not performed as the lesion was cystic with the possibility of hydatid disease. Patient was referred to thoracic surgery for surgical excision. After pre-operative work up, the patient underwent a left-sided thoracotomy. During surgery, a firm mass was noted in the left lower lung lobe. Approximately, 100 ml of clear, pleural fluid was noted in the pleural cavity. The upper lung lobe, chest wall, pleura and the aorta were normal. Left lower lung lobectomy was performed. Patient had a smooth post-operative recovery and was discharged 2 weeks after surgery.

The mass was a well circumscribed lesion, about 6.5cm x 6cm x 5cm in size located 2.5 cm from the bronchial margin of resection and 0.1 cm away from the closest peripheral margin of resection. The bronchial margin and bronchial vessels were grossly tumor free. Histologically, tumor mass showed interlacing fascicles and bundles of spindle cells. Individual cells showed moderate amount of eosinophilic cytoplasm containing spindle-shaped nuclei with fine chromatin and inconspicuous nucleoli. Approximately 6-7/10 mitosis was noted per HPF. Interspersed tissue demonstrated areas of

Figure 1: Pre-operative CT scan chest demonstrating mass in left hemithorax

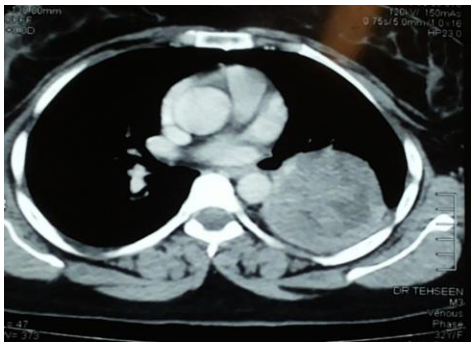


Figure 2: Forceps indicating tumor in left lower lobe

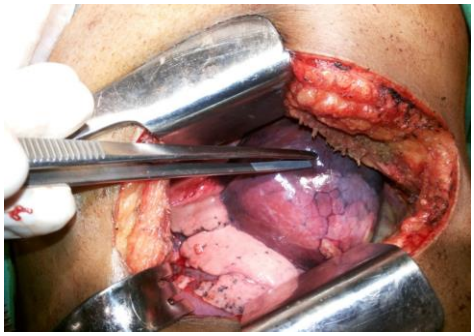


Figure 3: Tumor after excision

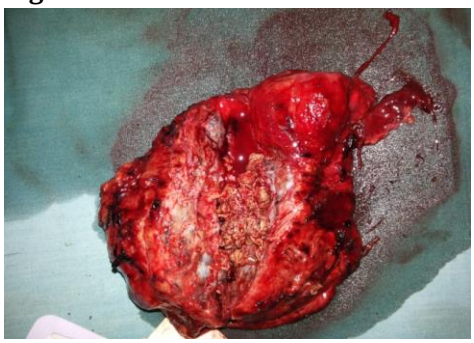
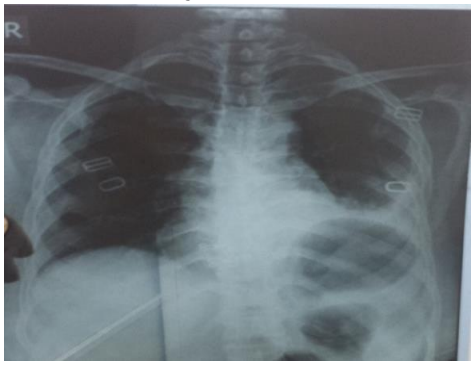


Figure 4: Follow-up X-ray after left lower lobectomy



necrosis. The tumor cells were positive for glycogen PAS +/- D. On immunohistochemistry, the tumor showed positive staining for cytokeratin, CD 99 , C35, EMA, CK AE1/AE3 and BCL-2. A diagnosis of synovial sarcoma, of the monophasic spindle cell variety, was made. Whole body CT scan and bone scan found no metastasis or primary malignancy elsewhere in her body. Patient was given adjuvant external beam radiotherapy with radical dose of 60 Gy in 30 cycles to the primary site, which the patient received uneventfully. She is now on regular follow-up without evidence of residual or recurrent disease.

DISCUSSION

Primary sarcomas of the lung are very rare in comparison to lung carcinomas or metastatic disease to the lung from extrapulmonary sarcomas. The most common lung sarcomas are malignant fibrous histiocytoma, fibrosarcoma and leiomyosarcoma [1]. Primary pulmonary synovial sarcoma is an extremely rare, but aggressive malignancy accounting for less than 0.5% of all lung tumors [2] and nearly 5-10% of all soft tissue sarcomas [3].

Synovial sarcoma is derived from immature mesenchymal elements [4] and not from synovial tissue as the name suggests. It typically presents in adolescents and young adults with a slight male predilection, male to female ratio of 1.2:1 [5], most commonly arising in the soft tissue of extremities, although neck, lung, heart and mediastinal sites have been reported [6]. Pulmonary synovial sarcoma presents with unusual histological features, showing only focal changes within an epithelial-like nodule of sometimes, even benign appearance [7]. There are two main histologic subtypes namely biphasic and monophasic. The biphasic type contains both epithelial and spindle cell components while the monophasic forms shows a uniform spindle pattern [6].

Patients commonly present with chest pain, cough, dyspnea and hemoptypis [8]. Diagnosis is based on radiologic investigations, immunohistochemical staining and cytogenetic analysis. On chest X-ray, pulmonary synovial sarcoma may present as focal pleural thickening, opacification of hemithorax, or consolidation and masses with well-circumscribed borders. Ipsilateral pleural effusion may also be seen [8].

On CT scan chest, primary synovial sarcomas are often described as well-defined heterogenous masses containing areas of fluid attenuation

compatible with hemorrhage or necrosis [8]. Metastasis is usually via the venous route but rarely can happen through lymphatic spread as well. Most synovial sarcomas are positive for epithelial markers such as cytokeratin and epithelial membrane antigen. Many of them will also show reactivity to CD99, vimentin, desmin and BCL2 [6]. Our patient was EMA, cytokeratin, CD99, CD35, CK AE1/AE3 and BCL-2 positive. Despite its high sensitivity, cytogenic evaluation for reciprocal chromosomal translocation (X;18 p11.2;q11.2) is often not performed if the diagnosis is certain on the basis on histopathology and immunochemistry [2]. Owing to its rarity and the paucity of data regarding its natural history, there are no guidelines on optimal treatment [6, 8]. Surgical removal of tumor with the aim to achieve negative resection margins is the current standard of care [7]. Role of adjuvant radiation and/or chemotherapy is unclear since there are no randomized control trials, however some authors advocate the use of adjuvant chemotherapy or radiotherapy [6]. Mullen J et al reported recurrence reduction with the use of conventional fractionation (1-x1.8 to 2 Gy/day) radiotherapy doses between 50-70 Gy [9].

The prognosis is generally poor. Spillane et al [10] demonstrated that an age > 20 years at diagnosis and size ≥ 5 cm were associated with a significantly worse prognosis.

CONCLUSION

Primary pulmonary synovial sarcoma is a rare neoplasm with only a few cases reported in literature. Diagnosis requires clinical, radiologic and immunohistochemical investigations. Treatment of choice is surgical excision with negative resection margins. Histopathology is helpful in confirmation of the diagnosis. Despite lung resection and adjuvant radiochemotherapy, the prognosis for these patients remains dismal.

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