Case Report

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Subannular Mitral Aneurysm and Rheumatic Heart Disease Diagnosed during Postpartum Cardiomyopathy Clinical Case with Literature Review

Kimbally-kaky EG[1,2*], Mongo Ngamami SF[1,2], Makani Bassakouahou JK[3], Kouala Landa CM[1,2], Taty RJ[3], Ngolo Letomo K[1], Bakekolo RP[1,2], Baragou S[4,5] and Ellenga Mbolla BF[1,2]

[1]Cardiology Department B, University Hospital Center of Brazzaville, Republic of the Congo.

[2]Faculty of Health Sciences, Marien Ngouabi University, Brazzaville, Republic of the Congo.

[3]Cardiology Department A, University Hospital Center of Brazzaville, Republic of the Congo.

[4]Cardiology Department, Sylvanus Olympio University Hospital, Lomé.

[5]Faculty of Health Sciences, Lomé, Togo.

Correspondence

Kimbally-kaky EG, Cardiology Department B, University Hospital Center of Brazzaville, Republic of the Congo.

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Abstract

Introduction: Mitral subvalvular aneurysm is a rare cardiac condition with a significant incidence in Africans. Its association with mitral regurgitation is even rarer. Its origin, not yet clearly established, could be congenital or related to infectious, inflammatory, degenerative, or autoimmune processes.

Case Report: A 28-year-old Congolese woman, married and mother of two, had a history of heart problems, including an episode of acute rheumatic fever requiring a prolonged hospitalization at age 14. There was no subsequent medical follow-up.

One month after her second childbirth, she was hospitalized for rapidly developing exertional dyspnea, followed by persistent dyspnea (NYHA class IV). On admission, she presented with global heart failure, a regular tachycardia of 100 beats/min, and a systolic murmur of mitral regurgitation in the apex of her arm.

The anteroposterior chest X-ray showed cardiomegaly (ICT = 0.67), a mitral-type cardiac silhouette and venous redistribution at the apex. The electrocardiogram revealed a sinus rhythm and hypertrophy of the left cardiac chambers. The Doppler echocardiogram revealed: a subannular mitral aneurysm of the left ventricle, grade 2 mitral regurgitation with thickened leaflets, dilated left ventricular chambers, reduced left ventricular systolic function to 35 per cent, and a minimal pericardial effusion associated with postpartum cardiomyopathy.

Medical treatment for heart failure was successfully administered. The patient was seen at her one-month follow-up appointment in good haemodynamic condition, although the mitral regurgitation persisted, requiring further monitoring, as did the aneurysm. However, the patient was subsequently lost to follow-up.

Conclusion: The authors emphasise the need to raise awareness amongst clinicians—particularly echocardiographers—of this rare cardiac condition when they are treating young patients presenting with mitral regurgitation and heart failure.

Keywords

Echocardiography, Submitral aneurysm, Rheumatic heart disease, Brazzaville.

INTRODUCTION

Subannular mitral ventricular aneurysm (ASAM) is considered a rare heart condition. Its association with mitral valve disease is even rarer. Its aetiology has not yet been clearly established. Several hypotheses have been put forward regarding its aetiology. However, it is currently accepted that this condition results from a congenital weakness of the posterior fibrous ring of the mitral valve and of the endocardium [1]. The condition is thought to occur predominantly in predisposed individuals exposed to certain infectious agents or chronic inflammatory conditions such as : rheumatic carditis, tuberculosis, infective endocarditis, Takayasu's arteritis [2-5].

In Africa, ASAM was first described in Nigeria

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in 1962 on the basis of a series of 12 cases [6]. For many years, this condition was associated almost exclusively with young adults of African origin, a perception that has been challenged over the last decade due to cases of ASAM observed in patients from various continents, such as India and Brazil [7,8] and even in children aged between 3 and 11 years [4,9-12].

Clinically, these aneurysms may be asymptomatic, or may lead to morbidity due to mitral regurgitation (MR), with or without left ventricular dysfunction. Clinical manifestations are varied and include: heart failure, thromboembolic complications, ventricular wall rupture, valve perforation, myocardial infarction due to compression of the coronary arteries, severe ventricular arrhythmias or, in some cases, sudden death [1,4,13-15]. Currently, thanks to new cardiac imaging technologies (computed tomography, magnetic resonance imaging), as well as the widespread availability of transthoracic echocardiography (TTE) – as is the case in our setting – the diagnosis of ASAM has become straightforward [3,8,15-19]. Surgery is considered the corrective method of choice for the management of both the aneurysm (particularly if it is large and/or ruptured) and mitral regurgitation [5,8,10,11,13,15-17,20-23].

We report the case of ASAM associated with rheumatic mitral valve disease, diagnosed in a 28-year-old Congolese woman during the postpartum period.

CASE REPORT

A 28-year-old Congolese woman, married and the mother of two children, had a history of acute rheumatic fever (ARF) with a prolonged hospital stay at the age of 14. She had not received any subsequent medical follow-up. One month after the birth of her second child, she was admitted to the Cardiology Department at the Brazzaville University Hospital with exertional dyspnoea that had developed rapidly, followed by persistent dyspnoea (NYHA Class IV). On admission, she presented with signs of global heart failure.

The clinical examination revealed normal consciousness, good skin and mucous membrane colour, a temperature of 37°C, a weight of 62 kg, a height of 158 cm, giving a body mass index of 24.8 kg/m^2. She complained of severe physical weakness and loss of appetite. Oxygen saturation in room air was 96 per cent, the WHO performance status was 2, and urine output was 3200 ml/24 hours.

The chest was normal in appearance, the pulse was regular at 100 beats per minute, and blood pressure was 105/60 mmHg. There was tachycardia, a proto-diastolic gallop rhythm, a holosystolic murmur at the apex of intensity 3/6, radiating to the axilla, indicative of mitral regurgitation (MR), and crackles at the lung bases. The remainder of the examination revealed: spontaneous jugular vein distension, painful hepatomegaly (liver span = 17 cm), hepatojugular reflux, and oedema of the ankles and feet. There was no Homans' sign. The rest of the physical examination was normal.

Paraclinical investigations

- The anteroposterior chest X-ray (Figure 1) showed: cardiomegaly (cardiothoracic index = 0.67), with a mitral-type cardiac silhouette (protrusion of the lower right arch with a double contour, consistent with dilatation of the left atrium (LA); a straight left middle arc, an elongated left lower arc); venous redistribution at the apexes; elevation of the right diaphragmatic dome.

-The standard 12-lead electrocardiogram showed a sinus rhythm and signs of left atrial and ventricular overload.
-Two-dimensional echocardiography (Figure 2) revealed: a) an image suggestive of an aneurysm in the subvalvular portion of the mitral valve, located at the level of the posterior leaflet, with a single neck, communicating freely with the left ventricle (LV) and measuring 3.5 mm × 1.5 mm; b) thickened mitral valve leaflets; c) dilated cardiac chambers, measuring 40 mm for the left atrium (LA) and, for the LV, 63 mm in telediastole and 52.8 mm in telesystole; a Simpson's LV ejection fraction reduced to 35%. Transesophageal echocardiography (Figure 3) also revealed hypokinesis predominantly affecting the interventricular septum and pericardial detachment. On Doppler, the mitral regurgitation was grade 2 and the pulmonary arterial pressure was 30 mmHg. Echocardiography conclusion: ASAM associated with rheumatic mitral regurgitation and postpartum cardiomyopathy.

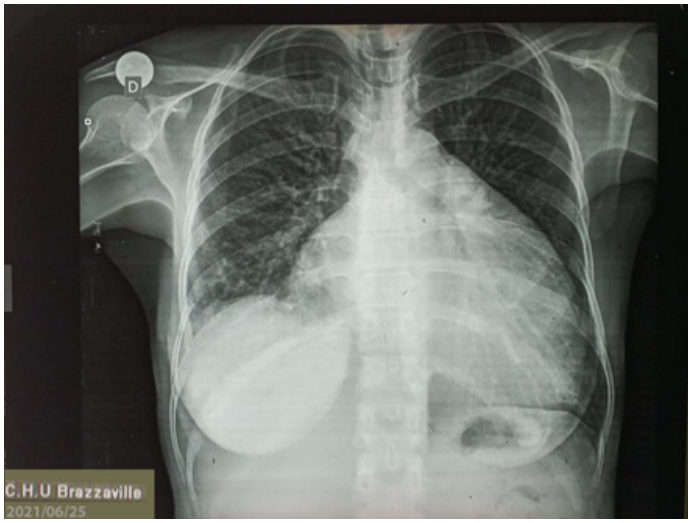

Figure 1: Anteroposterior chest X-ray showing cardiomegaly (cardiothoracic index = 0.67) and a mitral-type cardiac silhouette.

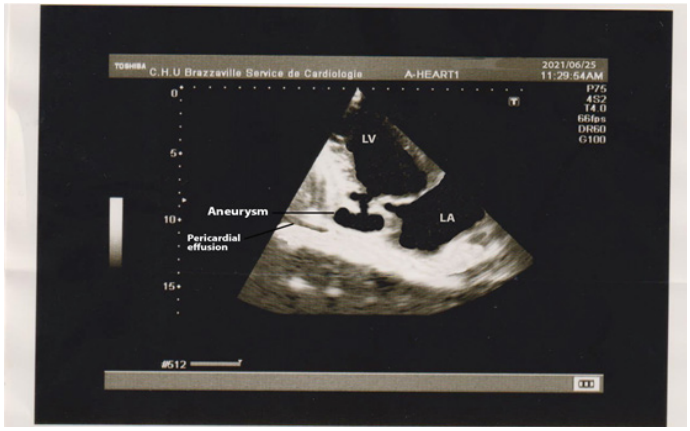

Figure 2: Two-echocardiography showing the sub mitral aneurysm (3.5 mm X 1.5 mm) opening into the left ventricle, the thickness of mitral valves in relation with rheumatic heart disease, and a little pericardial effusion.
Legend : LA = Left atrium ; LV = Left ventricle.

Treatment and clinical course

Initial treatment consisted of managing heart failure through: a low-sodium diet and fluid restriction; furosemide 40 mg administered parenterally, then orally at the same dose; ramipril 2.5 mg; dapagliflozin 10 mg; and spironolactone 25 mg. By day 15 of hospitalisation, the signs of heart failure had resolved. The patient was discharged. The one-month follow-up was satisfactory, confirming: the absence of signs of heart failure, and the persistence of a mitral regurgitation

murmur requiring regular medical follow-up. However, the patient was subsequently lost to follow-up.

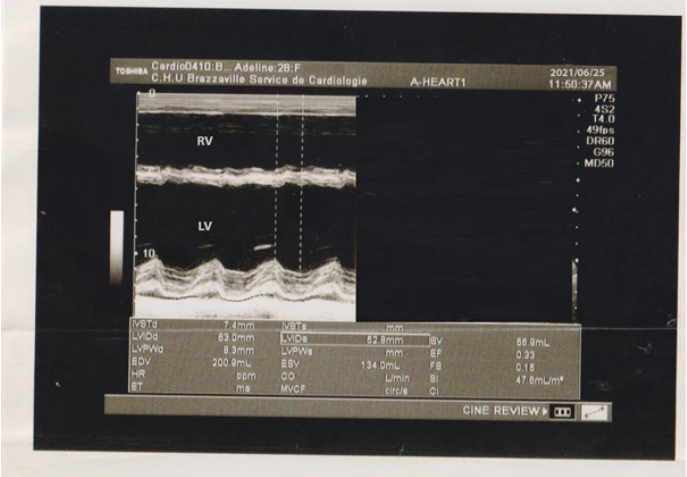

Figure 3: Échocardiography time movement showing dilated ventricles, hypokinesis of the interventricular septum and a little pericardial effusion.
Legend: LV= left ventricle ; RV = right ventricle.

-Routine laboratory tests were normal. For technical reasons, it was not possible to measure anti-Chlamydia pneumoniae antibodies, as was done in the three patients reported by Cenac et al. [24].

DISCUSSION

Epidemiology, aetiopathogenesis, anatomy

-Mitral subannular aneurysm (MSA) is a rare condition, diagnosed at any age, with no gender predominance and across all ethnic groups, although its documented incidence is higher among young African adults [16]. In the Congolese series [10], the youngest and oldest patients were 9 and 77 years old, respectively, and in Dakar, Senegal, the two children were 11 and 3 years old, respectively [4]. Our 28-year-old patient was Congolese of African origin.

-The prevalence of the condition varies from country to country. However, in a review of articles published between 1962 and 2018, Morais et al. [16] identified 150 cases of subvalvular aneurysms, 140 of which were submitral, and of these, 125 were congenital.

-It is now generally accepted that ASAM is associated with a congenital weakness of the posterior fibrous ring of the mitral valve. This may predispose individuals to the condition if they have been exposed to certain infectious, inflammatory or degenerative processes affecting the valvular endocardium [3,6,16,23], or to the presence of specific anti-Chlamydia pneumoniae antibodies, as reported by Cenac et al. in Niamey among female patients with peripartum cardiomyopathy [24]. Furthermore, the close association once observed between ASAM and young adults of African descent is no longer relevant, as this condition has been observed in India [7] and Brazil [8], where the latter authors even refute the hypothesis that ASAM is a phenomenon specific to those of African descent. In fact, it should be emphasised that Africa is a predominantly poor continent where infectious diseases are rife; amongst these, rheumatic heart disease and tuberculosis in particular continue to have the most significant negative impact on morbidity and mortality, with a possible causal link to the aetiopathogenesis of ASAM, as mentioned above [3,4,10,15,16,23,26].

-Thus, the anatomical vulnerability of the left ventricular structures at the subvalvular mitral level, linked to the specific epidemiology of the aforementioned infectious diseases, is thought to lead to chronic inflammation and progressive degeneration of the junction with the mitral-aortic annulus [8].

-In our patient, given her history of RAA and the thickened appearance of the mitral valve on transthoracic echocardiography (TTE) confirming the rheumatic aetiology, the cause of ASAM in this case is most likely rheumatic inflammation. This explains the association between rheumatic valvular disease (which in our case was moderate mitral regurgitation) and ASAM. Other authors have observed the same association [18,19]. In our case, TEE revealed a single-neck aneurysm measuring 3.5 mm × 1.5 mm, opening into the left ventricle. The aneurysm may be large [3,4,8,10,13,17-19,23,26], impairing the function of the mitral valve apparatus to the extent of causing significant to severe mitral insufficiency, or compressing the coronary arteries, thereby causing a myocardial infarction (MI) [5]. Du Toit et al. [26] classified ASAM into three types: type I, a single localised neck; type II, multiple necks; type III, involvement of the entire posterior mitral annulus.

-ASAM, particularly when extensive, may be complicated by: a) an in situ thrombus, visualised on echocardiography [27] or at autopsy, as in the case of the 3-year-old infant in Dakar [4]; b) an IDM [5]; c) severe ventricular arrhythmias [14,23,28]; d) valvular perforation, associated with a left atrial–left ventricular fistula, as reported by Harsanyi et al [29]; e) a rupture in the pericardium [15,23,30].

Clinical and Paraclinical Findings

The tell-tale clinical signs in our patient were: dyspnoea, liver pain and oedema of the lower limbs, indicative of a syndrome of global heart failure more likely to be associated with postpartum cardiomyopathy, as the mitral regurgitation was moderate (grade 2 on Doppler). This presentation of heart failure has been reported by several authors [2-4,8,10,11,13,17-19,23]. In our case, Doppler echocardiography enabled us: a) to visualise the mitral regurgitation and confirm its rheumatic aetiology, given the texture of the thickened mitral leaflets in a patient with a history of rheumatic heart disease; b) to assess the severity of the mitral regurgitation and the dilatation of the cardiac chambers. The significant LV dilatation with reduced systolic function and the minimal pericardial effusion were more consistent with postpartum cardiomyopathy at the stage of global heart failure. The TEE also enabled visualisation of the single-lumen ASAM opening into the left ventricle. TEE plays a key role in the assessment of ASAM. It has been used in several series [3-5,7,8,13,15-19,26,28-30]. Other non-invasive cardiological investigations (transoesophageal echocardiography, computed tomography, magnetic resonance imaging) also play an important role in the assessment of ASAM. However, their accessibility and availability are barriers to their widespread use in our setting.

Therapeutic and clinical course

Management is primarily medical [4,8,10,11,13,18,19,23], consisting of drug treatment for complications, foremost among which is heart failure, as in our patient, in whom this syndrome was exacerbated by the postpartum context. We successfully administered a low-sodium diet in combination with diuretics and vasodilators. This enabled the patient to be discharged. She was seen at her one-month follow-up in good haemodynamic condition, with persistent myocarditis of confirmed rheumatic aetiology, which consequently required medical monitoring alongside that of the aneurysm. However, the patient was subsequently lost to follow-up.

Surgical repair is considered the primary treatment option [22]. It was first reported by Shire and Barnard [20]. The procedure involves resection of the aneurysm, followed by the placement of a pericardial patch and repair of the mitral valve, either by valve plasty or replacement with a mechanical or biological prosthesis [6,8,10,11,13,18-20,22,23]. Large aneurysms with severe MI require this management as soon as possible to ensure a favourable post-operative prognosis [5,22]. However, the presence of shock [3] or the occurrence of a mechanical complication (perforation, parietal rupture of the LV or an aneurysm of the sinus of Valsalva) constitutes a surgical emergency [5,13,15,23,28-30].

CONCLUSION

A subannular mitral aneurysm may be congenital or secondary to an inflammatory process. It is frequently reported in people of African descent. The most common presentation is moderate or severe mitral regurgitation. This mitral regurgitation may be caused by the aneurysm, or may simply be associated with it. The case reported here is a rare association of rheumatic heart disease with moderate mitral regurgitation, accompanied by a small aneurysm, which was diagnosed during the investigation of postpartum heart failure. This review of the literature has been presented primarily to raise awareness amongst clinicians, particularly echocardiographers, of this rare cardiac condition when they are faced with young patients presenting with mitral regurgitation and heart failure.

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