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Case Report and Review of the Literature

Pulmonary Artery Sarcoma Diagnosed by Transbronchial Endosonographic (EBUS-TBNA) Approach

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ABSTRACT

Background: Pulmonary Artery Sarcomas are the rarest and the most aggressive vascular tumors. Their accurate diagnosis is challenging due to the lack of specific symptoms and clinical manifestations and they are often initially misdiagnosed and managed as chronic pulmonary thromboembolic disease until an alternate diagnosis is suspected based on the lack of response to anticoagulants. Proper management is thus often delayed until histological confirmation. Moreover, the lack of consensus regarding their diagnosis and treatment further contributes to their reported high mortality [1].

Case Presentation: We present the case of a 34-year-old male, nonsmoker, who presented with hemoptysis, fatigue and dyspnea on exertion 3 months prior to admission. His chest x-ray showed enlargement of the left hilum and chest CT revealed a large intraluminal low attenuation filling defect at the level of the left PA. Chest MRI confirmed the presence of a large endovascular tumor of the left PA. Before referring the patient for surgical biopsy, he underwent bronchoscopy during which the vascular tumor was successfully located, observed and sampled by endobronchial ultrasound (EBUS) transbronchial needle aspiration (TBNA). Tissue biopsy through EBUS-TBNA revealed an intimal sarcoma of the PA. The patient was subsequently referred for surgical excision with curative intent.

Conclusion: Bronchoscopy and EBUS-TBNA may safely and accurately diagnose even rare pathologies of the mediastinal vasculature adjacent to the airways through transbronchial approach.

Background

Pulmonary Artery sarcoma (PAS) arises from either the Intimal wall (PAIS) or the intramural wall of the PA. The incidence of PAIS is estimated to be fewer than 300 cases since it was first reported in 1923, with over 80% of cases being in the pulmonary trunk [2, 3]. It is a very aggressive and dismal prognosis tumor whose challenging diagnostic and management pitfall is that it very closely resembles PE both clinically and radiologically thus leading to important delay until final diagnosis. Those whose diagnosis is delayed have a significantly shorter overall survival than patients whose diagnosis is done in a timely manner However, regardless of the clinical and imaging diagnosis, tissue sampling is warranted before conclusive histopathological diagnosis is confirmed and therapeutic decisions are taken [4]. Most reported cases in the literature focus on surgical biopsy during thoracoscopy, thoracotomy, median sternotomy and other advanced surgical procedures. We report on a PAIS case diagnosed bronchoscopically through endobronchial ultrasound-transbronchial needle aspiration (EBUS-TBNA)

Case Presentation

A 34-year-old male, nonsmoker, with a history of liver sarcoma at the age of 10 treated then with surgical resection and chemotherapy without

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any sign of relapse since. He presented with 3 months history of hemoptysis easy fatigue and dyspnea on exertion. There was no history of chest pain, fever, weight loss or syncope. Vital signs, physical examination and routine laboratory tests were all unremarkable. Chest x-ray revealed enlarged left hilum. (Figure 1).

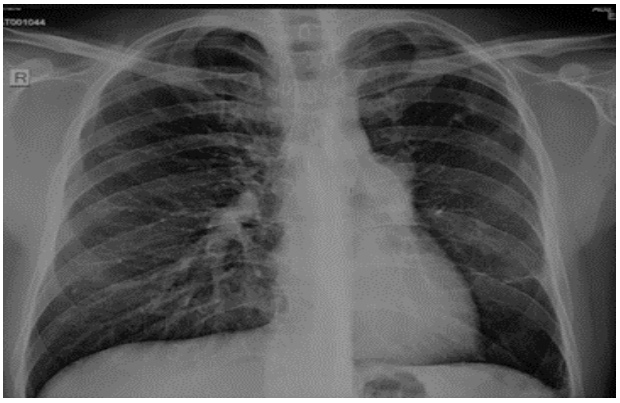

Figure 1: Chest x-ray showing abnormal left hilum.

In the contrast enhanced chest CT which followed a large intraluminal low attenuation filling defect at the level of the left pulmonary artery (PA) could be observed indicating either pulmonary embolism or a vascular tumor producing partial occlusion of the artery (Figure 2). A right upper lobe cavitary lesion was also observed. Flexible bronchoscopy revealed bronchial and tracheal varices. Bronchoalveolar lavage (BAL) came back positive for Aspergillus.

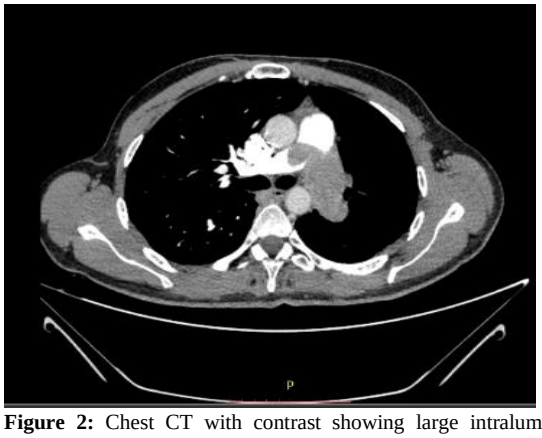

Figure 2: Chest CT with contrast showing large intraluminal low attenuation filling defect at the level of the PA.

Chest MRI confirmed the presence of a large (7.5x4.5 cm) endovascular tumor of the trunk of left PA invading its bifurcation to upper and lower branch producing an important filling defect (Figure 3). Due to the proximity of the left main bronchus to the involved part of the PA, the patient was referred for endobronchial ultrasound (EBUS) bronchoscopy. The lesion floating in the PA anterior to the left main bronchus was noted and a transbronchial/transvascular needle was advanced under direct observation to obtain biopsy (Figure 4).

Histopathological analysis of the biopsy revealed a fibrous stroma with pleomorphic spindle cells, partially giant or multinucleated, in a loose arrangement, with mature lymphocytes and plasma cells scattered in between. These lesions together with all the clinical and radiological findings were consistent with intimal sarcoma of the pulmonary artery (Figure 5).

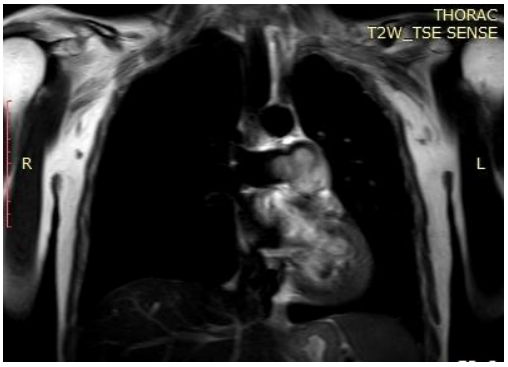

Figure 3: Chest MRI with contrast showing large endovascular tumor occluding the trunk and bifurcation of left PA.

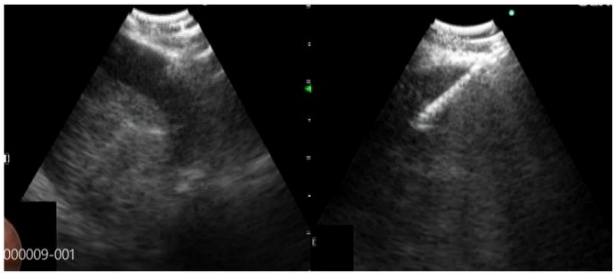

Figure 4: EBUS showing the PA tumor and the transvascular introduction of the EBUS-TBNA needle.

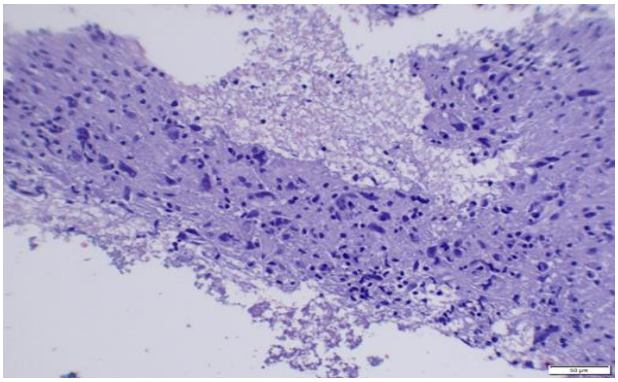

Figure 5: EBUS transbronchial biopsy. [H-E, X200].

Discussion

Clinical presentation of pulmonary artery sarcoma can be variable. A case series reported patients presenting with progressive shortness of breath, chest pain and syncope. Others presented with coughing, haemoptysis, fever, fatigue and weight loss [3]. All the aforementioned symptoms can be attributed either to pulmonary embolism or to Chronic Thrombo-embolic Pulmonary Hypertension (CTEPH). Some differentiating aspects are age and D-Dimers. PAIS tends to be more common in younger patients with no risk factors for PE. On the other hand, in patients with PAIS, d-dimers is lower so values dd > 2.81 ug/mL FEU, make PAIS less probable [4].

Both pathologies can also have similar imaging findings with low attenuation filling defects. However, a recent study suggested that PAIS can be differentiated from PE on CT scans by identifying the different

patterns reflecting the tumor's growth process. The spectrum varies depending on the stage and activity of the tumour. In initial stages polypoid tumour foci forming in the intimal layer of the PA can be observed which subsequently extend to form a cauliflower-like mass or wall thickening. In more advanced stages the tumour occupies almost the entire lumen of the PA causing the "tumour impaction" sign. Half of the patients with PE in the reviewed study, had Deep Venous Thrombosis (DVT), and the observed pattern on CT was tubular-polypoid. In the same study PAIS had higher attenuation than PE, which was attributed to the presence of intratumoral vessels. Other reports highlight the high specificity of the wall eclipse sign (tumour eclipsing the wall of the pulmonary artery prior to infiltrating outside the pulmonary artery) which was present significantly in patients with PAIS but none of the patients with PE [4, 5].

A study also examined the features suggestive of PAIS using Magnetic Resonance Imaging (MRI) and Positron-emission tomography (PET-CT). It concluded that MRI has the ability to distinguish PAIS combined with thrombus whilst PET-CT has the advantage of detecting other foci of PAIS and thus aiding in the staging and detecting distant metastases. The value of PET CT in PAIS evaluation still remains controversial. Tissue biopsy is of course warranted before any therapeutic decision is taken while no clear guidelines exist on the optimal assessment strategy and management of this aggressive and often lethal form of tumour. However regardless of the imaging characteristics, exact tissue confirmation is crucial for management decisions and most authors focus on extensive surgical interventions for obtaining tissue sample for diagnosis.

Endobronchial ultrasound guided transbronchial needle aspiration (EBUS-TBNA) is an invaluable tool for mediastinal lymph node staging in patients suspected to have lung cancer with a sensitivity and a specificity of 91.17% and 100.0% respectively with an excellent safety profile The use of EBUS in the diagnosis of PE has been reported with success in multiple studies [6-8]. This is especially important when CT pulmonary angiography is contraindicated. Recently, EBUS-TBNA has been used to diagnose intra PA masses given its proximity to the bronchial walls. A review of 12 cases, 5 of which had mass-like lesions in the PA and 3 which had persistent or progressive filling defects despite anticoagulation, EBUS-TBNA was used to obtain a biopsy. 6 of the cases sampled were diagnosed with sarcoma [9].

In another case report transbronchial needle aspiration of pulmonary intra-arterial masses proved a safe procedure especially when less invasive approaches failed [10]. EBUS-TBNA was used to sample a PA sarcoma using Pulmonary Rapid on Site cytology Examination (P-ROSE) which helped expedite the diagnosis and timely management of this rare but fatal malignancy using the characteristic clusters of malignant pleomorphic spindled and epithelioid cells [11]. Possible complications include haemorrhage from hypertrophied bronchial arteries especially in pulmonary hypertension which a common consequence to pulmonary artery occlusion [12].

Timely diagnosis is crucial as the prognosis is poor and survival is estimated to be 12-18 months [13]. The treatment modality related with longest survival is surgical excision with pulmonary endarterectomy. Other treatment modalities include targeted therapy with Panzopanib; the only agent approved for use in soft tissue sarcoma, radiotherapy, and chemotherapy. Chemotherapeutic agents include adriamycin plus isosfamide, gemcitabine plus taxane, and dacarbazine. Recent studies show survival benefit when multimodality treatment is applied with adjuvant or neoadjuvant chemotherapy and/or radiotherapy compared to monotherapy such as surgery alone [14, 15].

Conclusion

PAIS poses a dilemma in its diagnosis and management. EBUS-TBNA is highly sensitive and specific and offers a promising diagnostic modality, however, further studies are needed to establish its safety profile.

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