

# Primary Endobronchial Osteosarcomatoid Malignant Neoplasm Presenting with Pneumothorax

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## Abstract

**Introduction:** Pulmonary sarcomas are a rare, diverse group of neoplasms. Management is largely guided by limited case series.

**Presentation of the case:** We present a case of an elderly male who presented with acute respiratory failure due to pneumothorax, confirmed on bronchoscopy to have an obstructing mass in the left mainstem bronchus. Biopsy revealed the lesion to be consistent with a sarcomatoid malignant neoplasm with osteosarcomatoid features. A PET-CT showed no evidence of extrapulmonary disease.

**Conclusion:** There are several subtypes of pulmonary sarcoma. Pulmonary osteosarcomas carry an especially poor prognosis.

**Keywords:** pulmonary; sarcoma; osteosarcoma; endobronchial; pneumothorax

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**Competing Interests:** The authors have declared that no competing interests exist.

**Consent:** Consent was taken from the patient's next of kin for publication of this case report.

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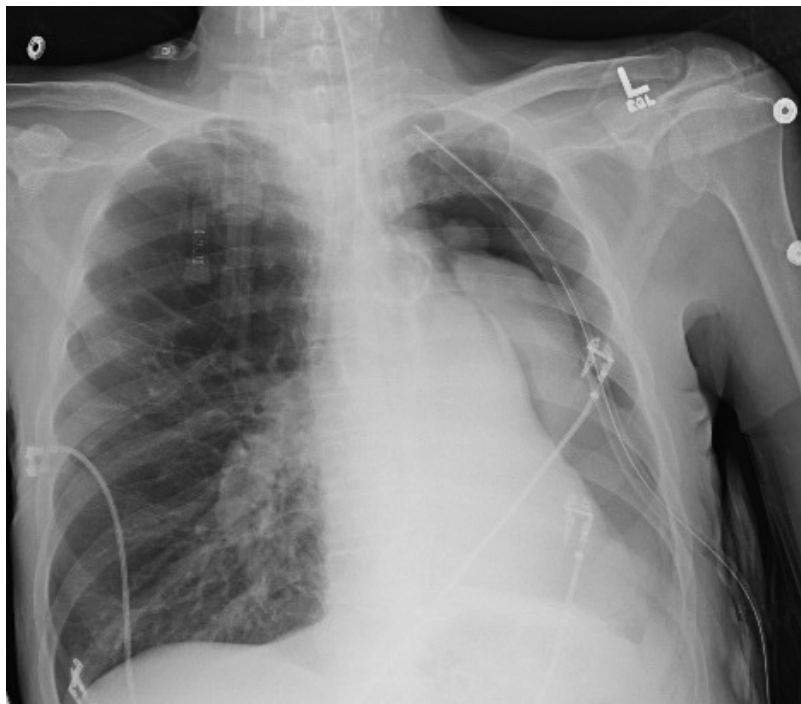
## Introduction

Primary pulmonary sarcomas (PPS) are a heterogeneous group of neoplasms that account for less than one percent of lung tumors. Given the low incidence of PPS, less is known about them compared to epithelial lung tumors. We present the case of a primary endobronchialosteosarcomatoid malignant neoplasm presenting with pneumothorax.

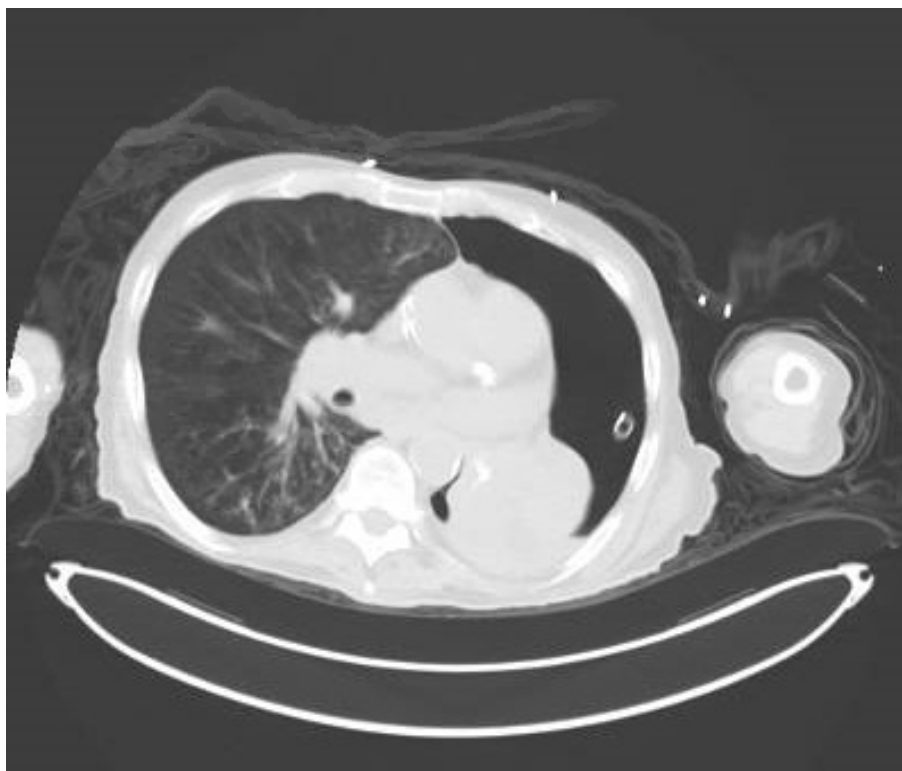
## Case Presentation

A seventy-one-year-old male was transferred from an outside hospital for management of respiratory failure due to suspected obstructing left mainstem bronchus endobronchial lesion. He had a past history of emphysema, former tobacco and alcohol abuse, diabetes mellitus type 2, localized bladder cancer fifteen years prior status post resection in remission, hypertension, hyperlipidemia and stage three chronic kidney disease.

He had initially presented to the outside hospital with hypoxic respiratory failure requiring intubation. Chest xray and CT had revealed a large left-sided pneumothorax and pleural effusion with occlusion of his left mainstem bronchus due to obstructing material (Figures 1 and 2).



**Figure 1** Chest radiograph demonstrating left pneumothorax.



**Figure 2** Axial section on CT chest showing a large mass in the left hilar region.

He received a chest tube and underwent bronchoscopy. Samples from an endobronchial washing showed inflammatory cells and no evidence of malignancy. He was transferred to our facility on hospital day ten for further workup.

Bronchoscopy at our facility on hospital day eleven allowed visualization of a large endobronchial mass which completely obstructed the left mainstem bronchus. The mass was noted to be firm and adherent to the airway. Tumoral specimens were sent for pathology and sputum was collected for cytology. Although attempts to fully excise the mass using hot diathermy forceps and electrocautery were unsuccessful, a partially patent airway was able to be formed allowing for extubation on hospital day fourteen. Sputum cytology revealed no malignant cells. A preliminary surgical pathology report described a sample of white, pink, and red-brown soft tissue that had a mesenchymal appearance with cartilagenous elements. The final surgical pathology report confirmed a sarcomatoid malignant neoplasm with osteosarcomatous features. A PET-CT showed intense activity in the left perihilar region corresponding to the endobronchial lesion, but no distant metastatic disease (Figure 3).



**Figure 3** PET-CT showed increased activity in the patient's endobronchial lesion with no metastatic disease identified.

Bronchoscopy was again performed on day eighteen with laser debulking and balloon recanalization. The tumor was visualized extending into both the left upper and lower lobes. His hospital course was complicated by recurrent pneumonia and respiratory failure requiring re-intubation. Given his severe underlying emphysema and acute decline in the setting of pneumonia, he was deemed a poor candidate for pneumonectomy. Radiation oncology was consulted and he was offered palliative radiotherapy. His family declined this, making him comfort measures only. He passed away shortly after terminal extubation on day 26.

## Discussion

The vast majority of primary lung malignancies are carcinomas including small cell, large cell, squamous cell and adenocarcinoma [1]. Less than one percent of lung tumors are primary pulmonary sarcomas (PPS), a diverse group of neoplasms. PPS account for approximately nine percent of all sarcomas. These tumors originate from the stromal elements of the vasculature, connective tissue and airways of the lungs [2]. As with other lung tumors, classification of pulmonary sarcomas is outlined in the 2015 WHO classification [3]. The PPS include pure sarcomas as well as mixed neoplasms. PPS are far less common than lung metastases from extrapulmonary sarcomas.

Compared to the other types of more common lung neoplasms, knowledge about PPS is lacking. Management of these cancers is largely guided by limited case series despite recent advancements in their characterization. For all varieties of PPS, surgery is the mainstay of treatment. If

excision of the tumor is incomplete, then this is followed by radiotherapy [4]. Chemotherapy is not yet a common practice.

Biopsy alone is often insufficient to make a definitive diagnosis. In this case, the patient's frailty prevented pneumonectomy which would have perhaps allowed better characterization. Nevertheless, histopathologic patterns provided insight. Based on the histologic characteristics of this lesion, the differential included primary pulmonary osteosarcoma, carcinosarcoma and myoepithelial tumor.

Primary osteosarcoma of the lung is exceedingly rare. It is quite possibly the rarest histologic subtype of PPS. In contrast to skeletal osteosarcomas, those of the lung arise in older patients. This subtype occurs in patients in their fourth to seventh decades [5]. Primary osteosarcomas of the lung show a male predominance as well as a predominance for the left lung [5]. Clinicians often cite calcification as a feature of benign nodules, but 10-15% of malignant lesions also show calcification [6]. The prognosis of this subtype is especially poor. Prognosis is generally generated based on the tumor size [6]. These osteosarcomas are further subclassified, with the osteoblastic subtype demonstrating an especially poor prognosis.

Carcinosarcomas originate when a carcinoma develops sarcomatous changes. The carcinoma is usually adenocarcinoma or squamous cell carcinoma [3]. It is recognized that TP53 mutations are commonly present. In one series of pulmonary carcinosarcomas, seventy four percent of patients harbored TP53 genomic alterations [7]. Smoking and asbestos exposure are strong risk factors. Carcinosarcomas can be characterized as either squamous or glandular. Metastases are infrequent [8]. This tumor also shows a male predominance. Peak age for these tumors is in the fifth to seventh decade of life [8]. The diagnosis can be confirmed by showing positive stains for both vimentin and cytokeratin.

Myoepithelial carcinoma originates from the bronchial glands. Similar to salivary glands, the bronchial glands play a role in secretion. Unlike carcinosarcomas, myoepithelial carcinomas are more prone to metastasize [9]. Myoepithelial carcinomas typically arise in the elderly. Fewer than a dozen cases are reported in the English literature.

Regardless of the type of lung malignancy, endobronchial tumors are uncommon. Tracheobronchial tumors comprise just 0.6% of lung tumors [10]. Only a minority of these tumors are benign. Squamous cell, small cell and mucoepidermoid carcinomas along with carcinoid tumors comprise the most common types of tracheobronchial tumors. The incidence of endobronchial PPS is not reported. Symptoms of tracheobronchial tumors may mimic those of obstructive lung disease. It is the size of the tumor, rather than the type that dictates its manifestation. It has been estimated that 2% of spontaneous pneumothoraces occur in the setting of lung malignancy [11]. Pneumothoraces due to lung neoplasms can be caused by one of two mechanisms. One way is air leakage following lung infarction from tumor emboli [12]. The alternative mechanism is via the formation of a ball-valve effect from tumor growth into the airway. Damaged alveoli allow air to escape into the subpleural space. Patients with lung malignancies are also more likely to have chronic obstructive pulmonary disease, in which bleb rupture is not uncommon. When presenting with a pneumothorax, lung tumors typically have a poor prognosis, as the diagnosis was often delayed [11].

## Conclusion

Primary pulmonary sarcomas represent perhaps the rarest type of primary lung malignancy. There remains much to understand about these neoplasms.

## Consent

Patient consent was obtained from the patient to publish this case report.

## Competing Interests

None.

## Acknowledgments

None.

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