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Caso clinico / *Case report*

Diagnosis of pulmonary artery sarcoma with esophageal endoscopic ultrasound-guided needle aspiration

Diagnosi di sarcoma dell'arteria polmonare con agoaspirati transesofagei sotto guida ecoendoscopica

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Summary

Intraluminal sarcoma of the pulmonary artery is a rare malignant neoplasm with aggressive behaviour. Differential diagnosis with pulmonary embolism, metastasis or even pseudotumors can be complex and the site of the tumour can pose a significant diagnostic challenge, as the pulmonary arteries are not easily accessible for diagnostic sampling. We describe a case of primary pulmonary artery sarcoma diagnosed with endoscopic ultrasound-guided needle aspiration/biopsy through transesophageal access.

Key words: EUS, FNA, primary pulmonary sarcoma, echoendoscopy

Riassunto

Il sarcoma dell'arteria polmonare è una rara neoplasia maligna dal comportamento clinico aggressivo. La diagnosi differenziale con embolia polmonare o metastasi, o addirittura pseudotumori può non essere facile da ottenere e può necessitare di un approccio molto invasivo. Presentiamo il caso di un paziente in cui abbiamo ottenuto la diagnosi di sarcoma primitivo dell'arteria polmonare con agobiopsie condotte sotto guida ecografica e con approccio transesofageo.

***Parole chiave**: EUS, FNA, sarcoma primitivo dell'arteria polmonare, ecoendoscopia*

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Conflitto di interessi
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Introduction

Primary pulmonary artery sarcoma (PPAS) is a rare malignant neoplasm which arises from the mesenchymal cells of the pulmonary artery with ominous prognosis regardless of treatment options [1]. On computed tomography (CT)-angiography or magnetic resonance imaging (MRI), PPAS causes an intraluminal filling defect of the pulmonary artery that resembles pulmonary emboli, metastatic angiosarcoma [2], or inflammatory pseudotumor [3]. Even if PPAS is considered as a differential diagnosis, acquiring pathological confirmation can be challenging.

Case presentation

A 54-year-old patient with no prior medical history was admitted to our

ward after a CT scan showed a partly cavitated left superior lobe consolidation, with an endoluminal mass in the pulmonary trunk and left pulmonary artery (Fig. 1: a,b,d,e). The endoluminal lesion appeared hypodense at the contrasted CT scan and suspicion of a neoplastic mass with secondary lesions in the upper lobe was raised. A positive positron emission tomography - computed tomography (PET-CT) scan supported this hypothesis, showing an increased fluorodeoxyglucose (FDG) uptake in both the upper left lobe, with maximal standardized uptake value (SUVmax) 9.8, and the lesion of the pulmonary artery, with SUVmax 5.

The patient had accessed the emergency ward because of a recent onset of cough, fever and a slight high-back pain while coughing, with no benefit from a 5-day antibiotic treatment with clarithromycin. He had no smoking history and was physically fit, working as a fireman. He was haemodynamically stable, his Blood Gas Analysis (BGA) was normal, blood tests revealed only slight neutrophilia and elevated C-reactive protein (CRP): 250 mg/L. Echocardiography showed a normal left ventricle ejection fraction (FE 65%), with no right-side overload.

For diagnostic purposes, we performed trans-esophageal endoscopic ultrasound-guided fine needle aspiration/biopsy (EUS-FNA/B) on the endo-arterial mass in the left pulmonary artery with an Olympus GF-UCT 160 echoendoscope and a 19G needle (Fig. 1c). The procedure was performed in deep sedation with anaesthesiologic assistance, without need for intubation. On echo-analysis, the lumen of the left pulmonary artery branch appeared completely obliterated by hypoechoic tissue. The samples resulted diagnostic for mixoid stroma-rich spindle cell sarcoma (Fig. 1f). Murine Double Minute Clone 2 (MDM2) expression on neoplastic cells was documented by immunohistochemistry and confirmed the diagnosis of PPAS.

Discussion

We present an unusual case of PPAS that was diagnosed with EUS-FNA. PPAS is a rare disease and, even when the hypothesis is inferred, differential diagnosis with chronic pulmonary embolism or other rare tumors can be challenging. CT can help differentiate PPAS from PE by indicating a low-attenuation filling defect occupying the entire luminal diameter of the proximal or main pulmonary artery, expansion of the involved arteries, or extraluminal tumor extension and PET-FDG avidity with high SUVmax is consistent with a neoplastic lesion[4]. To obtain a conclusive diagnosis, however, endoscopic strategies like those described in this case report can be extremely valuable. Although experience is limited, as the disease is rare, endobronchial ultrasound-guided trans-bronchial needle aspiration/biopsy (EBUS-TBNA/B) for the diagnosis of intravascular mediastinal lesions appears to be feasible, safe, and minimally invasive according to a review of available literature [5,6]; in a recent study aimed at evaluating transvascular EBUS-TBNA/B of intrathoracic lesions (in which in 33 cases the transversed vessel was the pulmonary artery or its

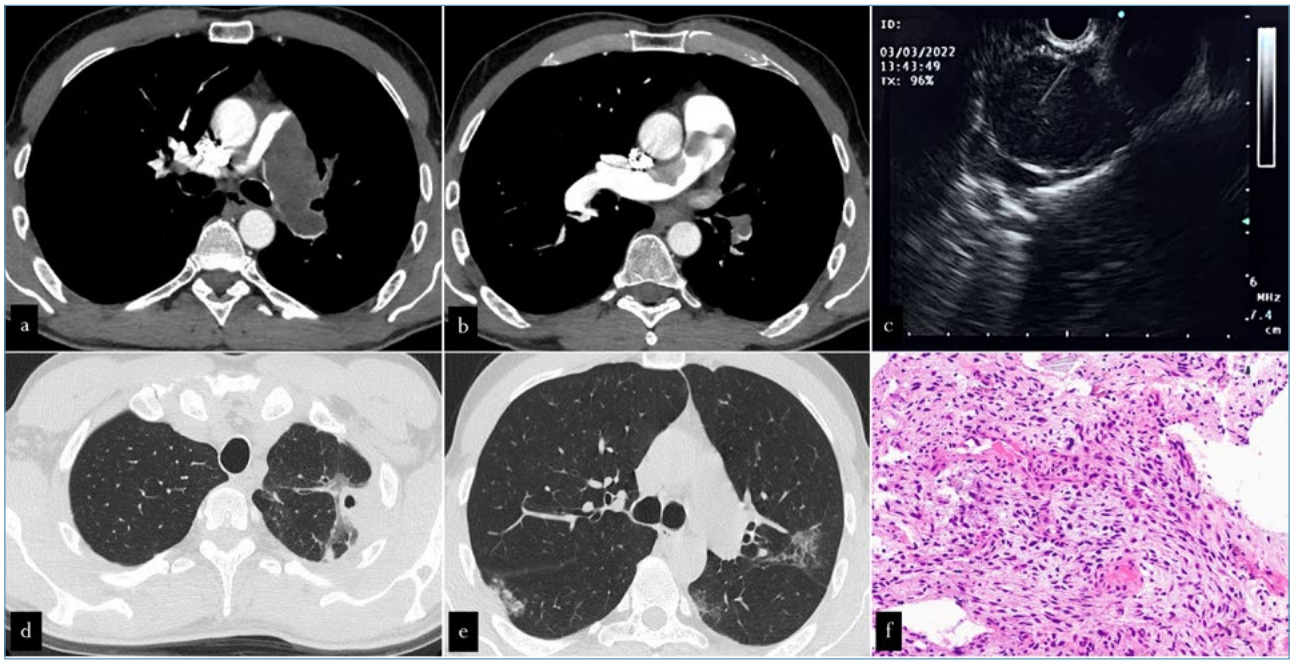

Figure 1. CT scan showing the endo-arterial lesion (a, b), cavitated upper lobe lesion (d), ground-glass lesions (e); EUS-FNA on endo-arterial lesion (c), hematoxylin and eosin-stained specimen showing atypical spindle cells in a mixoid stroma (f).

main branches) none of the patients had intraoperative or immediate postoperative complications [7], supporting the notion that piercing the pulmonary artery with ultrasound-guided techniques can be a safe diagnostic strategy. We found some case reports of EBUS-TBNA/B performed for diagnosing pulmonary artery intravascular lesions [5,8-10], but none regarding EUS-FNA/B.

EUS-FNA/B can be acceptable alternative to more invasive procedures such as catheter suction biopsy in the diagnosis of primary pulmonary artery sarcoma, avoiding the potential delay in diagnosis, and should be considered in the diagnostic approach to intravascular masses of the pulmonary artery.

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