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SURGICAL CHALLENGES IN A RARE CASE OF LUTEMBACHER SYNDROME

Cardiovascular

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ABSTRACT

Purpose: To describe the surgical challenges and clinical management in a young adult with Lutembacher syndrome who had a previously placed Amplatzer septal occluder device and later developed severe mitral stenosis requiring mitral valve replacement (MVR). **Methods:** This is a retrospective case report of a 20-year-old female who underwent percutaneous closure of an atrial septal defect (ASD) at the age of 7. She presented with worsening dyspnoea over 14 years. A thorough diagnostic work-up was done, followed by successful surgical management. **Results:** The patient had a well-epithelialized ASD occluder device in situ with severe mitral stenosis. Intraoperatively, the device did not obstruct surgical access to the mitral valve. MVR was performed with a TTK Chitra mechanical valve. The device was left in place. The postoperative course was uneventful, and follow-up showed normal prosthetic function and pulmonary pressures. **Conclusion:** Mitral valve replacement can be safely performed in patients with an in-situ ASD occluder device. This case highlights the importance of preoperative imaging and intraoperative vigilance in managing such dual-pathology patients.

KEYWORDS

Lutembacher syndrome, atrial septal defect, mitral stenosis, Amplatzer device, mitral valve replacement, cardiac surgery

INTRODUCTION

Lutembacher syndrome (LS) is a rare cardiovascular condition characterized by the coexistence of an atrial septal defect (ASD) and mitral stenosis (MS) (1,2). In its classical form, the ASD is congenital, while MS is most often acquired secondary to rheumatic heart disease (3). The presence of an interatrial communication allows partial decompression of the left atrium in the setting of MS, which may mask clinical symptoms and delay diagnosis (4,5).

In recent decades, advances in percutaneous transcatheter interventions have led to an increasing number of patients undergoing device closure of isolated ASDs in childhood. Some of these patients subsequently develop MS-most commonly due to rheumatic pathology-resulting in a "modified" Lutembacher syndrome where a septal occluder device coexists with significant mitral valve pathology (6). This modern variant presents unique diagnostic and surgical challenges compared with the classical form.

The presence of an in-situ ASD occluder device, such as the Amplatzer septal occluder, may potentially complicate surgical access to the mitral valve during mitral valve replacement (MVR), especially when a transseptal approach is considered (2,7). Preoperative echocardiographic evaluation is essential for determining the position, stability, and endothelialization status of the device (5,8). Intraoperatively, careful assessment guides the decision to remove or retain the device, depending on whether it obstructs the surgical field or poses hemodynamic concerns (2).

We present the case of a 20-year-old female with a previously placed Amplatzer septal occluder who later developed severe MS requiring MVR. This case emphasizes the importance of comprehensive preoperative imaging, meticulous surgical planning, and intraoperative adaptability in managing patients with this dual pathology.

Case Report

Patient Information

A 20-year-old female presented with progressively worsening exertional dyspnea (NYHA Class III) over the past 14 years. She was previously diagnosed with a large atrial septal defect (ASD) at age six and underwent transcatheter closure using an Amplatzer septal occluder device in 2011 at the age of 7.

Clinical Findings

On physical examination, she was hemodynamically stable with a loud first heart sound (S1), mid-diastolic murmur over the apex, and a wide fixed split of the second heart sound (S2). There were no signs of right heart failure or peripheral oedema.

Diagnostic Assessment

Transthoracic echocardiography revealed a well-seated and endothelialized Amplatzer ASD occluder device with no residual shunt. Severe mitral stenosis was confirmed with a mitral valve area of 0.9 cm^2 and mean pressure gradient of 14 mmHg. Left ventricular ejection fraction was preserved. Chest X-ray showed cardiomegaly with pulmonary venous hypertension. Routine blood investigations revealed mild anaemia but were otherwise within normal limits.

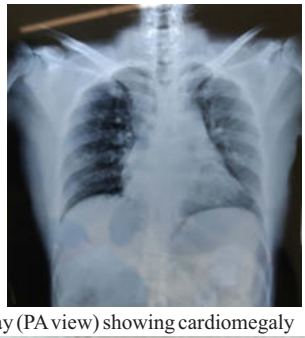

Fig. 1 Chest X-ray (PA view) showing cardiomegaly

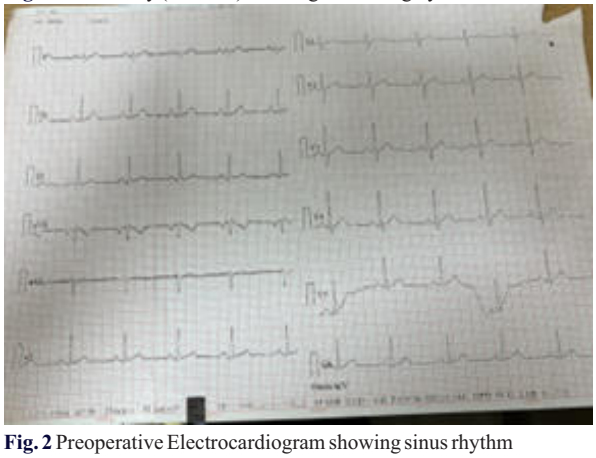

Fig. 2 Preoperative Electrocardiogram showing sinus rhythm

Therapeutic Intervention

The patient was scheduled for open-heart mitral valve replacement (MVR). Intraoperatively, the Amplatzer device was found well-epithelialized and positioned within the interatrial septum without interference in the operative field. The mitral valve was accessed through the left atrium, and a size 31 TTK Chitra mechanical valve was implanted. The ASD device was left in situ as it posed no hemodynamic or technical challenge.

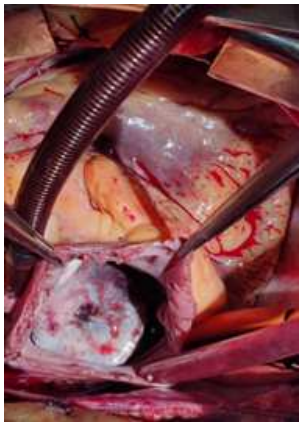

Fig. 3 Intraoperative image showing the ASD device epithelialized in situ

Fig. 4 Postoperative wound site during first follow-up visit

Follow-up And Outcomes

The patient's postoperative course was uneventful. She was extubated on postoperative day 1 and discharged on the 10th postoperative day. At 3-month follow-up, she was asymptomatic with improved effort tolerance (NYHA Class I). Echocardiography showed normal functioning of the prosthetic valve and normalized pulmonary artery pressures.

DISCUSSION

Lutembacher syndrome is a rare clinical entity defined by the coexistence of an atrial septal defect (ASD) and mitral stenosis (MS). The ASD is often congenital, while the MS is usually acquired, most commonly due to rheumatic heart disease. In contemporary settings, this syndrome increasingly involves patients who have undergone ASD closure through percutaneous devices and subsequently develop mitral stenosis — a modified presentation of the classical definition.

The presence of an in-situ ASD device, such as the Amplatzer septal occluder, poses unique surgical challenges during mitral valve replacement (MVR). The Amplatzer device is a self-expanding nitinol wire mesh with polyester patches, designed to promote endothelialization and long-term incorporation into the septal tissue. However, this device's position within the interatrial septum can sometimes hinder surgical access to the mitral valve, especially when approached via the transseptal route.

In this case, despite a large ASD having been closed in early childhood with an Amplatzer device, surgical evaluation showed that the device was well incorporated into the septum and did not interfere with visualization or access to the mitral valve. The decision to leave the device in situ avoided unnecessary manipulation or potential iatrogenic septal damage. The left atrial approach was employed effectively, allowing complete and safe excision of the stenosed mitral valve and implantation of a mechanical prosthesis.

This case illustrates that careful preoperative imaging and intraoperative assessment are crucial in planning MVR in patients with previously placed septal occluder devices. It also suggests that such devices do not always necessitate removal, provided they are stable and non-obstructive, and that mitral valve surgery can still be safely accomplished.

Statements And Declarations

Competing Interests The authors declare that they have no competing financial or non-financial interests.

Funding: No funding was received for this case report.

Ethical Approval: Not applicable.

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

Author Contributions: Prof. Dr. Harpreet Singh performed the surgery and supervised the clinical management and reviewed the manuscript.

Dr. AB Karthikeyan was the first assistant surgeon and was involved in the pre-op and post-op management and is the corresponding author for this report.

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