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CASE REPORT
Other

Complete resection of a pulmonary artery sarcoma involving the pulmonary valve and right ventricle outflow tract: a case report

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Background Pulmonary artery sarcoma (PAS) is an extremely rare tumour, preferably treated by surgery. However, the surgical management remains largely debatable, as only less than half of patients with PAS can undergo thorough excision.

Case summary A 32-year-old man with a tumour involving the right ventricle outflow tract, pulmonary trunk extending into the bifurcation, and right pulmonary arteries underwent complete resection using a homologous pulmonary valve and vascular grafts for reconstruction, combined with right pulmonary endarterectomy (PEA) for potential seeding metastasis. Histopathological examination demonstrated undifferentiated pleomorphic sarcoma with surgical margins free of disease. The patient remains asymptomatic, and follow-up computed tomography 5 months after surgery indicated no recurrence or metastasis.

Discussion Radical resection of a PAS with reconstruction using pulmonary valve allograft and polytetrafluoroethylene vascular grafts is technically feasible and successful. Additionally, PEA may eliminate the potential intima implantation metastasis.

Keywords Pulmonary artery sarcoma • Surgery • Complete resection • Case report

ESC Curriculum 4.10 Prosthetic valves • 6.8 Cardiac tumours • 7.5 Cardiac surgery • 9.5 Pulmonary thromboembolism

Learning points

- Pulmonary artery sarcoma (PAS) is a rare tumour with a catastrophic prognosis.
- Surgery remains the mainstay of treatment for PAS; however, only less than half of patients can undergo complete resection intra-operatively.
- When an initial or recurrent tumour involves only the right ventricle outflow tract and the main pulmonary artery trunk, radical resection with reconstruction using pulmonary valve allograft and polytetrafluoroethylene vascular grafts is feasible and safe, whereas pulmonary endarterectomy may eliminate potential intimal implantation metastasis.

Introduction

Since the first report on pulmonary artery sarcoma (PAS) by Mandelstamm in 1923, <500 cases of PAS have been published so far.[1,2] The mainstay of treatment for PAS is surgery; however, less than half of patients with PAS are suitable for complete resection intra-operatively.[2] As previously published,[1,3,4] the median survival after PAS is 17–36 months even after surgery, since most patients develop local tumour recurrence or distant metastases. We report the case of a patient with PAS undergoing complete resection followed by a reconstruction of the right ventricle outflow tract (RVOT) and pulmonary arteries.

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Timeline

Time	Events
2021 May	• The patient was initially diagnosed with a large pulmonary embolus at a local hospital • He received oral anticoagulant therapy with no improvement in symptoms
2021 July	• He was referred to our institution • Pre-operatively, his suspected diagnosis was pulmonary artery sarcoma, as the positron emission tomographic computed tomography (PET–CT) scan revealed increased fluorodeoxyglucose uptake in the right ventricle outflow tract and pulmonary artery trunk
2021 July	• The tumours were successfully resected, and reconstruction was done using a homologous pulmonary valve and polytetrafluoroethylene vascular grafts, combined with right pulmonary endarterectomy, for potential seeding metastasis.
2021 August	• He was discharged on post-operative day 7 in a satisfactory condition • Histopathological examination demonstrated undifferentiated pleomorphic sarcoma with surgical margins free of disease
2021 December	• He remains alive with no symptoms; follow-up CT indicated no recurrence or metastasis

Case presentation

A previously healthy 32-year-old man presented to a local hospital with dyspnoea upon exertion for 1 week in May 2021. He had no fever, no oral or genital ulcers, no oedema in the lower extremities, no related genetic history in the family, and no past medical history. On physical examination, he had normal respiratory sounds, and cardiac auscultation revealed a Grade 3/6 systolic ejection murmur on the left upper sternal border. Laboratory examination for cardiac biomarkers (cardiac troponin and N-terminal pro-B-type natriuretic peptide), coagulation function tests, antiphospholipid antibodies, lupus antibodies, and tumour markers were negative. He was initially diagnosed with a large pulmonary embolus on imaging (*Figure 1A*), for which he received oral anticoagulation therapy (rivaroxaban—15 mg twice a day for the first 21 days and 20 mg once a day afterward); however, there was no improvement.

Subsequently, he was referred to our institution in July 2021. Transthoracic echocardiography (*Video 1*) showed a large mass of 47 × 18 mm in the pulmonary artery with a partial occlusion of the RVOT and surrounding the lateral pulmonary valve. The blood flow in the residual pulmonary artery was significantly accelerated, with a peak flow rate of 4.0 m/s and a peak pressure gradient of 64 mmHg across the valve. Re-evaluation using computed tomography (CT) revealed filling defects in the RVOT, the pulmonary trunk extending into bifurcation, and the right pulmonary arteries (*Figure 1B*). A positron emission tomographic CT scan revealed increased fluorodeoxyglucose uptake with a maximum standardized uptake value (SUV_{max}) of 15.6 in the RVOT and pulmonary artery trunk and no metastases to other structures (*Figure 1C and D*). The pre-operative suspected diagnosis was pulmonary artery malignancy.

Through a median sternotomy with hypothermic cardiopulmonary bypass, the tumour was excised, and the pulmonary valve, a part of the RVOT, the main pulmonary artery, and the bilateral pulmonary trunk, including the bifurcation, were resected (see Supplementary material online, *Video S1*; *Figure 2A and B*). Pulmonary endarterectomy (PEA) was undertaken on the right side with deep hypothermic circulatory arrest (9 min). A polytetrafluoroethylene (PTFE) vascular graft was used, and end-to-end anastomosis was performed between the tube and the native bilateral pulmonary arteries. Pulmonary root reconstruction was performed using a pulmonary allograft—a 22 mm homologous pulmonary valve—along with a part of the RVOT and the main pulmonary artery; end-to-side anastomosis was made for the pulmonary allograft and the middle of the vascular grafts. Finally, the RVOT was closed with a PTFE patch.

Histopathological examination of the resected specimen revealed an undifferentiated pleomorphic sarcoma (*Figure 2C*) with surgical margins free of disease. On immunohistochemical staining, the neoplastic cells were positive for alpha-smooth muscle actin, vimentin, and CD 68, partially positive for desmin and murine double minute 2, and negative for myoglobin and myogenin. The patient was discharged on Day 7 post-operatively in a favourable condition. He refused to receive adjunctive chemotherapy, radiotherapy, or targeted therapy due to the associated risks and tentative efficacy. He remains otherwise healthy with no symptoms. A follow-up CT 5 months post-operatively indicated no recurrence or metastasis (*Figure 2D*).

Discussion

Pulmonary artery sarcoma is believed to originate from the pluripotential mesenchymal cells of the intima of the pulmonary artery trunk or the bulbous cordi.[5] While surgery remains the first line of treatment for PAS, pneumonectomy, PEA, and graft reconstruction of the pulmonary artery constitute the mainstream choices. Krüger *et al.*[6] reported a 58% incidence of pulmonary metastases in patients with PAS and described pneumonectomy as the most reasonable oncological concept. On the other hand, scholars from the University of California, San Diego,[7] stated that PAS was always bilateral if the patient presented with pulmonary hypertension, and even in a unilateral case, bilateral seeding had probably occurred by the time the patient presented. Therefore, they insisted on PEA as a palliative surgery for extending the lifespan and did not recommend pneumonectomy for this disease.

In our centre, our earlier practice was to perform PEA instead of pneumonectomy for PAS, since most patients had evidence of bilateral disease, with half of them having coexisting pulmonary hypertension at baseline.[1] As tumours grow, they are usually limited to pulmonary artery intima, rather than invading through the artery into the surrounding organs. When an initial or recurrent tumour involves only the RVOT and the main pulmonary artery trunk, it is technically possible to achieve complete surgical resection and reconstruction.[3,4]

Some reports have demonstrated that patients with PAS, who receive post-operative adjuvant therapy, have significantly improved survival rates.[2,8] There is great variability in the chemotherapy regimens prescribed to patients with PAS, of which the ifosfamide and epirubicin regimen is the most typical combination.[1] The recent success of immune checkpoint inhibitors that target either programmed death-1 or programmed death-ligand 1 in cancer treatment suggests that immunotherapy may play an important role in sarcoma therapy. A recent case report described promising treatments for metastatic pulmonary angiosarcoma with pembrolizumab.[9] However, further research on immunotherapy should be undertaken to investigate its efficacy and safety in this disease.

In the present case, the tumour mass involved the RVOT, the pulmonary trunk extending into bifurcation, and the right pulmonary artery. Nevertheless, it was safely resected and reconstructed by using a homologous pulmonary valve and PTFE vascular grafts, combined with right PEA for potential seeding metastasis. Complete resection might improve long-term survival for this patient.

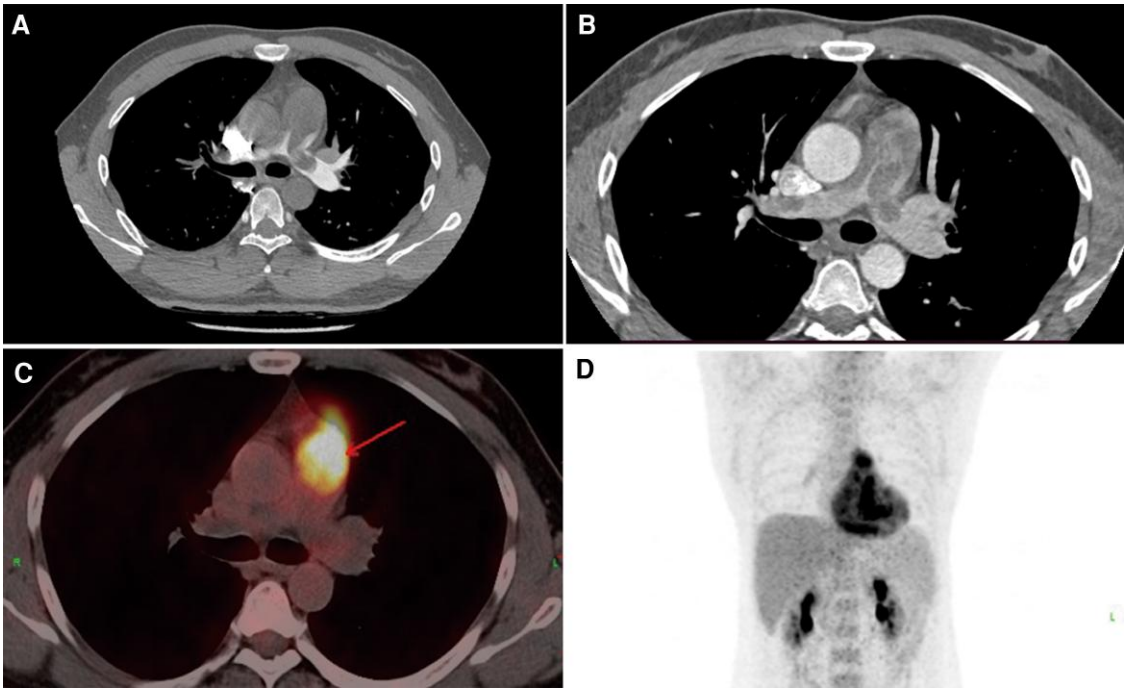


Figure 1 Computed tomography shows a mass in the right ventricle outflow tract, pulmonary trunk extending into bifurcation, and right pulmonary arteries (*A* and *B*); positron emission tomographic computed tomography reveals increased fluorodeoxyglucose uptake ($SUV_{max} = 15.6$) in the right ventricle outflow tract and pulmonary artery trunk (*C* and *D*).

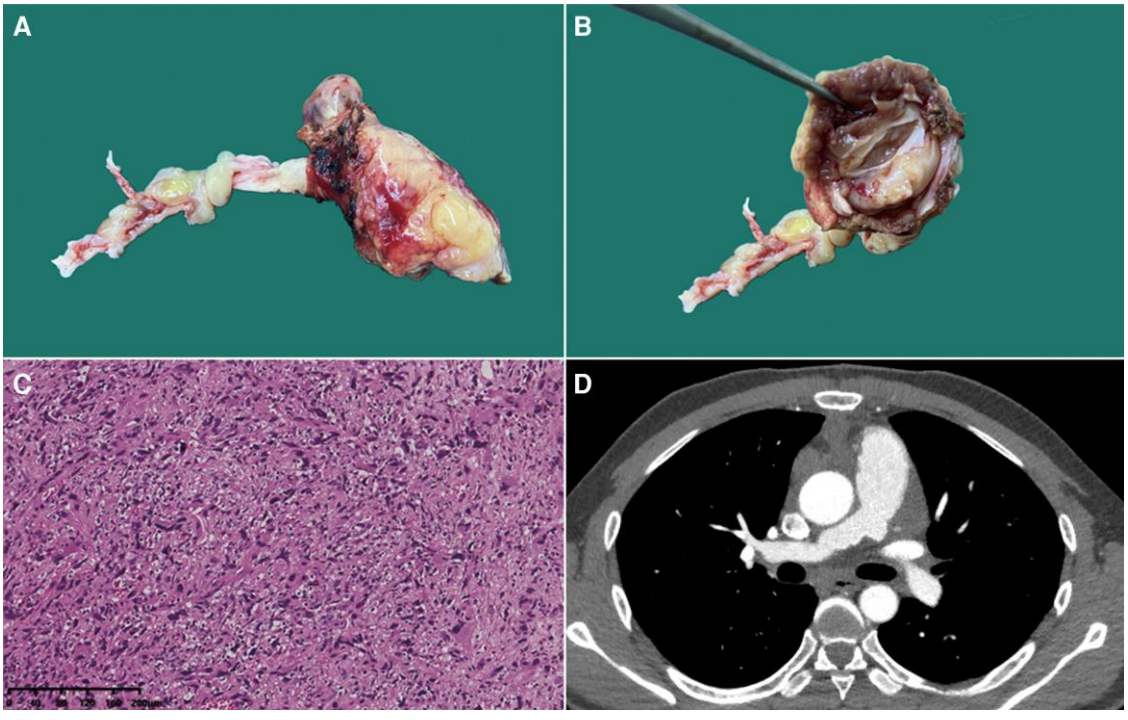
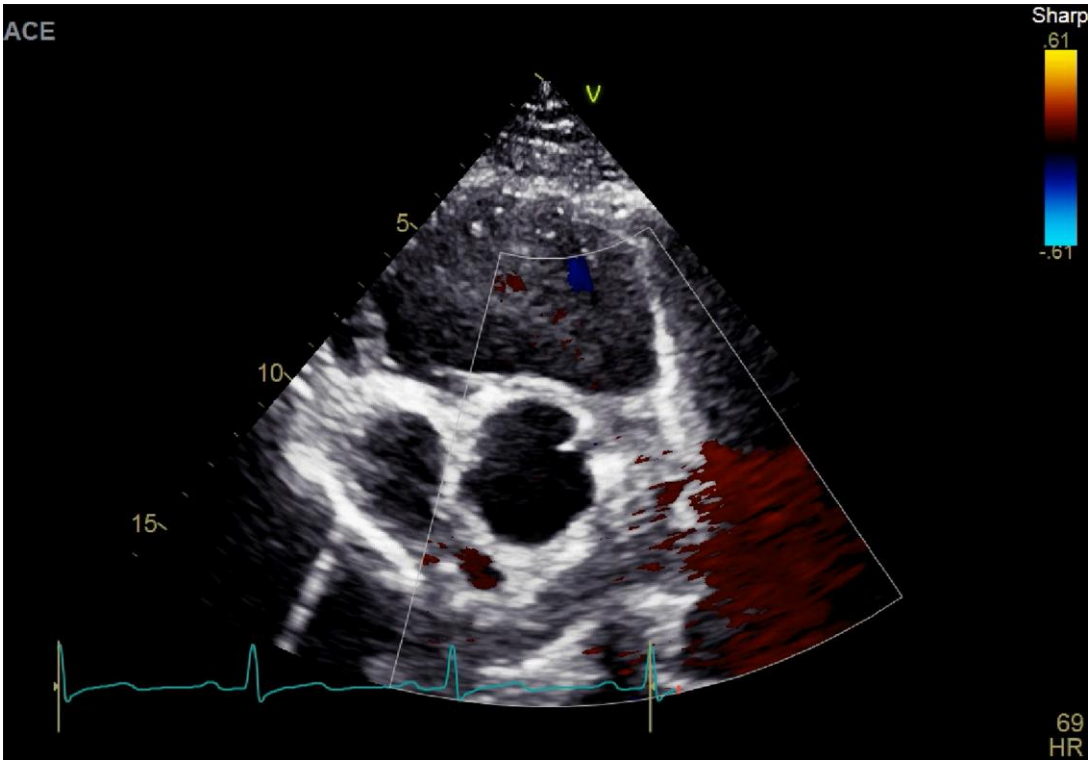


Figure 2 The surgical specimens (*A* and *B*) of a patient with pulmonary artery sarcoma. (*C*) Haematoxylin and eosin staining of the tumour demonstrates undifferentiated spindle cells. (*D*) The follow-up computed tomography scan 5 months after surgery indicates no recurrence or metastasis.



Video 1 Transthoracic echocardiography section for the right ventricle outflow tract of a patient with pulmonary artery sarcoma.

Conclusion

Creative surgical strategies are required to achieve complete resection in PAS. Radical resection with reconstruction using pulmonary valve allograft and PTFE vascular grafts is technically feasible and safe, while PEA eliminates the chances of potential intimal implantation metastasis.

Lead author biography

Dr Sheng Liu graduated from Peking Union Medical College, Beijing, China. He is the director of the Cardiac Surgery Department and leader of the Cardiac Transplantation Center in FuWai Hospital. Dr Liu is an expert in the surgical treatment of end-stage heart disease, chronic thrombo-embolic pulmonary hypertension, and coronary heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report, including image(s) and associated text, has been obtained from the patient in line with COPE guidance.

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References

1. Song W, Deng L, Zhu J, Zheng S, Wang H, Song Y, Liu S. Surgical treatment of pulmonary artery sarcoma: a report of 17 cases. *Pulm Circ* 2021;**11**:2045894020986394.
2. Bandyopadhyay D, Panchabhai TS, Bajaj NS, Patil PD, Bunte MC. Primary pulmonary artery sarcoma: a close associate of pulmonary embolism—20-year observational analysis. *J Thorac Dis* 2016;**8**:2592–2601.
3. Chan EY, Ravi V, Ali A, Nguyen DT, Graviss EA, MacGillivray TE, Reardon MJ. Surgical management of primary pulmonary artery sarcoma. *Semin Thorac Cardiovasc Surg* 2021:S1043-0679(21)00472-X.
4. Kim HK, Choi YS, Kim K, Shim YM, Sung K, Lee YT, Park PW, Kim J. Surgical treatment for pulmonary artery sarcoma. *Eur J Cardiothorac Surg* 2008;**33**:712–716.
5. Anderson MB, Kriett JM, Kapelanski DP, Tarazi R, Jamieson SW. Primary pulmonary artery sarcoma: a report of six cases. *Ann Thorac Surg* 1995;**59**:1487–1490.
6. Krüger I, Borowski A, Horst M, de Vivie ER, Theissen P, Gross-Fengels W. Symptoms, diagnosis, and therapy of primary sarcomas of the pulmonary artery. *Thorac Cardiovasc Surg* 1990;**38**:91–95.
7. Jamieson SW. Pulmonary artery sarcoma. *Eur J Cardiothorac Surg* 2013;**43**:793–794.
8. Yin K, Zhang Z, Luo R, Ji Y, Zheng D, Lin Y, Wang C. Clinical features and surgical outcomes of pulmonary artery sarcoma. *J Thorac Cardiovasc Surg* 2018;**155**:1109–1115.e1.
9. Nguyen QT, Pham AT, Nguyen TT, Nguyen TTT, Van Le K. Role of immunotherapy in pulmonary angiosarcoma: a case report. *Case Rep Oncol* 2021;**14**:797–801.