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Rare giant RVOT aneurysm, encasing and compressing the ascending aorta, superior vena cava, and pulmonary trunk along with right ventricle: Surgical excision with RVOT reconstruction and outcome – A case report

***Corresponding Author:** Rakesh Kumar Verma*

Email: rakeshcvts11@gmail.com

Abstract

Introduction: Right Ventricular Outflow Tract (RVOT) aneurysm is an uncommon entity, most frequently described as a late complication following repair of congenital heart disease, particularly Tetralogy of Fallot. Pulmonary Stenosis (PS) increases the hemodynamic burden on the Right Ventricle (RV), exacerbating RVOT pathology.

Case presentation: We report a rare case of a 28-year-old male presented with a three-year history of chest heaviness, palpitations, and progressively worsening orthopnea. The patient presented with a giant RVOT aneurysm that encased and compressed the ascending aorta, superior vena cava, pulmonary trunk, and right ventricle. Surgical management required meticulous dissection to separate the aneurysm from adjacent mediastinal structures, followed by complete excision, transannular patch augmentation of the RVOT and main pulmonary artery, and pulmonary valve replacement.

Conclusion: RVOT aneurysm associated with pulmonary stenosis represents a complex pathology that mandates definitive surgical intervention for excellent anatomical restoration and functional outcomes.

Rakesh Kumar Verma*; Kumar Saurabh Gaur; Prabhat Biswas; Lakshman Balasubramanian; Parinita Chelleng

LPS Institute of Cardiology & Cardiac Surgery in Kanpur, LPS Institute of Cardiology, India.

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Keywords: Pulmonary artery aneurysm; RVOT (Right Ventricular Outflow Tract) aneurysm; Pulmonary valve stenosis; Pulmonary valve replacement; Axillo-bicaval cardiopulmonary bypass.

Introduction

The preoperative challenge in giant RVOT aneurysms lies in their mass effect on adjacent mediastinal structures. Progressive expansion often results in severe extrinsic compression of the Main Pulmonary Artery (MPA) and distortion of the pulmonary valve apparatus [1]. This will exaggerate problems when there is prior disease in the pulmonary valve, the main pulmonary artery, the left pulmonary artery, and the right pulmonary artery. This creates a complex hemodynamic condition in which the Right Ventricle (RV) must overcome both intrinsic valvular resistance and extrinsic arterial compression, ultimately leading to RV failure.

Given these anatomical constraints, preoperative planning is critical. Traditional surgical approaches may be complicated by the aneurysm's proximity to the sternum and major vessels, necessitating alternative strategies for Cardiopulmonary Bypass (CPB) initiation. This report details the preoperative evaluation and strategic planning for a patient with a giant RVOT aneurysm, in which elective axillo-bicaval cannulation and pericardial patch augmentation were identified as the primary maneuvers to ensure safe surgical entry and restore pulmonary outflow tract patency.

Case report

A 28-year-old male presented with a three-year history of chest heaviness, palpitations, and progressively worsening orthopnea. He had no history of prior cardiac surgery or known congenital heart disease.

On examination, a palpable thrill was noted in the pulmonary area, along with a right parasternal heave. A systolic murmur of variable intensity was best heard at the left upper sternal border.

Transthoracic echocardiography revealed a large aneurysmal dilatation (110×80 mm) arising from the base of the right ventricle and abutting the superior vena cava, main pulmonary artery, and RV free wall. Severe pulmonary valvular stenosis was present, with a peak gradient of 80 mmHg and a peak velocity of 4.5 m/s. Severe pulmonary regurgitation and mild tricuspid regurgitation were also observed. Transesophageal Echocardiography (TEE) confirmed similar findings in the mid-esophageal aortic long-axis view (Figure 1). Cardiac Computed Tomography (CT) with pulmonary angiography, performed using a 128-slice scanner, identified a large outpouching aneurysm measuring 119×88 mm originating from the right Ventricular Outflow Tract (RVOT) proximal to the pulmonary valve, through a narrow neck measuring 18 mm (Figure 2). The aneurysm encased and compressed the ascending aorta, superior vena cava, pulmonary trunk, and body of the RV (Figure 3). Laboratory investigations, including complete blood count, metabolic panel, and coagulation profile, were within normal limits. Electrocardiography demonstrated a heart rate of 150 bpm, right axis deviation, right ventricular hypertrophy, and ST-segment depression in leads V1 and V3.

Surgical technique

Establishing Cardiopulmonary Bypass (CPB)

Due to the complexity and size of the aneurysm, we used a specific cannulation strategy

- **Axillo–bicaval cannulation:** Instead of the standard aortic cannulation, the Axillary Artery cannulation was used for arterial inflow (16-F cannula), while the Superior Vena Cava (SVC) (22-F cannula) and Inferior Vena Cava (IVC) (28-F cannula) handled venous return for CPB, along with a snare in the SVC and IVC.
- **Myocardial protection:** The patient was cooled to 32°C. After cross-clamping the aorta, blood cardioplegia was delivered. A vent was placed in the Right Superior Pulmonary Vein to prevent the heart from overfilling (distension).

Aneurysm excision and RVOT reconstruction

The surgeon's primary goal was to remove the aneurysm while restoring the compressed vessel anatomy, as the origin of the ostium of the aneurysm is subannular and is below the pulmonary valve, anterior to it.

- **Dissection:** Careful retrograde and antegrade dissection of the right pulmonary artery, left pulmonary artery, and main pulmonary artery was carried out to delineate the anatomy of the pulmonary trunk, and it was separated from the aneurysm
- **PA augmentation:** A long longitudinal incision was made over the aneurysm, which was found to originate from the RVOT proximal to the pulmonary valve. The main pulmonary artery was noted to be severely compressed by the aneurysm with a slit-like opening. The whole aneurysmal sac was fully excised along with drainage of frank blood, which was its content. The pulmonary artery was calibrated with Hegars dilator and augmented using a transannular pericardial patch, which extended up to MPA, RVOT, and pulmonary annulus, and it was sutured with continuous 5-0 Prolene (Figure 4).
- **Valve replacement:** Because the pulmonary valve was "tethered, stenotic, and irreparable," a 23A TTK Chitra tilting-disc valve was implanted using pledgeted sutures in the neo pulmonary annulus.

Weaning and postoperative results

Once the repair was complete, the "restart" process began:

- **De-airing:** Crucial steps were taken to ensure no air remained in the heart chambers (using the pulmonary vein vent and CPAP) before removing the aortic cross-clamp.
- **Inotropic support:** The patient was weaned off the bypass machine with the help of low-dose inotropic support (**Dobutamine, Adrenaline, and Milrinone**).
- **Closure:** Protamine was given to reverse the Heparin, hemostasis was confirmed, and the chest was closed.

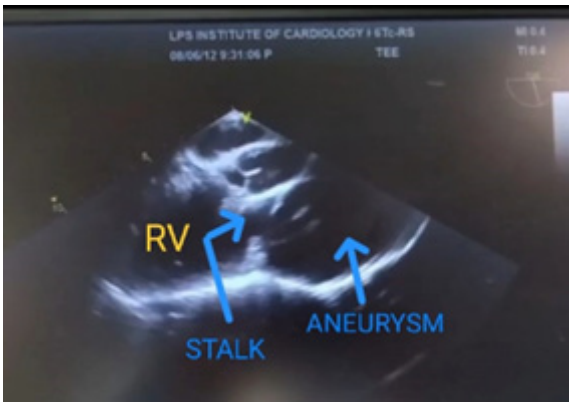

Figure 1: TEE shows mid esophageal aortic long axis view showing large RV aneurysm with stalk of 1.8 cm.

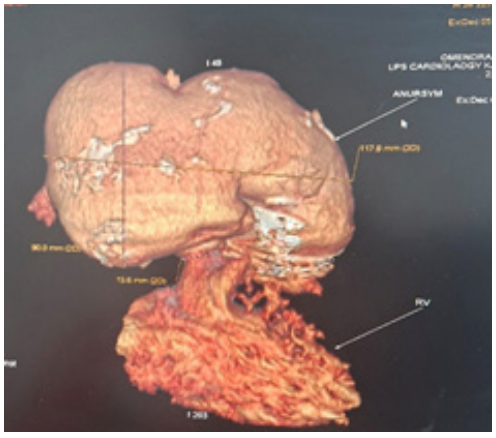

Figure 2: 3D image showing large outpouching through a narrow neck from RV measuring 119×88 mm.

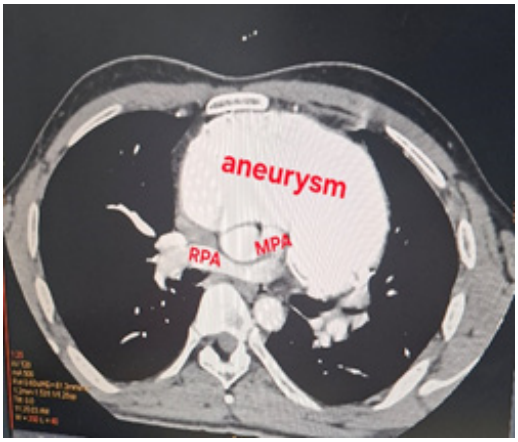

Figure 3: Cardiac CT showing giant RVOT aneurysm encasing of ascending aorta, SVC, pulmonary trunk and most of RV.

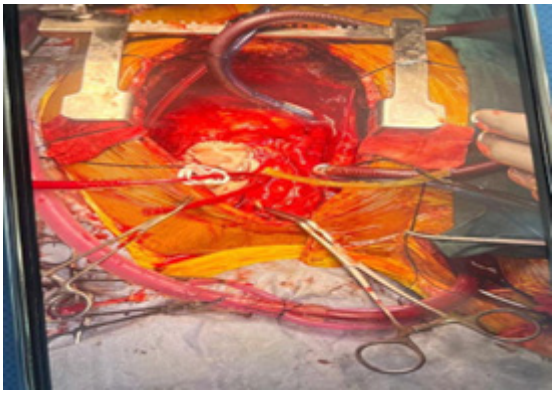

Figure 4: MPA was augmented with pericardial patch, which was fixed with 5-0 prolene.

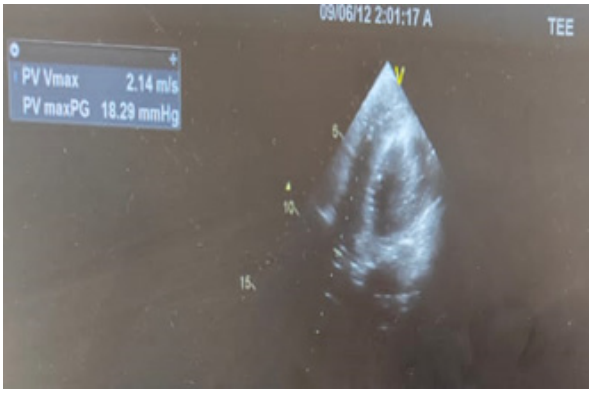

Figure 5: Postoperative TEE demonstrated a reduction in pulmonary valve gradient to 18mmHg

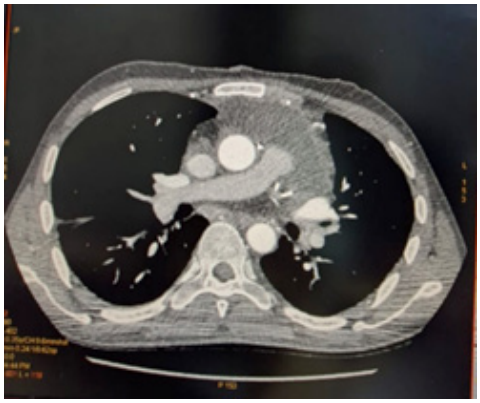

Figure 6: Postoperative CT Pulmonary angiography shows complete excision of aneurysm.

Key outcomes

Postoperative TEE demonstrated a reduction in pulmonary valve gradient to 18 mmHg (Figure 5) with normal RV and LV function, with an ejection fraction of 58%. The postoperative course was uneventful, and the patient was extubated within 12 hours. The patient was discharged after 7 days. Post-operative CT Pulmonary angiography shows complete excision of aneurysm (Figure 6). Histopathology was not sent in this case.

Discussion

Idiopathic RVOT dilatation is rare. RVOT dilatation/aneurysm can develop due to complications of surgical procedures like ventriculotomy for the repair of TOF and creation of an aorta to main pulmonary artery shunt, but an idiopathic etiology has been rarely reported [3]. Our patient never had cardiac surgery and was found to have RVOT dilatation on imaging for dyspnea workup. The Framingham Heart Study established the mean MPA diameter as 2.51+0.28 cm; sex-specific reference values were 2.9 cm in men and 2.7 cm in women [4]. 2D echo/TEE measured MPA diameter to be 1 cm, which is less than 2.9 cm. CT Pulmonary Angiogram (CT-PA) is the gold standard for diagnosis of RVOT aneurysm as it can establish the diagnosis and describe the size, number, location, and outcome. CPB plays a crucial role in treating complex cardiac conditions such as coronary artery disease and valvular heart disease, thereby improving patient outcomes and extending lives [7]. We have found that axillary artery cannulation provides a safe and feasible alternative to the historically used femoral artery, by enabling antegrade aortic flow through the true lumen, ensuring adequate arterial inflow, and minimizing the risk of retrograde atheroemboli [8]. Our choice of a multi-pronged approach—simultaneous PVR (pulmonary valve replacement), MPA augmentation, RVOT reconstruction with patch, and aneurysm excision—was crucial. PVR addresses the volume/pressure overload from the pulmonary stenosis, preventing recurrence of high RV pressure, which is a major contributor to aneurysm formation and expansion [5]. The use of a pericardial patch provides a biologically compatible material for anatomical reconstruction and augmentation, although long-term follow-up is essential due to the known risk of patch degeneration or late aneurysmal recurrence [6]. Precise and careful preoperative assessment, using echocardiography and CT, is important for ensuring the safety of surgery performed on these patients [9].

Conclusion

RVOT aneurysm associated with pulmonary stenosis represents a complex pathology that mandates definitive surgical intervention. A comprehensive approach of aneurysm excision, pulmonary valve replacement, and RVOT, along with pulmonary artery augmentation using a pericardial patch, can result in excellent anatomical restoration and functional outcomes. Lifelong follow-up is essential to monitor prosthetic valve function and patch durability, and long-term anticoagulation is required. To the best of our knowledge, a case of this nature has not been previously reported.

Declarations

Author contributions: Dr RKV – Main operating surgeon, idea, critical review, Content review, Supervising the work; Dr KSG – Assisting surgeon, review of article, Supervising the work; Dr PB - verification of articles; Dr LB - writing of the initial draft; Dr PC- creation of the published work from original research articles.

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Consent to participate: Consent was obtained from the patient for publication of this case report and accompanying images.

Written consent for publication consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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