Aortic and Mitral Disease due to an Unusual Etiology

Juan I. Cotella, Julio Dantur, Eduardo Hasbani, Javier Hasbani, Marcela Ortiz Mayor, Aldo Prado
Centro Privado De Cardiología, Tucumán, Argentina

Abstract

Relapsing polychondritis (RP) is a rare multisystem disease characterized by inflammation in cartilaginous structures and other connective tissues throughout the body, affecting the ears, nose, eyes, joints, respiratory tract, heart, and blood vessels. Cardiovascular involvement is the second most common cause of mortality after laryngotracheal involvement. Here, we report a successful surgical case of RP in which the patient underwent aortic and mitral valve replacement and concomitant coronary artery bypass grafting.

Keywords: Multivalvular involvement, relapsing polychondritis, tissue disease

INTRODUCTION

Relapsing polychondritis (RP) is a rare multisystem disease characterized by inflammation in cartilaginous structures and other connective tissues throughout the body, affecting the ears, nose, eyes, joints, respiratory tract, heart, and blood vessels. Cardiovascular compromise is the second most common cause of mortality after laryngotracheal involvement. We report a successful surgical case of RP in which the patient underwent aortic and mitral valve replacement and concomitant coronary artery bypass grafting.

CASE REPORT

A 54-year-old patient without a previous history of heart disease was admitted to our institution with a diagnosis of congestive heart failure (Class IV according to the New York Heart Association). He was diagnosed with RP 6 years ago. Diagnosis was made due to recurrent polyarthritis and by right auricle biopsy. The patient was given an anti-tumor necrosis factor (TNF) agent (etanercept) to treat recurrent arthritis. During admission, dyspnea at rest, jugular venous elevation, rales, lower limb edema, and third heart sound were found. Transthoracic echocardiography (TTE) confirmed a left ventricular enlargement (two-dimensional parasternal long-axis view diameters: left ventricular end-diastolic diameter = 69 mm/left ventricular end-systolicdiameter = 40 mm), left ventricular ejection fraction of 53% (left ventricular 18 end-diastolic volume = 157 mL and left ventricular end-systolic 19 volume = 67 mL), marked thickening of anterior mitral leaflet with restriction in its mobility, without commissural fusion or any findings associated with rheumatic disease [Figure 1 and Videos 1-3]. Normal transmitral gradients were found (mean gradient: 2.1 mmHg). The aortic valve showed increased echogenicity and retraction of its three cusps, with abnormal closure. Moderate mitral regurgitation (MR) (effective regurgitant orifice: 0.3 cm² and regurgitant volume/beat: 48 mL) and severe aortic regurgitation (AR) (pressure half-time = 152 ms and vena contracta width = 8 mm; holodiastolic flow reversal in the descending aorta) were reported [Figure 2 and Video 3].

According to the clinical and echocardiographic findings, surgical treatment with double valve replacement was decided. A severe proximal obstruction of the left descending artery was found upon coronary angiography [Figure 3].

The aortic valve was excised and the connective tissue surrounding the sinus of Valsalva and the annulus was removed, and a prosthetic mechanical aortic valve (St Jude 23 mm) was implanted. The mitral valve was replaced by a mechanical valve prosthesis (St Jude 29 mm). A left

Address for correspondence: Mr. Aldo Prado, Virgen de La Merced 550, Tucuman, Argentina.
E-mail: prado.a@icloud.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

Submitted: 27-Jul-2020 Revised: 03-Sep-2020 Accepted: 11-Sep-2020 Published: 09-Nov-2020

How to cite this article: Cotella JI, Dantur J, Hasbani E, Hasbani J, Mayor MO, Prado A. Aortic and mitral disease due to an unusual etiology. J Cardiovasc Echography 2020;30:179-82.
artery bypass to the left anterior descending artery was done after valve replacement.

All tissues removed during operation were sent for histological analysis. At the mitral level, a multifocal inflammatory infiltrate was observed, predominantly at subendocardial level, with vascular evolvement. New vessels, edema, and necrosis of connective tissue associated with myxomatous changes and fibrosis were also detected [Figure 4].

At the aortic valve, fibrosis, perivascular inflammatory infiltrates, and a high level of plasmocytes and neovessels were also found. In addition, vascular damage with hemosiderin pigment and lymphoplasmacytic infiltrate with Russell and Dutcher bodies was described, as well as destruction of the connective tissue, focus of karyorrhexis, and nodular calcifications [Figure 5].

At the left coronary ostium level, endothelial and muscular hyperplasia and focal inflammatory tissue infiltration were observed [Figure 6].

Two days after, anticoagulation and anti-TNF therapies were started. No adverse events were reported during the postoperative recovery period. Home discharge was given on the 5th postoperative day.

At 3-month follow-up, the patient remained well, and an echocardiography was performed, which showed normal left ventricular function, no paravalvular regurgitation, and normal prosthetic gradients.

**DISCUSSION**

RP is a rare connective tissue disease, affecting cartilaginous structures rich in proteoglycans, with chronic and recurrent behavior.[1] Thus, organs such as the nose, ears, larynx, tracheobronchial tree, and the cardiovascular system are usually more affected, either individually or simultaneously. Among the proposed mechanisms, circulating autoantibodies against type II, IX, and XI collagens have been described, as well as deposition of IgG, IgM, IgA, and C3 immune complexes, among others.[2]

Cardiovascular involvement occurs in 7.1%–51% of the patients with this disease.[3] The leading cause of death is laryngotracheal involvement with subsequent airway obstruction followed by cardiac complications. In a recent publication,[4] cardiovascular compromise was observed in 7.1% of the cases, affecting mainly patients older than 70, which reflects the late presentation of these manifestations in this disease.

Other reports found that cardiovascular involvement occurs in 24%–52% of the patients most typically presenting AR, MR, pericarditis, aortic aneurysms, and conduction disturbances.[3]

AR and aortic root involvement (dilation or retraction) represent the most common manifestations. MR is less frequent than AR, and mitral valve replacement has seldom been reported.[5] In cases of MR with RP, not enough data are available to support urgent surgical intervention. The association of mitral valve, aortic valve, and coronary involvement in the same patient is not a usual presentation.

Mitral and aortic valve replacement in patients with RP is associated with a poor prognosis, and postsurgical events such as valve dehiscence and periprosthetic leak were reported as frequent complications.[5,6] In our patient, the connective tissue surrounding the sinus of Valsalva and the annulus and at mitral annulus was removed to prevent future complications.
Since the mechanisms involved in the appearance and recurrence of this disease are not well understood, there is no specific treatment established, although steroids have been shown to be effective in suppressing disease activity and reducing the severity of recurrences. Biological anti-TNF or anti-CD4 agents are promising, though they have been associated with the rise of infections.

There are case reports that show rapidly progressive coronary stenosis after valve replacements. In the case of our patient, the coronary affection was observed during coronary angiography. Evaluation of the coronary artery should be performed in all patients, and the ostium should be checked by the surgeon during the operation to decide if a bypass is necessary.

We consider it important to emphasize the need for periodic cardiovascular and echocardiographic evaluations in patients with connective tissue conditions, during early stages of systemic disease diagnosis and after recurrent episodes.

**CONCLUSION**

We performed a successful surgery in a patient with RP who showed a rare combination of AR, MR, and coronary stenosis. Echocardiography is crucial to identify cardiac involvement related to RP. TTE was essential to understand the underlying mechanism and etiology of mitral and aortic valve disease and develop a successful surgical plan.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.
REFERENCES

1. McAdam LP, O’Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: Prospective study of 23 patients and a review of the literature. Medicine (Baltimore) 1976;55:193-215.
2. Letko E, Zafirakis P, Baltatzis S, Voudouri A, Livir-Rallatos C, Foster CS. Relapsing polychondritis: A clinical review. Semin Arthritis Rheum 2002;31:384-95.
3. Shimizu J, Oka H, Yamano Y, Yudoh K, Suzuki N. Cardiac involvement in relapsing polychondritis in Japan. Rheumatology (Oxford) 2016;55:583-4.
4. Oka H, Yamano Y, Shimizu J, Yudoh K, Suzuki N. A large-scale survey of patients with relapsing polychondritis in Japan. Inflamm Regen 2014;34:149-56.
5. Dib C, Moustafa SE, Mookadam M, Zehr KJ, Michet CJ Jr, Mookadam F. Surgical treatment of the cardiac manifestations of relapsing polychondritis: Overview of 33 patients identified through literature review and the Mayo Clinic records. Mayo Clin Proc 2006;81:772-6.
6. Dormoy L, Belin A, Labombarda F. Heart transplantation as last resort treatment for relapsing polychondritis with severe cardiac involvement. Prog Transplant 2017;27:321-3.
7. Vaidyanathan RK, Byalal JR, Sundaramoorthi T, Sankar NM, Thomas JM, Meenakshi NA, et al. Rapidly progressive coronary ostial stenosis after aortic valve replacement in relapsing polychondritis. J Thorac Cardiovasc Surg 2006;131:1395-6.
8. Lang-Lazdunski L, Hvass U, Paillole C, Pansard Y, Langlois J. Cardiac valve replacement in relapsing polychondritis. A review. J Heart Valve Dis 1995;4:227-35.