

Rheumatic Heart Disease with Quadrivalvular Stenosis: A Case Report and Review of Literature

W. Belkho^{1*}, Z. Wakrim¹, M. El Jamili¹, S. El Karimi¹, M. El Hattoui¹

¹Department of Cardiology, Mohammed VI University Hospital Center, Marrakech, Morocco

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*Corresponding author: W. Belkho

Department of cardiology, Mohammed VI University Hospital Center, Marrakech, Morocco

Abstract

Case Report

Multivalvular heart disease (MVD), particularly of rheumatic nature, continues to be a widespread condition in developing countries such as Morocco, despite implemented preventive measures. The intricate hemodynamic interactions between coexisting valve lesions may influence the clinical expression of each individual lesion. It makes diagnosis difficult and management often at the discretion of the physician given the lack of specific guidelines to assist in strategy. In the available literature, rheumatic quadrivalvular stenosis is exceptional. Here we describe a very rare case of rheumatic involvement of all four heart valves discovered in a 34-year-old woman with a history of untreated recurrent tonsillitis and joint pain in childhood, who presented with progressively worsening dyspnea and fatigue. Transthoracic echocardiography revealed severe left valves stenosis in addition to severe tricuspid and mild pulmonary stenosis, with evidence of congestive heart failure. Our case highlights the diagnostic difficulties encountered in the context of a developing country with limited access to primary care and treatment options.

Keywords: Valvular heart disease, rheumatic heart disease, complications.

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INTRODUCTION

Acute Rheumatic Fever (ARF) is a multisystemic, non-suppurative, post-streptococcal inflammatory disease; Rheumatic heart disease (RHD), is an almost unavoidable long-term complication in about half of ARF cases and is a major prognostic factor [1].

Despite preventive measures, RHD is a matter of concern in many parts of the world, particularly in developing countries such as Morocco [2].

The hallmark lesion in RHD is valve regurgitation, but chronic inflammation leads to valvular stenosis (most often mitral stenosis); Right-sided valve disease is not rare, generally affecting the tricuspid valve, however, the pulmonary valve is often spared [1].

In the available literature, there are scarce data on multiple valvular disease, a fact highlighted by the European guidelines [3], which include a limited number of evidence-based recommendations.

We report a case of a very rare entity in rheumatic heart disease presenting with quadrivalvular stenosis in a woman with a history of untreated ARF.

CASE REPORT

A 34-year-old woman from a rural area of Morocco was admitted to the emergency with exertional dyspnea, cough, and fatigue for two months with recent worsening; no weight loss was reported.

Background finding showed a history of untreated repetitive tonsillitis in childhood, self-medication for joint aches; no history of throat infection in the past month was found, no relatives or family members had similar symptoms.

General examination found an orthopneic, polypneic patient at 25 cycles per minute, her blood pressure was 92/62 mmHg, with a pulse of 99 bpm, she had signs of congestive heart failure with bilateral lower extremity oedema, jugular vein distention, and bilateral crackles. Cardiac auscultation revealed a latero-sternal mesosystolic murmur radiating to the carotids.

The admission ECG was in sinus rhythm with a rate of 96 beats/min and electrical left ventricular hypertrophy, there were no dysarrhythmia or atrioventricular block. A chest X-ray showed cardiomegaly, a pneumonia with parapneumonic pleural effusion.

Transthoracic echocardiography revealed calcified, thickened valve leaflets (Fig 1) and restricted diastolic opening, with severe mitral stenosis and a mitral valve area of 0,8 cm² by planimetry and a mean pressure gradient of 13 mmHg associated with a moderate mitral regurgitation with a proximal isovelocity surface area (PISA) of 7 mm.



Figure 1: Abnormally thickened mitral valve leaflets in parasternal long-axis view on transthoracic echocardiography

In addition to this, aortic valve leaflets were severely thickened, with a severe aortic stenosis and a mean pressure gradient of 46,1 mmHg, peak aortic

valve velocity of 4,11 m/s associated to a moderate aortic regurgitation (Fig 2).

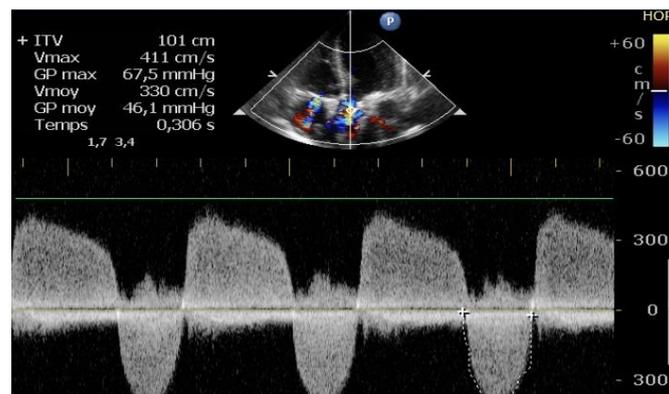


Figure 2: Severe aortic stenosis measured in 5 chambers view on transthoracic echocardiography

We also found a severe tricuspid stenosis with a mean pressure gradient of 7 mmHg and a severe tricuspid regurgitation. The mobility of pulmonary cusps was restricted with mild pulmonary stenosis and a peak gradient of 24,9mmHg (Fig 3). The patient had bi-

atrial enlargement and a dilated inferior vena cava. Biventricular function was normal, with a Left Ventricular Ejection Fraction (LVEF) of 54%. Left ventricular dimension during systole and diastole were respectively LVDs = 32 mm/m², LVDd = 43 mm/m².

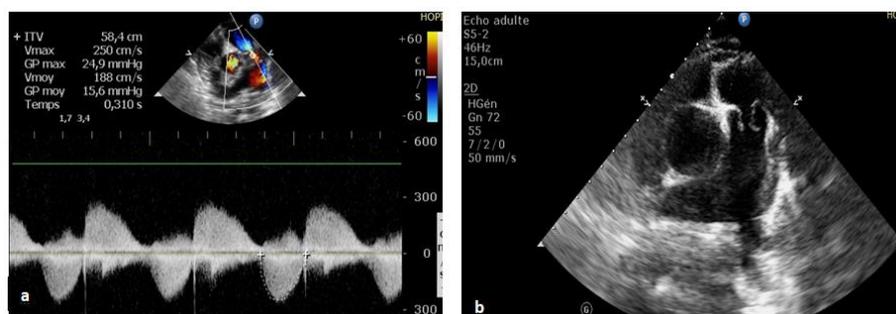


Figure 3: Transthoracic echocardiography of parasternal short axis view showing pulmonary stenosis and regurgitation (a) and restricted mobility of the pulmonary leaflets (b)

Infectious workup was positive in favor of a pneumopathy, the standard assessment for overload and carcinoid heart disease was negative - The diagnosis of quadrivalvular stenosis in a setting of rheumatic heart disease was retained.

DISCUSSION

Multiple valve heart disease (MVD) is defined by at least 2 moderate VHDs and is most frequently acquired, with rheumatic fever in the acute phase being the predominant mechanism. Hemodynamic interactions between valve lesions can either exaggerate or attenuate the clinical expression of each singular lesion, which can make the diagnosis, quantification, and management of MVD difficult [4].

Rheumatic heart disease (RHD) carries a significant burden in developing countries, contributing to most cardiovascular morbidity and mortality in young people [5], it continues to be a public health crisis in countries such as Morocco, with a reported incidence of 18,3 per 100 000 inhabitants in 2010 [2].

A gap in strong population-level epidemiological data for ARF and RHD is lamented, and it is likely that the burden of disease is much higher than estimates published to date [6].

Where medical care is deficient or absent, the natural course of the disease is likely to prevail due to poor accessibility to treatment. Modifiable risk factors identified for rheumatic fever include poverty, overcrowding, malnutrition, and maternal education and employment [5].

In general, the initial manifestations most frequently seen in ARF (the "major criteria" for diagnosis) are carditis and arthritis, followed in frequency by chorea, subcutaneous nodules, and erythema marginata; a more insidious clinical course with no clear extra-cardiac signs will require Doppler echocardiography. This imaging modality is considered the cornerstone of diagnosis and follow-up of patients with ARF in order to satisfy the recently revised Jones criteria [7].

The writing group of these criteria recommends [7] with a class I, level of evidence B, that echocardiography with doppler should be performed in all cases of confirmed or suspected ARF.

The characteristic acute mitral valve disease shows mitral annulus dilatation, cordage elongation, and anterior leaflet prolapse; aortic valve involvement is seen in 20% to 25% of cases, the predilection for left-sided valvular heart disease (VHD) is probably related to the expression of specific endothelial surface proteins and exposure to higher pressures [8]; Right-sided VHD is not as common, it tends to affect the tricuspid valve in primary valvulitis or as a result of the adverse

hemodynamic consequences of left-sided VHD, particularly mitral stenosis; it rarely involves the pulmonary valve [1].

Acute rheumatic valvulitis manifests as valvular regurgitation, but over time, chronic inflammation leads to valve stenosis by thickening of the leaflets and restriction of motion, commissural fusion, with varying degrees of involvement of other parts of the valve apparatus in the later stages of the disease [1, 7]. Quadrivalvular involvement is extremely rare in the chronic phase [9].

Although chronic RHD occurs only as a sequel to ARF, the majority of patients with RHD lack a history of past ARF [10]; They generally present at the age of 20-50 years after the onset of dyspnea [5]; The present case is particularly interesting because of the diagnostic delay due to the absence of suggestive symptoms in the early phase, thus making it difficult to label a quadruple valve blockage at first presentation.

Patients with multiple or mixed VHD should be followed by a specialized heart valve team with an individualized treatment strategy that includes multiple parameters: the complexity of each valve lesion, the patient-specific level of risk, the increased long-term morbidity associated with multiple prostheses, and the natural course of each valve lesion in the absence of treatment [4].

Spontaneous RHD in most developing countries is still progressing to severe multiple VHD with consequent risk of congestive heart failure, thromboembolic events and infective endocarditis [8].

Informed consent: The patient gave her informed consent and permitted the writing of this case report.

CONCLUSION

RHD has a high prevalence in developing countries and is a condition of social inequality.

Multiple VHD is a late development of ARF, but the concomitant involvement of all four valves is an exceptional entity in which the scarcity of data does not allow a standardized management.

A case-by-case therapeutic management strategy should be made by a Heart Team based on echocardiography, depending on symptoms, severity, hemodynamic interactions of the valve lesions, surgical risk profile and co-morbidities.

Future studies, however, are needed to guide therapeutic decisions and evaluate the real benefits of emerging treatments.

Declaration of Conflicting Interests: The author(s) declare(s) that there is no conflict of interest.

REFERENCES

1. Kumar, R. K., Antunes, M. J., Beaton, A., Mirabel, M., Nkomo, V. T., Okello, E., ... & American Heart Association Council on Lifelong Congenital Heart Disease and Heart Health in the Young; Council on Cardiovascular and Stroke Nursing; and Council on Clinical Cardiology. (2020). Contemporary diagnosis and management of rheumatic heart disease: implications for closing the gap: a scientific statement from the American Heart Association. *Circulation*, *142*(20), e337-e357.
2. Ghanem, D. N., & Jroundi, D. I. (2010). Epidémiologie du Rhumatisme Articulaires Aigu au Maroc: Description des données de surveillance collectées entre 2000 et 2010, 33.
3. Beyersdorf, F., Vahanian, A., Milojevic, M., Praz, F., Baldus, S., Bauersachs, J., ... & Wojakowski, W. (2021). 2021 ESC/EACTS Guidelines for the management of valvular heart disease: Developed by the Task Force for the management of valvular heart disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). *European journal of cardio-thoracic surgery*, *60*(4), 727-800.
4. Unger, P., Pibarot, P., Tribouilloy, C., Lancellotti, P., Maisano, F., Iung, B., ... & European Society of Cardiology Council on Valvular Heart Disease. (2018). Multiple and mixed valvular heart diseases: pathophysiology, imaging, and management. *Circulation: Cardiovascular Imaging*, *11*(8), e007862.
5. Marijon, E., Mirabel, M., Celermajer, D. S., & Jouven, X. (2012). Rheumatic heart disease. *The Lancet*, *379*(9819), 953-964.
6. Zühlke, L. J., & Steer, A. C. (2013). Estimates of the global burden of rheumatic heart disease. *Global heart*, *8*(3), 189-195.
7. Gewitz, M. H., Baltimore, R. S., Tani, L. Y., Sable, C. A., Shulman, S. T., Carapetis, J., ... & Kaplan, E. L. (2015). Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography: a scientific statement from the American Heart Association. *Circulation*, *131*(20), 1806-1818.
8. Woldu, B., & Bloomfield, G. S. (2016). Rheumatic heart disease in the twenty-first century. *Current Cardiology Reports*, *18*(10), 1-11.
9. Picó-Aracil, F., García, J., Ruipérez, J. A., Campos, V., Ruiz, J. A., Jaen, E., & Ramos, F. (1983). Quadrivalvular heart stenosis. *American Heart Journal*, *106*(6), 1447-1447.
10. Zühlke, L., Engel, M. E., Karthikeyan, G., Rangarajan, S., Mackie, P., Cupido, B., ... & Mayosi, B. M. (2015). Characteristics, complications, and gaps in evidence-based interventions in rheumatic heart disease: the Global Rheumatic Heart Disease Registry (the REMEDY study). *European heart journal*, *36*(18), 1115-1122.