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Diagnosis of Pulmonary Artery Sarcoma with Multi-modality Imaging

Pulmoner Arter Sarkomunun Multi-modalite Görüntüleme ile Değerlendirilmesi

CASE REPORT
OLGU SUNUMU

ABSTRACT

Pulmonary artery sarcoma is an extremely uncommon malignancy with a poor prognosis. It is often difficult to distinguish it from pulmonary thromboembolic disease because of nonspecific signs and symptoms as well as similar imaging findings. We present a 46-year-old man who had initially been diagnosed with presumed asthma that later proved to be pulmonary artery sarcoma. The patient was evaluated with multi-modality imaging studies which showed a mass in the pulmonary artery, its extension, mobility and invasion, and attachment to the artery wall. Pulmonary artery mass was excised and pulmonary artery endarterectomy was performed. The histopathological diagnosis was undifferentiated sarcoma with pleomorphic morphology.

Keywords: Multi-modality imaging, pulmonary artery sarcoma, thromboembolism

ÖZET

Pulmoner arter sarkomu, kötü prognozlu seyreden, son derece nadir görülen bir malignitedir. Spesifik olmayan belirti ve bulguların yanı sıra benzer görüntüleme bulguları nedeni ile pulmoner tromboembolik hastalıktan ayrımı zordur. Bu olguda, başta astım tanısı alan, daha sonra PAS tanısı koyulan, 46 yaşında bir erkek hastayı sunmaktayız. Hastada pulmoner arterde kitle, yayılımı, mobilitesi ve invazyonu, arter duvarına tutunması, multi-modalite görüntüleme ile değerlendirildi. Pulmoner arter kitlesi eksize edilerek pulmoner endarterektomi yapıldı. Histopatolojik olarak pleomorfik morfolojiye sahip farklılaşmamış sarkom tanısı koyuldu.

Anahtar Kelimeler: Multi-modalite görüntüleme, pulmoner arter sarkomu, tromboembolizm

Pulmonary artery sarcoma (PAS) is a rare and aggressive malignancy, usually presenting with dyspnea, cough, chest pain, and hemoptysis. Its diagnosis is often missed or delayed due to these nonspecific symptoms. It can mimic pulmonary hypertension, various pulmonary thromboembolic (PTE) diseases, and pneumonia. Early diagnosis and radical surgery are the only proven therapeutic options for survival.[1,2] We present a case of PAS initially mistaken for asthma.

Case Report

A 46-year-old man presented to the cardiology outpatient clinic with a 6-month history of nonproductive cough and dyspnea. Three months earlier, he had received a treatment for presumed asthma but his symptoms persisted. Electrocardiography showed sinus tachycardia at a rate of 120 bpm and his chest x-ray was normal. Transthoracic echocardiography (TTE) showed an ejection fraction of 60%, mild to moderate tricuspid regurgitation, and a pulmonary artery systolic pressure (PAPs) of 70 mm Hg with a normal right ventricular function and dimension. A small pericardial effusion surrounding the heart was also noted. Contrast-enhanced computed tomography (CT) of the thorax showed a pulmonary artery mass causing a filling defect without parencymal involvement (Figure 1A, 1B). Transesophageal echocardiography (TEE) was performed to evaluate pulmonary hypertension. In the aortic short-axis view (SAW), the main pulmonary artery was dilated, with no pathology. However, in the upper esophageal position, a color turbulance in the main pulmonary artery was noted. The aortic arch SAW (45°) revealed a mass 61 × 32 mm in size, attached to the artery wall, and extending to the right pulmonary artery from the main pulmonary

Büşra Güvendi Şengör[1], **M.D.**
Alev Kılıçgedik[1], **M.D.**
Tuba Unkun[1], **M.D.**
Emine Bozkurtlar[2], **M.D.**
Ahmet Erdal Taşçı[3], **M.D.**
Serpil Taş[4], **M.D.**
Cevat Kırma[1], **M.D.**

[1]*Department of Cardiology, Kartal Koşuyolu Training and Research Hospital, İstanbul, Turkey*
[2]*Department of Pathology, Marmara University School of Medicine, İstanbul, Turkey*
[3]*Department of Thoracic Surgery, Kartal Koşuyolu Training and Research Hospital, İstanbul, Turkey*
[4]*Department of Cardiovascular Surgery, Koşuyolu Training and Research Hospital, İstanbul, Turkey*

Corresponding author:
Büşra Güvendi Şengör
✉ guvendi2861@hotmail.com

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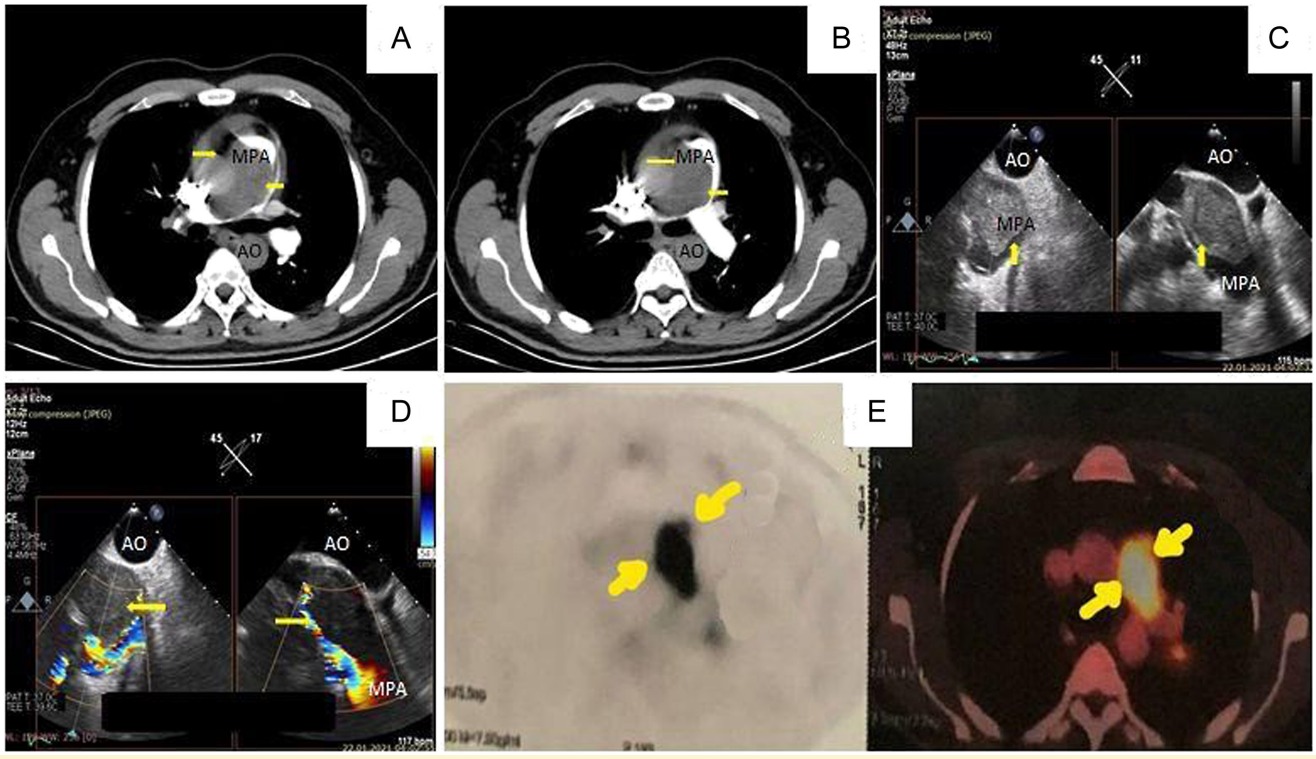

Figure 1. A, B. Contrast-enhanced computed tomography (CT) of the thorax showed a pulmonary artery mass (arrows) causing a filling defect. C. Transesophageal echocardiography (TEE) revealed a mass (arrows), attached to the pulmonary artery wall. D. Color flow doppler images of the pulmonary artery was seen. E. Fluorodeoxyglucose positron emission tomography (FDG-PET) showed a mass of soft tissue density with hypermetabolism (yellow arrows). Written informed consent was obtained from the patient. AO, aorta; MPA, main pulmonary artery.

artery (Figure 1C, 1D; Video 1,2,3*). PAS or chronic thromboembolic pulmonary hypertension was considered in the differential diagnosis. T2-weighted magnetic resonance imaging (MRI) showed hyperintensity, and fluorodeoxyglucose positron emission tomography (FDG-PET) showed a mass of soft tissue density with hypermetabolism, findings suggestive of an angiosarcoma (Figure 1E). The patient underwent excision of the pulmonary artery mass and pulmonary artery endarterectomy (Figure 2A, 2B). After an uneventful postoperative course, he was discharged at day 7. Histopathological examination showed spindle neoplastic cells presenting with pleomorphism and extensive necrotic areas and an increased mitotic activity (Figure 2C, 2D, 2E). The Ki-67 proliferation index of the tumor was 50%. The neoplastic cells were positive for vimentin and negative for pankeratin, desmin, smooth muscle actin (SMA), S100, CD34, CD31, and PDGFR-α. Neither a histomorphologic nor immunohistochemical differentiation could be made. Fluorescence in situ hybridization for MDM2 amplification was also negative. The pathological diagnosis of the tumor was made as an undifferentiated sarcoma with pleomorphic morphology. Chemotherapy and radiotherapy were scheduled.

ABBREVIATIONS

CT	Computed tomography
FDG-PET	Fluorodeoxyglucose positron emission tomography
MRI	Magnetic resonance imaging
PAPs	Pulmonary artery systolic pressure
PAS	Pulmonary artery sarcoma
PTE	Pulmonary thromboembolic
SAW	Short-axis view
SMA	Smooth muscle actin
TEE	Transesophageal echocardiography
TTE	Transthoracic echocardiography

Discussion

Primary pulmonary artery tumors are rare, with an estimated incidence of 0.001%-0.03%.[3] PAS, originating from the intimal layer of the pulmonary artery, is the most common primary tumor of the pulmonary artery, with a poor prognosis as well as a median survival of 17 months. Patients with PAS are usually asymptomatic till the occlusion of the artery. It is often misdiagnosed as thromboembolic pulmonary hypertension because of nonspecific symptoms such as dyspnea and chest pain.[2] Since the radiographic findings mimic those of PTE disease, thrombolytic therapy is inappropriately administered in many cases before the diagnosis of PAS. Therefore, multi- modality imaging may be useful (Table 1).[4] Computeed tomographt can depict the extention of the tumor and differentiate PAS from PTE. Yi et al[5] reported that CT findings such as a low-attenuation filling defect occupying the entire luminal diameter of the proximal or main PA, expansion of the involved arteries, and extraluminal extension of the tumor favored the diagnosis of PAS. A hyperintense filling defect in the main pulmonary artery on T2-weighted MRI and hypermetabolism on FDG-PET is suggestive of PAS.[6,7] TTE can also

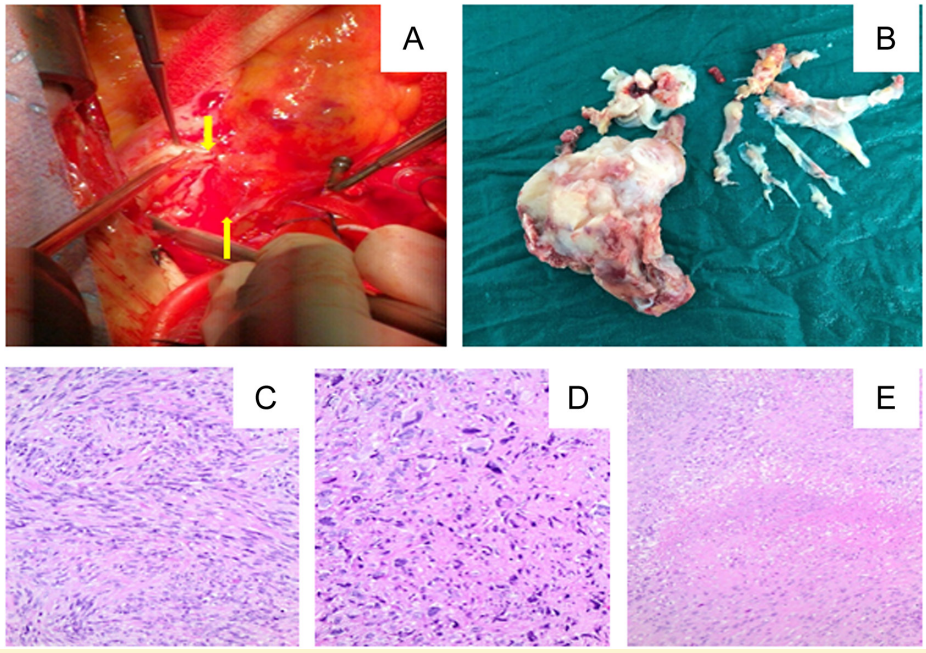

Figure 2. A. Intraoperative image of PAS (arrows) was noted. B. Resected specimen was seen. C, D, E. Histopathological examination revealed spindle neoplastic cell (2C; HE, 20×), pleomorfic spindle neoplastic cells (2D; HE, 20×) and necrotic areas of the tumor (2E; HE, 10×). Written informed consent was obtained from the patient. PAS, pulmonary artery sarcoma.

provide valuable information to differentiate PAS from PTE.[8] Mass mobility, attachment to the PA wall or pulmonic valve, the absence of echolucent areas, and bulging rather than linear morphology favor PAS. TTE can show planar invasion of the tumor into the arterial wall.[1] Immunohistochemical staining is the gold standard for the definitive diagnosis. The tumor may express vimentin, SMA, desmin, and endothelial cell marker such as CD31 and MDM2.[9] Surgery, including pulmonary artery endarterectomy, tumor debulking, pneumonectomy, and lobectomy, is the mainstay of the treatment for a better long-term outcome. Postoperative chemotherapy and radiotherapy may provide survival benefits.[2,10]

In our case, findings of CT, MRI, and FDG-PET were consistent with PAS. TTE revealed pulmonary hypertension with a PAPs of 70 mm Hg. TTE provided no additional information but TEE did.

Table 1. Radiologic Findings of Pulmonary Artery Sarcoma and Pulmonary Thromboembolism

	Pulmonary Artery Sarcoma	Pulmonary Thromboembolism
TTE	Inhomogeneous parenchymal filling in the pulmonary arteries Pulmonary hypertension, enlargement of the right heart and enlargement of the right ventricle Mobility, absence of echo lucent areas, bulging rather than linear morphology, and attachment to the pulmonary valve or pulmonary artery wall	Uniform echo of fresh thrombus Pulmonary hypertension, enlargement of the right heart and enlargement of the right ventricle
TEE	Planar invasion of the tumor into the arterial wall Mobility, absence of echo lucent areas, bulging rather than linear morphology	Sessile, mural, immobile, echodense, partially obstructive masses (CTEPH) Mobile, snake-like appearance (acute PTE)
CT	Heterogeneous enhancement Expansive growth and a bulging appearance against the direction of blood flow, a lobulated structure at the proximal part of the tumours, a grape-like appearance at the distal end of the tumour Presence of "wall eclipsing sign" Pulmonary nodules, parenchymal changes, hilar adenopathy, pleural effusions	Uniform density of filling defect Straight and cup-like structure (caused by blood flow against the surface of blood clots) Absence of "wall eclipsing sign" Wedge shaped infarct, pleural effusion
MRI	Non-homogeneous delayed enhancement Higher T2 signal intensity The grape-like appearance	Different signal intensities related to various stages of thrombus Calcifications
PET-FDG	Higher maximal standardized uptake value (SUVmax) (10.2 ± 10.8) - hypermetabolism	Lower SUVmax uptake (1.7 ± 0.3)

TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; CT, computed tomography; MRI, magnetic resonance imaging; FDG-PET, fluorodeoxyglucose positron emission tomography; SUV, standard uptake value.

In the upper esophageal position, the aortic arch SAW (45°) revealed the mass and its extension, mass mobility, attachment to the artery wall, the absence of echolucent areas, and bulging. These finding ruled out PTE disease, allowing the diagnosis of PAS.

Conclusion

The diagnosis of PAS may be challenging, requiring early diagnosis and radical surgery. It should be kept in mind that PAS can manifest as nonspecific symptoms and be misdiagnosed as PTE disease. When a high clinical suspicion of PAS is present, patients should be evaluated by multi-modality imaging.

*Supplementary video files associated with this article can be found in the online version of the journal.

Informed Consent: Written informed consent was obtained from the patient.

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