Received: 18 September 2023 | Revised: 25 November 2023 | Accepted: 14 December 2023

DOI: 10.1002/ccr3.8401

CASE REPORT

Clinical Case Reports Open Access WILEY

Bioprosthetic pulmonary valve dysfunction in a primary cardiac sarcoma survivor: Clinical considerations and treatment options

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Key Clinical Message

The case highlights the good survival after radical surgery and chemotherapy of a cardiac sarcoma, and the need for close follow-up due to possible early postsurgical complications.

KEYWORDS

cardiac sarcoma, prosthetic valve dysfunction, valve-in-valve

1 | INTRODUCTION

Primary sarcomas are a rare and aggressive group of cardiac malignancies. Despite the advances in medical therapy and surgical techniques, the prognosis for patients with cardiac sarcomas remains poor, with limited treatment options and low survival rates.[1] We describe the good postsurgical survival of a patient affected by cardiac sarcoma and the early prosthetic valve dysfunction successfully treated with a transcatheter valve-in-valve procedure.

2 | CASE PRESENTATION

We have already described the case of a 69-year-old woman with a diagnosis of right ventricular outflow tract (RVOT) and pulmonary artery pleomorphic sarcoma, treated with radical excision, reconstruction of the pulmonary artery, and bioprosthetic pulmonary valve replacement.[2] After surgery, the patient started adjuvant chemotherapy with epidoxorubicin and had a close oncological and cardiological follow-up.

After 2 years, the patient was alive and did not show signs of recurrence. However, she presented at the emergency department because of signs and symptoms of heart failure (HF): she reported shortness of breath and weight gain in the last weeks, and was found with peripheral edema, increased CVP and mild reduction of vesicular murmur at the basis. Laboratory tests showed an increased propeptide of Brain Natriuretic Peptide (NT-proBNP 1280 pg/mL; normal values: <300 pg/mL). Collaterally, elevated serum parathyroid hormone

was found (85 pg/mL; normal values: 11–73 pg/mL), screened after incidental finding of hypercalcemia at arterial blood gas analysis. A transthoracic echocardiography was performed showing right ventricular (RV) dilatation (RVOT basal diameter 38 mm) and dysfunction, with both stenosis and severe regurgitation of the pulmonary bioprosthesis (Figure 1A): PHT 75 msec; Vmax 2,5 m/sec. A transesophageal echocardiography confirmed the signs of RV overload and prosthetic valve dysfunction (Figure 1B,C). To better evaluate the anatomy of the RVOT and the pulmonary artery, the patient underwent cardiac computed tomography, showing thickened prosthetic cusps and confirming the absence of oncologic disease recurrence. Moreover, a positron emission tomography/computed tomography (PET/CT) scan with 18F-choline was performed, revealing a hypercaptation in the inferior right parathyroid gland.

To address the pulmonary valve dysfunction, after the heart team discussion, the patient underwent transcatheter valve-in-valve implantation.

The procedure was performed under general anesthesia. Aortography and angiography of the left coronary circulation were performed to assess its relation to the pulmonary artery trunk (Figure 2A), considering that CA compression following TPV implantation is significantly associated with the presence of abnormal CA anatomy, so preimplantation coronary angiography is an important step to evaluate for the presence of CA compression during TPV implantation. Anterograde catheterization of the pulmonary bioprosthesis confirmed severe prosthetic regurgitation. Subsequently, an extra-stiff guide was placed in the left pulmonary artery (Figure 2B), and then a Sapien 3 23 mm balloon expandable bioprosthesis was implanted with good procedural result (Figure 2C,D). The patient was already on an antiplatelet regimen with 100 mg of acetylsalicylic acid, which was continued.

Following the transcatheter procedure, the patient experienced a significant improvement in symptoms and cardiac function, and was stable after 4-months. Postoperative echocardiography showed normalization of RV dimensions and function and normal prosthetic valve function. Subsequently, the patient was referred to an endocrinologist to manage the primary hyperparathyroidism.

3 | DISCUSSION

Primary cardiac malignancies are rare and associated with a poor prognosis due to their aggressive nature and limited treatment options.[3] Radical surgical resection remains the main therapeutic approach, although achieving a complete surgical excision can be challenging.

In our case, the patient was affected by a pleomorphic sarcoma. This aggressive tumor is associated with an extremely poor prognosis due to its rapid progression.[1] Our patient was alive after 2 years following the initial diagnosis, surgical treatment, and adjuvant chemotherapy. However, she developed an early degeneration of the bioprosthetic pulmonary valve.

Bioprosthetic valves are commonly used in cardiac surgery due to their reduced risk of thrombosis and avoidance of long-term anticoagulation therapy. However, they are known to have a limited lifespan, especially in younger patients.[4]

In our patient, the early bioprosthetic dysfunction introduced new challenges. In fact, medical therapy alone was not sufficient to control HF symptoms and redo-surgery was considered at extremely high risk. The subsequent transcatheter valve-in-valve implantation proved to be feasible, safe and effective, leading to a significant clinical improvement and normalization in RV function and dimensions.

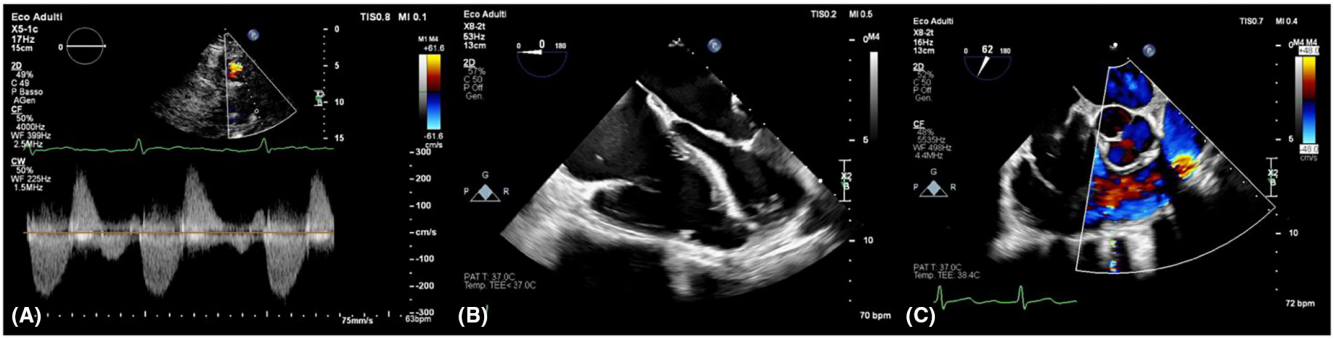

FIGURE 1 (A) Transthoracic echocardiography, continuous wave Doppler of the pulmonary bioprosthesis, showing an increased transprosthetic gradient and proto-diastolic retrograde flow with markedly decreased pressure half-time, consistent with prosthetic stenosis and severe regurgitation. (B) Transesophageal echocardiography, mid-esophageal four-chamber view, showing enlarged right ventricle with signs of volume overload. (C) Transesophageal echocardiography, mid-esophageal short axis view with color-Doppler study, showing severe intraprosthetic regurgitation.

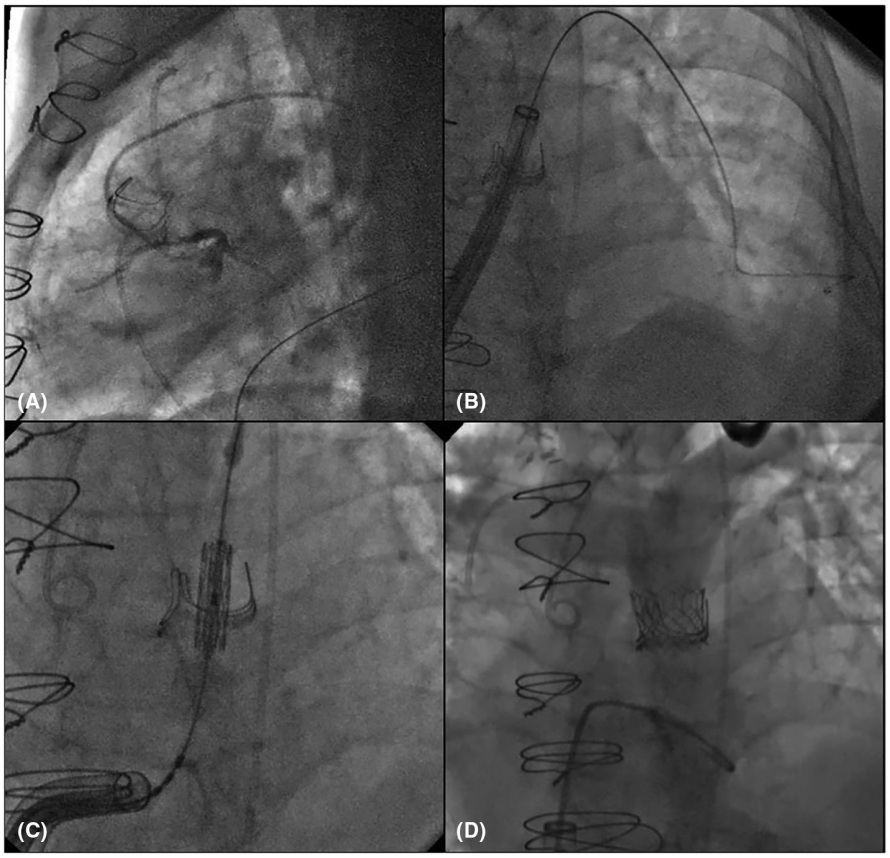

FIGURE 2 (A) Coronary angiography showing the course of left main coronary artery. (B) Placement of an extra-stiff guide in the left pulmonary artery and the 24F introducer over the pulmonary bioprosthesis. (C) Placement of the Sapien 3 23 mm bioprosthesis over the previous pulmonary bioprosthesis. (D) Implantatation of Sapien 3 23 mm bioprosthesis with good result and without any residual regurgitation.

The case highlights the good survival after radical surgery and chemotherapy of a cardiac sarcoma, and the need for close follow-up due to possible early postsurgical complications, particularly in patients receiving cardiotoxic chemotherapy.

The correlation between anthracycline-based chemotherapy agents, including epidoxorubicin, and native valve degeneration has been previously reported.[5] The mechanisms underlying anthracycline-induced valve dysfunction are not fully understood. However, it is thought that the direct toxicity of anthracyclines to valvular tissue, oxidative stress, and inflammation contribute to valve degeneration and fibrosis over time.[6] In our case, the patient's exposure to epidoxorubicin as part of adjuvant chemotherapy raises the possibility of anthracycline-induced bioprosthetic pulmonary valve degeneration, even if to our best knowledge no evidence of such effect is currently reported in literature.

The potential association between primary hyperparathyroidism and the observed bioprosthetic valve degeneration is another interesting point. Hyperparathyroidism can lead to elevated serum calcium levels and disturbances in mineral metabolism. Prolonged hypercalcemia and increased parathyroid hormone levels can promote calcification and mineralization of tissues, including bioprosthetic valves. These processes may contribute to accelerated degeneration over time. While the exact mechanisms linking hyperparathyroidism and bioprosthesis degeneration require further investigation, several studies have explored the association between hyperparathyroidism and valvular calcification.[7]

Percutaneous valve interventions have emerged as an alternative and effective treatment option for high-surgical-risk patients. Several studies showed the safety and efficacy of valve-in-valve techniques, leading to improved hemodynamics and enhanced clinical outcomes in patients undergoing transcatheter pulmonary valve-in-valve replacement.[8]

4 | CONCLUSIONS

The management of patients with primary cardiac tumors remains challenging. Improved patient survival leads to new clinical scenarios, where patients may experience complications unrelated to the oncological history. In this context, multidisciplinary care proved to be fundamental for the effective treatment of a complex cardiac condition.

AUTHOR CONTRIBUTIONS

Davide Maione: Project administration; writing – original draft; writing – review and editing. **Antonio De Luca:** Conceptualization; data curation; supervision; writing – review and editing. **Andrea Pezzato:** Resources. **Giancarlo Vitrella:** Resources. **Andrea Perkan:**

Resources; supervision. **Elisabetta Rauber:** Resources. **Gianfranco Butera:** Supervision; writing – review and editing. **Gianfranco Sinagra:** Supervision; validation; writing – review and editing.

FUNDING INFORMATION

The present submission was not supported by external fundings.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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How to cite this article: Maione D, De Luca A, Pezzato A, et al. Bioprosthetic pulmonary valve dysfunction in a primary cardiac sarcoma survivor: Clinical considerations and treatment options. *Clin Case Rep*. 2024;12:e8401. doi:10.1002/ccr3.8401