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CASE REPORT

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Pulmonary artery sarcoma treated with pulmonary endarterectomy and leaflet reconstruction using Ozaki technique

Gokhan Arslanhan MD[1] | Koray Ak MD[1] | Fatih Öztürk MD[1] | Alperen Türker MD[1] | Emine Bozkurtlar MD[2] | Sinan Arsan MD[1] | Bedrettin Yıldızeli MD[3]

[1]Department of Cardiovascular Surgery, Marmara University School of Medicine Pendik Research and Training Hospital, Istanbul, Turkey

[2]Department of Pathology, Marmara University School of Medicine Pendik Research and Training Hospital, Istanbul, Turkey

[3]Department of Thoracic Surgery, Marmara University School of Medicine Pendik Research and Training Hospital, Istanbul, Turkey

Correspondence
Gokhan Arslanhan, MD, Department of Cardiovascular Surgery, Marmara University Pendik Research and Training Hospital, Muhsin Yazıcıoglu Caddesi No. 10 Pendik Istanbul 34890, Turkey.
Email: dr.gokhanarslanhan@gmail.com

Abstract

Pulmonary arterial intimal sarcomas (PAIS) are rare malignancies with a poor prognosis. Sarcomas present with signs and symptoms mimicking pulmonary thromboembolic disease, delaying the diagnosis. We present a 29-year-old male patient diagnosed with PAIS in the right and main pulmonary arteries extending to the left pulmonary leaflet. The patient was treated with pulmonary endarterectomy and pulmonary leaflet reconstruction using the Ozaki technique.

KEYWORDS

aorta and great vessels, cardiovascular pathology, valve repair/replacement

1 | INTRODUCTION

Pulmonary arterial intimal sarcomas (PAIS) are rare malignancies with an incidence of 0.001%–0.03%.[1] Due to their rare occurrence, the genetic and environmental risk factors are not clear and a consensus is lacking for its treatment. Surgery, chemotherapy, and radiotherapy are all applied with a poor prognosis.

Sarcomas present with signs and symptoms mimicking pulmonary thromboembolic disease, causing delays in diagnosis.[1,2] With the current Covid-19 pandemic, venous thromboembolism is seen often during hospitalization and after discharge with clinicians vigilant for their early detection.[3] We present a case of PAIS initially diagnosed with Covid-19 associated pulmonary embolism treated with pulmonary endarterectomy (PEA) and pulmonary valve repair using the Ozaki technique.

Approval was obtained from the patient for this case report and International Review Board was waived.

2 | CASE REPORT

A 29-year-old male patient who recovered from Covid-19 infection 3 months ago presented with increasing dyspnea and pleuritic chest pain. Computed tomography (CT) showed filling defects in the pulmonary vasculature on the right and left lower lobes consistent with chronic thromboembolic disease. Referred to our center for PEA operation, the patient was NYHA class III with resting oxygen saturation of 93% on ambient air. The D-Dimer was 0.56 mg/L (upper limit 0.05 mg/L). Transthoracic echocardiography revealed pulmonary hypertension with a peak pulmonary pressure of 88 mmHg and an enlarged right ventricle of 42 mm in diameter. In addition, a mass was seen on the pulmonary valve. On chest CT, a lobulated mass obstructing the right and main pulmonary arteries, and extending onto the pulmonary valve causing right ventricular outflow tract obstruction was seen (Figure 1A). Positron emission tomography-CT (PET-CT) was performed on suspicion of malignancy which suggested

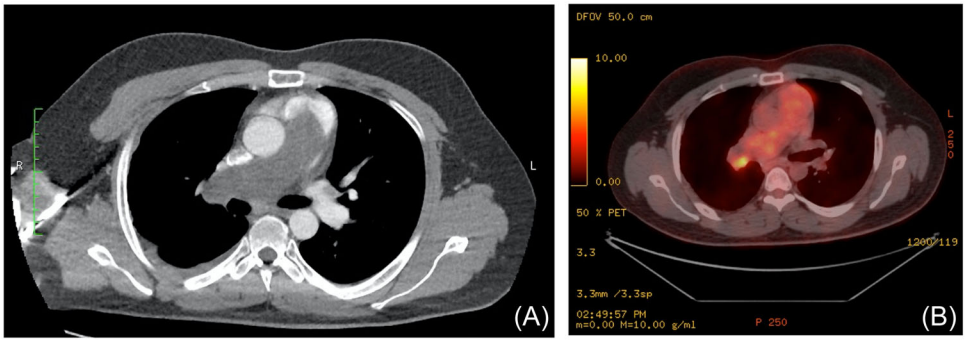

FIGURE 1 Contrast enhanced thoracic CT (A) and PET-CT (B) of the pulmonary arterial intimal sarcoma. CT, computed tomography; PET-CT, positron emission tomography-CT

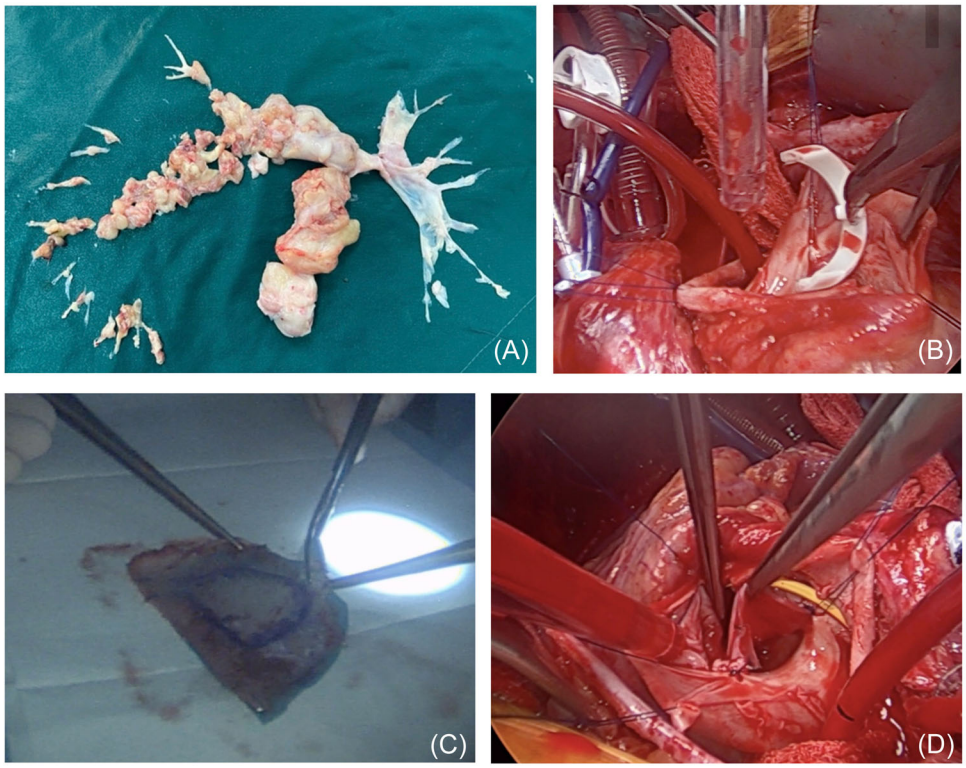

FIGURE 2 (A) The tumoral mass and chronic thrombi removed with pulmonary endarterectomy. The mass extending to the right ventricular outflow tract covers the left pulmonary valve lying posterior to the mass. The Ozaki sizer (B) and template (C) used for leaflet reconstruction. (D) The reconstructed pulmonary leaflet (held with forceps)

tumoral activity with high fluorodeoxyglucose uptake (SUV max 10.34) at the main and right pulmonary arteries (Figure 1B). A decision for PEA was made because of suspected pulmonary artery sarcoma, the patient's NYHA III functional class, pulmonary hypertension, and lack of significant comorbidities.

Our surgical approach has been reported previously.[4] A longitudinal incision from the main pulmonary artery towards the right pulmonary artery revealed a lobular yellow-white mass obstructing the pulmonary arteries. A dissection plane between the intima and media layers of the pulmonary artery was used to continue the endarterectomy to completely remove the mass distally within the right pulmonary tree (Figure 2A). The tumor was found to invade the left pulmonary valve leaflet and the infundibular septum. The left pulmonary leaflet and part of the infundibular septum was excised with the mass. The pulmonary valve leaflet was repaired with the Ozaki technique using glutaraldehyde-treated autologous pericardium. The commissural distance of the resected leaflet was measured using the Ozaki sizer (Figure 2B) and the pericardium was tailored using the Ozaki template (Figure 2C). Sufficient valve coaptation was observed (Figure 2D) with trivial pulmonary insufficiency on transesophageal echocardiography.

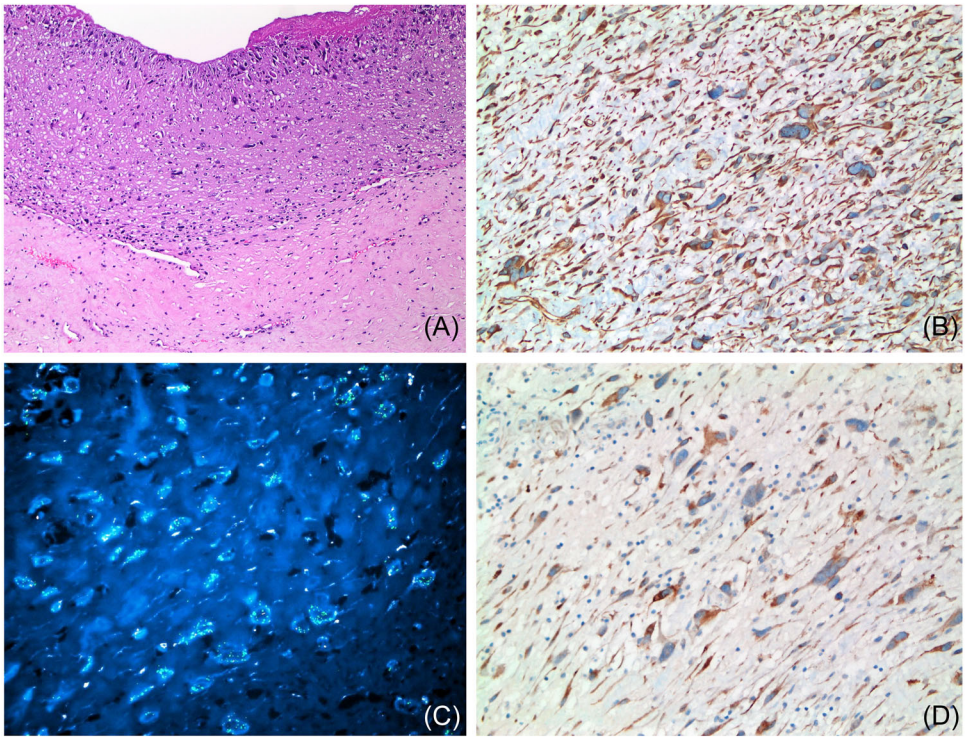

FIGURE 3 (A) Hematoxylin-eosin stained spindle shaped and pleomorphic neoplastic cells showing endoluminal growth). (B) Vimentin positive neoplastic cells. (C) Amplification of mouse double minute 2 gene and (D) positivity with platelet derived growth factor receptor alpha immunohistochemical stain

Histopathologically, neoplasti cells were spindle-shaped and pleomorphic. Endoluminal growth of the tumor was seen (Figure 3A). Neoplastic cells were positive for vimentin and negative for pankeratin and desmin (Figure 3B). The proliferation index with Ki67 was 60%. Amplification of mouse double minute 2 gene and positivity with platelet derived growth factor receptor alpha immunohistochemical stain were observed (Figure 3C,D). The diagnosis of an intimal sarcoma was made. The resected leaflet was free of neoplastic cells outside of the area with tumor invasion.

The patient has received two rounds of chemotherapy with ifosfamide and doxorubicin. Radiotherapy is planned after completing four rounds of chemotherapy. The patient is currently doing well with NYHA I symptoms 6 months postoperatively.

3 | COMMENT

Pulmonary artery sarcomas are rare malignancies arising from mesenchymal cells of the tunica intima. Survival with radiotherapy and chemotherapy is reported at 1.5 months without surgical debulking which improves to 10 months with incomplete resection and 36.5 months with complete resection.[5,6] Therefore, surgical debulking is indicated in patients who can tolerate surgery. These tumors have been removed with pneumonectomy, however, recent recommendations have evolved to near-complete tumor excision with PEA in experienced centers, providing the benefit of reduced pulmonary pressures in cases without distant metastases.[1,2,6] The residual tumoral microembolism in the distant pulmonary arterial tree can be treated with adjuvant chemotherapy and radiotherapy. Our patient was assessed as low risk high benefit for surgery and was recommended debulking surgery with PEA.

Clinicians are wary of the thromboembolic complications of the Covid-19 disease which are considered promptly in the differential diagnosis of patients with a recent history of Sars-Cov-2 infection. Patients with PAIS present with clinical and radiological findings that resemble pulmonary thromboembolism, leading to delays in true diagnosis.[1] Characteristic findings of PAIS include filling defects with low attenuation in the main and proximal pulmonary arteries, enlargement of artery segments, extraluminal presence of tumoral mass, and right ventricular outflow tract obstruction with pulmonary valvular involvement in some cases.[1,5,7] The diagnosis can be confirmed radiologically with PET-CT or gadolinium-enhanced MRI.[2,5]

Sarcomas primarily progress with intravascular invasion. Pulmonary valve or right ventricular involvement is reported in 30% of PAIS cases.[2,5] Per the classification by Han et al.,[2] our case had type II involvement with retrograde extension to the pulmonary valve. When the pulmonary valve needs to be resected due to invasion of the sarcoma, replacement of the outflow tract and the valve with a homograft can be considered.[5] Without an available homograft, the Ozaki technique is a

reasonable method of valvular reconstruction which avoids the adverse events associated with prosthetic valves. PAIS is an aggressive sarcoma and priority should be given to the completeness of tumor resection. If the surgeon is confident that invasion is limited to a single leaflet, the described surgical method may be considered to prevent the adverse events associated with prosthetic valves. Our patient had only the involvement of the left leaflet which was excised and reconstructed with autologous pericardium, preserving the remaining native leaflets. The Ozaki technique has been performed for pulmonary valve infective endocarditis, and we achieved good results in our case with PAIS.[8]

The diagnosis of PAIS is challenging because the disease may mimic chronic thromboembolic pulmonary hypertension. Suspicion should be aroused when anticoagulant treatment is unsuccessful. PEA is indicated both for definitive histologic diagnosis of sarcoma and for symptomatic benefit of patients which is a complex procedure that should be performed in experienced high-volume centers with a multidisciplinary team.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

ORCID

Gokhan Arslanhan http://orcid.org/0000-0002-6846-1589
Koray Ak http://orcid.org/0000-0002-8608-5951

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