CARDIOLOGIE TUNISIENNE

CASE REPORT

Severe Calcified Rheumatic Mitral Stenosis with severely Mixed Pulmonary Hypertension: A case with High-Risk Surgical challenge

Sténose mitrale rhumatismale sévèrement calcifiée avec hypertension pulmonaire mixte sévère : un défi chirurgical à haut risque

Aymen Boubzizi, Hela Bouzidi, Ihsen Zairi, Alaeddine Ayadi, Ghassen Tlili, Sofien Kamoun, Sondos Kraiem

Cardiology department of Habib Thameur hospital of Tunis

Summary

Background: Rheumatic mitral stenosis remains a major cause of morbidity in developing countries. Despite percutaneous balloon mitral commissurotomy (PBMC), restenosis with severe calcification and pulmonary hypertension may occur, posing therapeutic challenges.

Case Presentation: A 66-year-old woman with diabetes, hypertension, atrial fibrillation, and prior PBMC was admitted with global heart failure. Echocardiography showed severe calcified mitral stenosis (valve area 1.0 cm², gradient 10 mmHg), left atrial appendage (LAA) thrombus, and severe pulmonary hypertension with right ventricular dysfunction. PBMC was contraindicated. Surgery was proposed but considered prohibitive due to extreme pulmonary hypertension and an Euro-SCORE II of 6 (mortality estimation greater than 10%)

Conclusion: This case illustrates advanced rheumatic mitral stenosis after PBMC, complicated by atrial fibrillation, LAA thrombus, and severe pulmonary hypertension. It underscores the dilemma between high-risk surgery and palliative management, highlighting the need for multidisciplinary decision-making.

Keywords

Rheumatic mitral stenosis; Pulmonary hypertension; Heart failure; PBMC; Valve surgery

Résumé

Introduction : La sténose mitrale rhumatismale reste une cause majeure de morbidité dans les pays en développement. Malgré la commissurotomie mitrale percutanée (CMP), une resténose avec calcification et hypertension pulmonaire peut survenir, posant un défi thérapeutique.

Présentation du cas : Une femme de 66 ans, diabétique, hypertendue, en fibrillation auriculaire et ayant bénéficié d'une CMP antérieure, a été admise pour insuffisance cardiaque globale. L'échocardiographie a montré une sténose mitrale sévèrement calcifiée (surface valvulaire 1,0 cm², gradient moyen 10 mmHg), un thrombus auriculaire gauche et une hypertension pulmonaire sévère avec dysfonction ventriculaire droite. La CMP était contre-indiquée. Le remplacement valvulaire mitral chirurgical a été proposé mais jugé prohibitif en raison de l'hypertension pulmonaire extrême et d'un Euro-SCORE II à 6 (estimant une mortalité supérieure à 10%)

Conclusion : Ce cas illustre l'évolution d'une sténose mitrale rhumatismale après CMP, compliquée de fibrillation auriculaire, thrombus et hypertension pulmonaire sévère. Il met en évidence le dilemme entre chirurgie à haut risque et prise -en charge palliative, soulignant l'importance d'une décision multidisciplinaire.

Mots-clés

Sténose mitrale rhumatismale; Hypertension pulmonaire ; Insuffisance cardiaque; CMP ; Chirurgie valvulaire

Correspondance

Aymen Boubzizi
Cardiology department of Habib Thameur hospital of Tunis

INTRODUCTION

Rheumatic mitral stenosis (MS) remains a frequent cause of valvular heart disease in developing countries. Percutaneous balloon mitral commissurotomy (PBMC) is the preferred treatment for suitable candidates, but late restenosis, extreme valve calcification, atrial fibrillation, and pulmonary hypertension significantly deteriorate the prognosis. Severe pulmonary hypertension associated with right ventricular (RV) dysfunction is one of the most challenging-to-treat conditions, where the benefit of surgery must be carefully balanced against its disastrous risks.

We present the case of a 66-year-old woman with calcified end-stage rheumatic MS, atrial fibrillation, and severe mixed pulmonary hypertension in which both surgical and percutaneous methods were limited.

CASE PRESENTATION

A 66-year-old female patient with a previous history of type 2 diabetes mellitus, systemic hypertension, and permanent atrial fibrillation on anticoagulation was admitted with deteriorating dyspnoea (NYHA III), peripheral oedema, and abdominal distension. She had a previous PBMC 10 years ago for rheumatic mitral stenosis.

Clinical examination has revealed blood pressure of 140/70 mmHg and an irregular rhythm at 110 bpm. On cardiac auscultation, a loud pulmonary component of S2 and a diastolic rumble at the apex. Pulmonary auscultation found bilateral crackles at the basal regions. She had also developed grade 3 pitting oedema of the lower limbs.

The electrocardiography showed atrial fibrillation with a rapid ventricular rate (110 bpm), narrow QRS complexes, and T-wave inversion in inferior and lateral leads.

Chest radiography showed large left atrial enlargement and bilateral pulmonary congestion but no suspected image of infection (Figure 1).

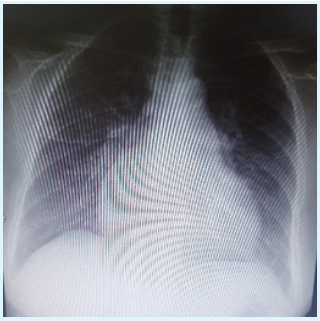

Figure 1. chest radiography

The blood test showed haemoglobin at 13 g/dL, white blood cell count 6,800/mm^3, platelets 167,000/mm^3, creatinine 95 µmol/L, CRP 6 mg/L, and subtherapeutic anticoagulation (TP 37%) with a normal liver function.

On transthoracic echocardiography we found a normal size of the left ventricle and good systolic function with a highly dilated left atrium (75 ml/m^2), a spontaneous echo contrast, and a highly thickened and calcified mitral valve with a hockey stick morphology, commissures already split, a mean gradient of 10 mmHg, and a valve area of 1.0 cm^2 (figure 2) by planimetry. There is also an extensive subvalvular involvement with minimal mitral regurgitation. (WILKINS SCORE = 12).

The right atrium and ventricle were dilated with a moderate RV dysfunction. The estimated systolic pulmonary artery pressure was more than 105 mmHg.

Transoesophageal echocardiography confirmed the diagnosis (figure 2) and revealed an organised thrombus in the left appendage. Significant mitral valve calcification and thickening with restricted leaflet mobility were present with an opened commissure.

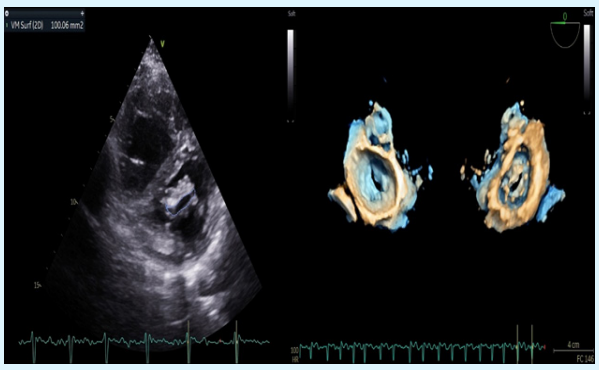

Figure 2. mitral valve area short axis TTE view, B: 3D TEE reconstruction of mitral valve (left: atrial view, right: ventricular view)

A right heart catheterisation had revealed a pulmonary artery pressure of 109/70 mmHg (mean 85) with a pulmonary capillary wedge pressure at 24 mmHg.

The pulmonary vascular resistance calculated from the arterio-venous (A-V) difference was 9 Wood units, consistent with severe combined pre- and post-capillary pulmonary hypertension.

The EuroSCORE II was 10%, equivalent to an estimated operative mortality of 9.1% besides, the morbidity and mortality from the STS score was predicted at 25% .

So, we are in discussion with the heart team to

perform surgical mitral valve replacement with tricuspid repair and left atrial appendage exclusion despite the high surgical risk.

DISCUSSION

The severe valve calcification, commissural opening, and the presence of a left atrial thrombus rendered PBMC strictly contraindicated in our patient(1). The Heart Team therefore suggested mitral valve replacement, combined with tricuspid annuloplasty and left atrial appendage exclusion. However, the coexistence of extreme pulmonary hypertension (PH) and right ventricular dysfunction represented a prohibitive surgical risk, raising serious concerns about perioperative and postoperative outcomes. PH increases morbidity and mortality in adult patients undergoing heart surgery(2) .it is an independent risk factor for the development of acute right ventricular failure especially with cardio-pulmonary bypass which exacerbates PH(2).

In addition, Patients with severe valvular and sub valvular apparatus (SVA) calcification are known to present with more advanced hemodynamic compromise before intervention. Interestingly, some studies have suggested that despite these unfavourable anatomical features, patients with severe SVA deformities may still derive significant hemodynamic benefit from PBMC, with no negative impact on immediate or intermediate outcomes (3). Nevertheless, such deformities increase the technical complexity of the procedure and can negatively influence procedural success.

Furthermore, Symptomatic restenosis is a well-recognized complication after surgical commissurotomy or PBMC. While most cases ultimately require surgical valve replacement, redo PBMC may be considered in carefully selected patients with favourable anatomy, particularly when commissural refusion is the predominant mechanism (1). Unfortunately, our patient presented with severe calcification, open commissures, and an organized thrombus in the left atrial appendage features that definitively precluded a percutaneous approach.

Also, Emerging transcatheter strategies, such as Transcatheter Mitral Valve Lithotripsy (TMVL), have been tested as a pretreatment to PBMC in patients with heavily calcified rheumatic MS, showing promise in optimizing procedural results and reducing the risk of mitral regurgitation (4). Similarly, Transcatheter Mitral Valve Implantation (TMVI) may be considered in symptomatic patients with severe mitral valve dysfunction at specialized centres (1). However, these novel approaches remain experimental, require advanced technology, and are not available in most resource-limited settings.

In this context, surgical valve replacement remains the only definitive therapy. Yet, the presence of extreme PH and right ventricular dysfunction significantly increases operative risk and worsens long-term prognosis. Although mitral surgery patients tend to have higher preoperative pulmonary artery pressures, PH was associated with a lower risk for mitral outcomes compared with CABG (5).Moreover, there is a pharmacological treatment for PH during cardiac surgery that has been tested . In the most recent trial, in patients with PH undergoing mitral valve replacement for mitral stenosis it was demonstrated that Inhaled prostacyclin and nitric oxide were given just before the end of cardiopulmonary bypass can reduce significantly the PH indices as well as increase in cardiac output (CO) and RV ejection fraction compared to conventional treatment (6). Besides, for the non-pharmacological approach, the chest temporarily can be left open to reduce the surrounding pressures(6). Finally, pulmonary artery balloon pumps, RV assist devices, and Cavo pulmonary diversion have been mentioned as a potential treatment for severe PH and right ventricular dysfunction(6).

This case underlines the importance of early referral and timely intervention in rheumatic heart disease, before irreversible pulmonary vascular remodelling and right ventricular dysfunction develop. It also emphasizes the crucial role of a multidisciplinary Heart Team in assessing risks, discussing novel treatment strategies, and tailoring management to each patient's clinical context especially patients present late with end-stage disease, leaving clinicians to choose between high-risk surgery and purely palliative management.

CONCLUSION

Severe rheumatic mitral stenosis complicated by severe pulmonary hypertension and RV dysfunction is a difficult therapeutic dilemma. This case highlights the limitations of percutaneous and operative management in far-advanced disease and emphasises the need for early referral, early intervention, and multidisciplinary assessment to maximise outcome

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