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Case Report

Pleural Vascular Tumor Presented with Conflicting Laboratory and Pathologic Findings

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Abstract

Various etiologies contribute to the development of pleural effusion, although differentiating these causes can be challenging in some cases, especially in patients who have inconsistence clinical and laboratory evidence. We present a 79-year-old female patient with massive right sided pleural effusion along with an air-fluid level and pleural thickening in chest X-ray. In the thoracentesis, the patient had pleural effusion with neutrophil predominance, and in the preliminary pathological examination of the thoracoscopic biopsy of the pleura, severe chronic inflammation with fibrous thickening and mesothelial hyperplasia was reported. Finally, a re-examination of the pathology revealed a final diagnosis of pleural epithelioid hemangioendothelioma, a tumor of vascular origin uncommonly seen in the thorax.

Introduction

Pleural effusion is the most common disease of the pleural space with various causes, among which the most common causes include heart failure, malignant pleural effusion, and parapneumonic effusions [1]. In the management of pleural effusion, diagnosing and evaluating its cause is of great importance, although routine diagnostic criteria sometimes fail to differentiate between underlying causes such as infections and malignancies [2]. Therefor, correct diagnosis of the cause of pleural effusion depends on taking a proper history and examination, performing appropriate radiological and laboratory studies, and ultimately performing a biopsy and pathological examination [3]. In this regard, medical thoracoscopy has a high diagnostic yield in differentiating the causes of pleural effusion especially in cases with undiagnosed pleural effusion [3,4]. It is important to note that conventional cytological evaluation of pleural fluids has a relatively low sensitivity and thus limited diagnostic accuracy for the diagnosis of malignant pleural effusions [5]. We report a patient with a pleural vascular tumor who had conflicting clinical and laboratory evidence, negative cytological result, and a misleading initial pathology report.

Case Report

A 79-year-old female patient presented with progressive dyspnea and productive cough for the past three months. She also complained of right flank pain. On admission, vital signs were stable and oxygen saturation was 97% on room air with no respiratory distress. Physical examination revealed decreased breath sounds over the right lung. Laboratory tests were unremarkable, except for CRP:10 mg/L and ESR:56 mm/h. In echocardiography, EF was reported as 55%. Chest X-ray demonstrated a massive right-sided pleural effusion (Figure1), along with an air–fluid level and mediastinal pleural thickening. The patient underwent a CT scan at another center, which revealed severe right-sided effusion with smooth pleural thickening. Diagnostic and therapeutic thoracentesis was also performed without a specific diagnosis.

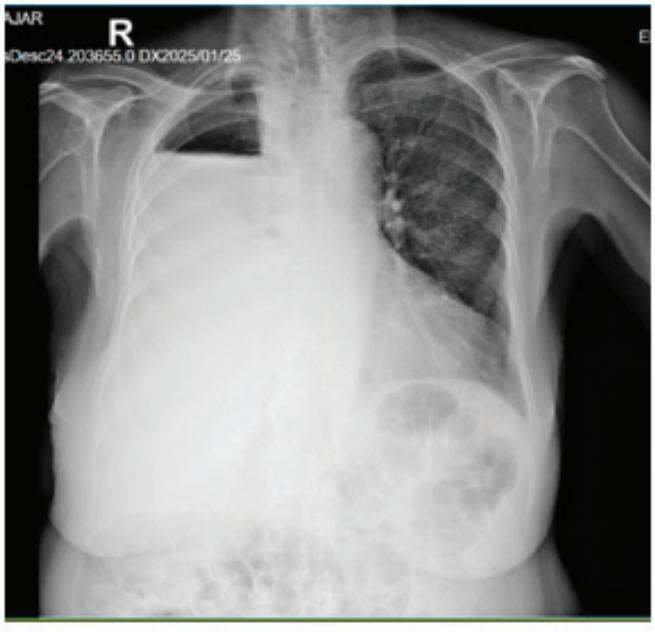

Figure 1: Massive right-sided pleural effusion with an air-fluid level and mediastinal pleural thickening.

The patient underwent diagnostic thoracentesis and the results obtained included: Protein:5 gr/dl, Glucose:125 mg/dl, LDH:245 IU/L, RBC:2200/µL, WBC:110/µL (Lymph:30%, PMN:70%) with serum protein and LDH of 8.4 gr/dl and 307 IU/L, respectively. Fluid bacteriological culture and ADA and cytology were reported negative. Since there was an inconsistency between the patient's clinical presentation and laboratory results, the patient underwent medical thoracoscopy that revealed markedly thickened pleura with rough surface and dense fibrinous deposits (Figure 2). Multiple biopsies were obtained from the parietal pleura. Initial microscopic examination was reported as severe chronic inflammation with fibrous thickening and mesothelial hyperplasia.

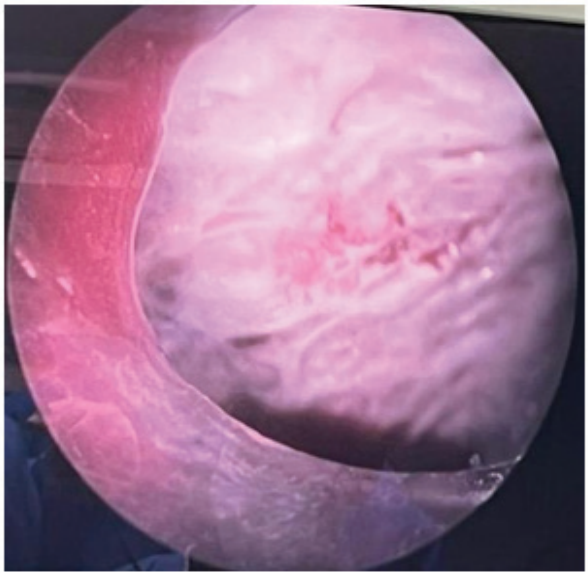

Figure 2: Thoracoscopic view with markedly thickened pleura with rough surface and dense fibrinous deposits.

Considering the non-specificity of the result, a pathology review was requested. On subsequent review of the pathology slides, it was reported infiltration of cleft like spaces lined by cuboid uniform epithelial cells within hyalinized stroma. Immunohistochemistry (IHC) staining was positive for AE1, AE3, CD31, CD34, CD10 and D2-40. These findings are in favor of final diagnosis of *pleural epithelioid hemangioendothelioma* that is a tumor of vascular origin rarely encountered in the thorax.

Discussion

Epithelioid Hemangioendothelioma (EHE) is a rare vascular tumor originates from vascular endothelial or pre-endothelial cells characterized histopathologically by the features of epithelioid and histiocytoid cells. In addition to the lungs, EHE in the chest also involves the pleura and mediastinum [6]. In contrast to EHE that affects women more than men and generally peaks in the fourth or fifth decade of life, pleural EHE is more common in older men. The severity of its clinical behavior is somewhere between the benign nature of hemangiomas and the highly aggressive angiosarcomas [7,8]. Common clinical symptoms of pleural EHE include dyspnea, pleuritic chest pain, cough and sputum production, and weight loss. Less frequent symptoms included hemoptysis, shoulder or back pain, constitutional symptoms (fever, night sweating, fatigue), nausea and vomiting, hypoxemia, and hoarseness. Typical radiologic manifestations of pleural EHE include pleural effusion and nodular or smooth pleural thickening [8]. There is still no standard treatment for pleural EHE and various treatments including chemotherapy, radiotherapy, and surgery have been performed on these patients, although associated with a lack of response and mortality rates of up to 80% [9].

Conclusion

In cases of pleural effusion with an unknown cause, paying attention to the clinical presentation and evaluating laboratory and pathological results based on it can be helpful in determining the cause of the effusion, especially in cases where laboratory and pathologic results are inconsistent with the patient's presentation.

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