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Case Report

Palpitations Revealing a Primary Pulmonary Artery Sarcoma

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ABSTRACT

Primary pulmonary artery sarcoma is an exceedingly rare and aggressive malignancy that carries poor prognosis. Clinical manifestations are nonspecific and include chest pain, dyspnea, syncope, palpitations, and asthenia, among others. Delay to diagnosis is common and compromises the prognosis. Here, we report an interesting case of primary pulmonary artery sarcoma presenting with frequent monomorphic premature ventricular contractions arising from the right/left ventricle outflow tract. Cardiac imaging is key in the evaluation of patients with frequent premature ventricular contractions to rule out rare pathologies such as tumour compression.

RÉSUMÉ

Le sarcome primaire de l'artère pulmonaire est une tumeur maligne extrêmement rare et agressive de mauvais pronostic. Les manifestations cliniques sont non spécifiques et peuvent inclure de la douleur thoracique, de la dyspnée, des syncopes, des palpitations et de l'asthénie. Le retard diagnostic est fréquent et compromet le pronostic. Nous rapportons ici un cas intéressant de sarcome primaire de l'artère pulmonaire pour lequel le patient présentait des extrasystoles ventriculaires prématurées monomorphes qui provenaient des chambres de chasse des ventricules gauche et droit. L'imagerie est essentielle à l'évaluation des patients présentant de fréquentes extrasystoles ventriculaires afin d'écarter des pathologies rares comme la compression tumorale d'une chambre cardiaque.

A 60-year-old man presented to the emergency room with palpitations of a few months' duration. Physical examination revealed a 2/6 systolic ejection murmur at the left upper sternal border, louder with inspiration. The 12-lead electrocardiogram (ECG) showed ventricular bigeminy. These premature ventricular contractions (PVCs) had a positive morphology in the inferior leads and a positive concordance in the precordial leads. Thus, an origin from the posterior site of the right-left ventricle outflow tract was suspected (Fig. 1A). A 24-hour Holter monitor, recently ordered by the patient's family physician for palpitations, revealed 15% burden of monomorphic PVCs.

The initial differential diagnosis of PVCs arising from the outflow tract should include acute pulmonary embolism (PE), chronic thromboembolic pulmonary hypertension, congenital pulmonary stenosis, and cardiomyopathy with outflow tract arrhythmias. Other conditions that deserve consideration include obstructive pulmonary hypertension caused by a primary tumour into the pulmonary artery (PA), a cardiac lymphoma, or metastases obstructing the right ventricular outflow tract.[1,2]

A thoracic contrast-enhanced computed tomography (CT) scan was first ordered to exclude a PE. It showed a polylobulated 3.9 × 6.2-cm mass invading the pulmonary trunk. The morphology was described as atypical for a thrombus; the diagnosis of a pulmonary artery sarcoma (PAS) was considered (Fig. 1C-E). Echocardiography confirmed a large mass in the PA, causing supravalvular stenosis with a maximal gradient of 46 mm Hg (Fig. 1B). Cardiac magnetic resonance imaging (MRI) suggested that the mass was invading the anterior wall of the PA, concordant with the hypothesis of a sarcoma (Fig. 2A). The right ventricle was mildly dilated (volume 129 mL/m^2), with a systolic function at the inferior limit of normal. Moreover, there was a thrombotic component to the mass. Finally, a positron emission tomography (PET) with fluorodeoxyglucose (FDG) scan showed significant hypermetabolism, compatible with a neoplastic process (Fig. 2B). There was no evidence of metastasis.

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Novel Teaching Points

- Pulmonary artery sarcoma can present with frequent premature ventricular contractions, particularly when they are arising from the right ventricle.
- Imaging, such as transthoracic echocardiography, should be promptly ordered to exclude rare causes of frequent ventricular extrasystoles such as tumour compression.
- Multimodality imaging should be performed when the initial workup is inconclusive or yields atypical results with a broad differential diagnosis.
- Multidisciplinary discussions are important in the diagnosis and management of primary pulmonary artery sarcoma.

The patient was prescribed low-molecular-weight heparin and bisoprolol, and he went to surgery a few days later. The origin of the mass was located in the anterolateral wall of the PA, invading the pulmonary valve (Fig. 2C). The frozen biopsy confirmed the diagnosis of a sarcoma (Fig. 2D). The mass was removed, and the pulmonary valve and trunk were replaced with a bioprosthesis.

The patient was seen at the oncology follow-up clinic 1 month later. The final biopsy analysis revealed an undifferentiated spindle cell sarcoma with negative margins. A chest CT scan showed no evidence of tumour, and no additional treatment was proposed. The patient remained arrhythmia free.

Discussion

Approximately 20% of cardiac masses are malignant, including sarcoma, pericardial mesothelioma, primary lymphoma, and—most commonly—metastases (from either lung, breast, or hematologic cancer).[2] Cardiac sarcomas are the most common malignant primary cardiac tumours, arising from any chamber, but mostly from the right atrium. PAS is very rare, with approximately 400 cases reported since first described in 1923. The average age at diagnosis is 50 years.[3] The most common presenting symptom is shortness of breath. Other symptoms include chest pain, cough, hemoptysis, dizziness, and other systemic symptoms. Palpitations are reported in 22.2% of cases, mostly as nonspecific tachycardia.[4] Physical examination may show signs of right-ventricular failure.[2] Because of the nonspecificity of symptoms, a misdiagnosis of PE is common. Bandyopadhyay et al. reported that 47% of patients with PAS were initially diagnosed with PE.[5] The delay from symptoms to the diagnosis ranges from 1.7 to 18.2 months.[4]

No imaging findings are considered pathognomonic. Multislice gated CT is commonly the initial imaging study and facilitates the distinction from PE. PAS involves the PA trunk and its main branches, is heterogeneous, and exhibit a globular appearance with possible extravascular tumour extension and delayed enhancement.[5] The tumour periphery is usually hypervascular, with a necrotic centre. Moreover, PAS projects in the direction of blood flow, whereas PE will

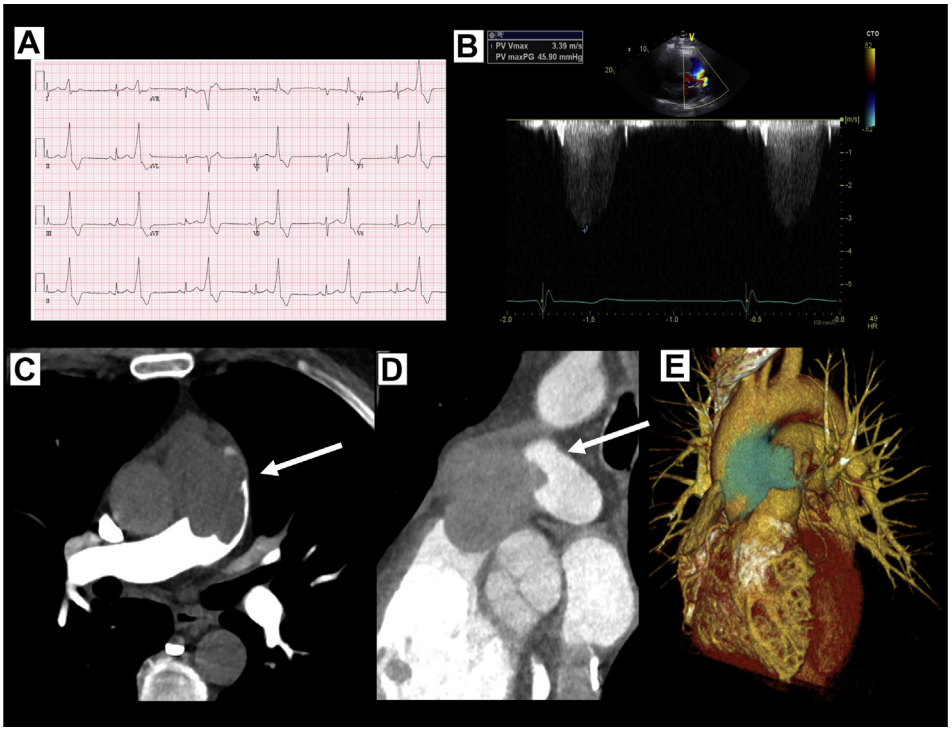

Figure 1. (**A**) A 12-lead electrocardiogram on admission demonstrating ventricular bigeminism. (**B**) Pressure gradient via the pulmonary valve, measured by continuous Doppler echocardiography. (**C**) Axial and (**D**) sagittal contrast-enhanced chest computed tomography (CT) images shows intraluminal filling defect in the main pulmonary artery (**white arrows**). (**E**) 3D reconstruction CT image shows the location of the mass (**blue**) between the aorta and the main pulmonary artery.

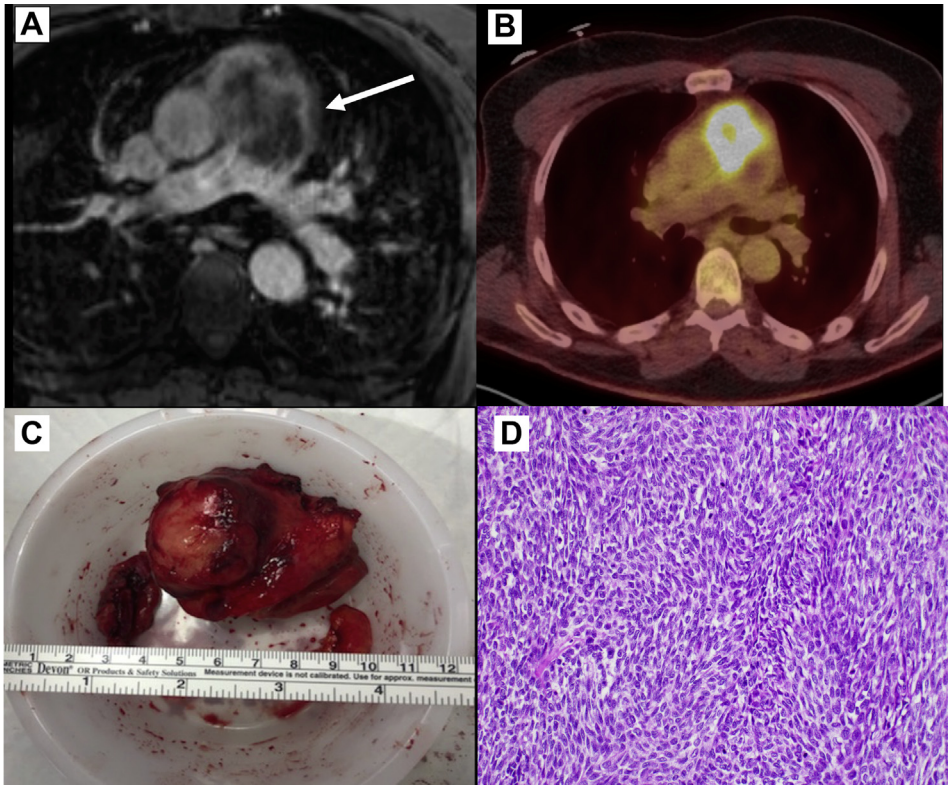

Figure 2. (**A**) On contrast-enhanced magnetic resonance imaging, the lesion (**white arrow**) shows intense heterogeneous enhancement. (**B**) The axial positron-emission tomography-computed tomography fused image shows increased fluorodeoxyglucose uptake of the main pulmonary artery mass. (**C**) Macroscopic photo of the tumour. (**D**) Surgical biopsy of the cardiac mass (200× magnification). The tumour is composed of highly cellular fascicles of spindle cells with prominent mitotic activity.

appear as a cup-like structure against blood flow.[3] MRI can detect soft tissue components of the tumour within the lumen of the PA and its extension. Functional assessment can provide information such as flow and gradients across the PA.[6] Gadolinium-enhanced MRI shows heterogeneous enhancement for PAS.[5] The FDG-PET scan is another valuable tool to differentiate PAS from PE on the basis of differences in standardized uptake value levels and is used for staging purposes.[7] Echocardiography provides information about the degree of PA obstruction, pulmonary valve function, RV dilation, and function.[3]

Without treatment, the survival of patients with PAS has been estimated to be as short as 6 weeks.[5] Surgery is the mainstay of therapy and includes palliative PA stenting, pulmonary endarterectomy, pneumonectomy, debulking, and wide surgical excision. An aggressive approach—including resection of the main PA and its branches, followed by graft reconstruction—is suggested.[6] The 5-year survival for patients undergoing a curative resection attempt was reported as 49.2%, compared with 0% for those undergoing incomplete resection.[8]

Neoadjuvant chemotherapy is proposed to allow tumour shrinkage and enhance resectability. If chosen, the initial use of adryamicin combined with ifosfamide, gemcitabine plus taxane, or dacarbazine regimens has been reported.[8] Postoperative chemotherapy and/or radiotherapy may improve treatment efficacy, but there is no clear consensus nor convincing data about this strategy.[7]

Conclusions

PAS is a rare disease that presents with nonspecific symptoms, including isolated palpitations. Misdiagnosis is common and can compromise the prognosis. To our knowledge, this is the first case presenting with frequent ectopic ventricular beats secondary to mass compression. Physicians have to be aware of this entity and consider it in their differential diagnosis to ensure proper management.

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References

1. Mussot S, Ghigna MR, Mercier O, et al. Retrospective institutional study of 31 patients treated for pulmonary artery sarcoma. Eur J Cardiothorac Surg 2013;43:787-93.

2. Gupta R, Meghrajani V, Desai R, Gupta N. Primary malignant cardiac tumors: a rare disease with an adventurous journey. J Am Heart Assoc 2020;9:e016032.

3. Xu R, Zhao Y, Xu X, et al. Pulmonary intimal sarcoma involving the pulmonary valve and right ventricular outflow tract: a case report and literature review. Medicine (Baltimore) 2020;99:e18813.

4. Pu X, Song M, Huang X, et al. Clinical and radiological features of pulmonary artery sarcoma: a report of nine cases. Clin Respir J 2018;12:1820-9.

5. 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;8:2592-601.

6. Wyler von Ballmoos MC, Chan EY, Reardon MJ. Imaging and surgical treatment of primary pulmonary artery sarcoma. Int J Cardiovasc Imaging 2019;35:1429-33.

7. Yin K, Zhang Z, Luo R, et al. Clinical features and surgical outcomes of pulmonary artery sarcoma. J Thorac Cardiovasc Surg 2018;155:1109-1115.e1101.

8. Blackmon SH, Rice DC, Correa AM, et al. Management of primary pulmonary artery sarcomas. Ann Thorac Surg 2009;87:977-84.