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Clinical case

Intrapericardial rupture of a mitral subannular aneurysm: A case report and review of the literature

Anévrisme sous-annulaire mitral rompu dans le péricarde : cas clinique et revue de la littérature

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Abstract

Mitral subannular aneurysm is a rare heart disease that can have many different forms of clinical presentations. It was first described in young men of African descent and was later reported in other geographical areas of the world. The etiopathogenesis as per data from the literature can be congenital, acquired or idiopathic. We report the case of a 19-year-old male in whom we made the diagnosis of mitral subannular aneurysm. The evolution was fatal following a rupture of the aneurysm into the pericardium. Through this case report, we stress the importance of echocardiography in the diagnosis of this condition. In resource-limited countries, the prognosis is unfortunately often unfavorable especially the ruptured forms. Therefore, a high degree of suspicion is needed to make a prompt diagnosis and timely surgical intervention.

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Keywords: Mitral subannular aneurysm; Intrapericardial rupture

Résumé

L'anévrisme sous-annulaire mitral est une cardiopathie rare qui peut avoir des présentations cliniques multiples. Il a été initialement décrit surtout chez de jeunes patients d'origine africaine, puis secondairement signalé dans différentes zones géographiques du monde. Des données de la littérature, il ressort au plan étiopathogénique, qu'il peut être congénital, acquis ou idiopathique. Nous rapportons le cas d'un patient de 19 ans chez qui nous avons fait le diagnostic d'anévrisme sous-annulaire mitral, l'évolution a été fatale suite à une rupture intra-péricardique de l'anévrisme. À travers cette observation, nous soulignons l'importance de l'échographie cardiaque dans le diagnostic de cette affection. Dans les pays à faibles ressources, le pronostic est malheureusement souvent défavorable dans ses formes rompues.

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Mots clés : L'anévrisme sous-annulaire mitral ; Rupture intra-péricardiale

1. Introduction

Mitral subannular aneurysm was first described in Nigeria [1]. This disease was later reported in other countries in sub-Saharan Africa and also in other parts of the world [2–4]. Its clinical presentation varies and its diagnosis has been facilitated by transthoracic and transesophageal echocardiography, multidetector scanner and more recently by magnetic resonance imaging [2,4,5]. Surgery is the cornerstone of treatment.

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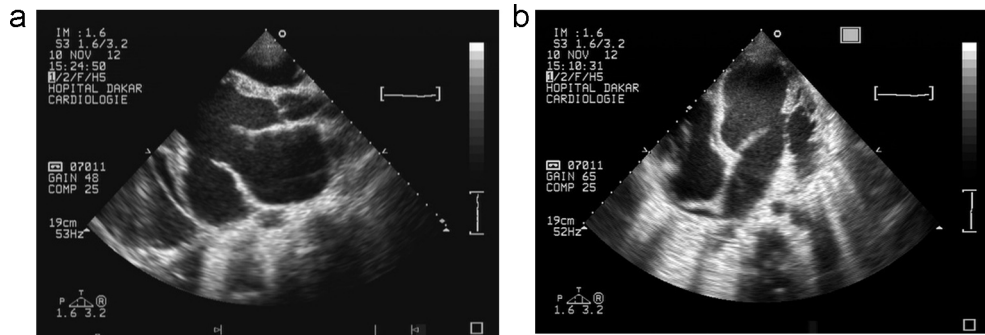


Fig. 1. a: 2D transthoracic echocardiography longitudinal long axis view showing mitral subannular aneurysm in diastole; b: 2D transthoracic echocardiography apical 4-chamber view showing aneurysm.

However, the prognosis is often not favorable even with surgery [2,4,6].

2. Case presentation

We report the case of a 19-year-old male who presented with a two-month history of chest pain with stage III dyspnea of the New York Heart Association classification. There was no significant past medical history. He was hemodynamically stable with an oxygen saturation of 99% on room air, heart rate of 118 beats per minute and a blood pressure of 90/50 mmHg. The apical impulse was deviated from the mid-clavicular line. Cardiac auscultation found regular heart sounds with an apical systolic murmur with an intensity of 3/6 radiating to the left axilla. The peripheral pulses were well palpated, and were of good volume and symmetrical. The rest of the clinical examination was normal apart from cryptic hypertrophied tonsils.

Echocardiography (Figs. 1 and 2) found a significant dilatation of the left heart chambers with a left atrial antero-posterior diameter of 49 mm, left ventricular diameter of 68 mm and 45 mm in telediastole and telesystole respectively. There was a preserved systolic left ventricular function. We noted a severe mitral insufficiency (regurgitant orifice area of 0.47 cm² and a regurgitant volume of 42 mL/cycle) by dilatation of the mitral annulus (53 mm) and faulty coaptation of the two mitral leaflets.

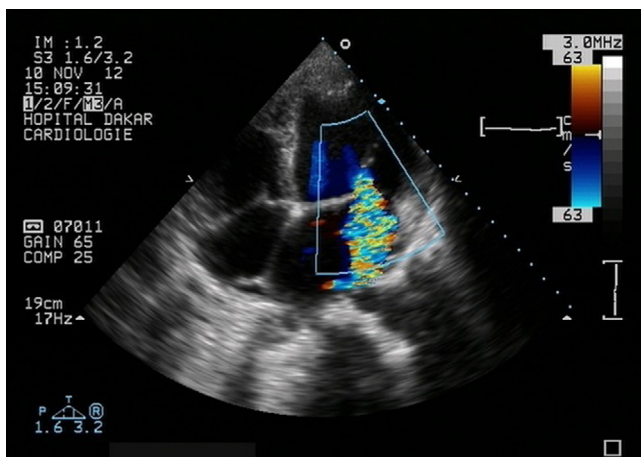


Fig. 2. Colour Doppler transthoracic echocardiography 4-chamber view in systole showing mitral subannular aneurysm and a severe mitral insufficiency.

The mitral leaflets were moderately thickened with the subvalvular apparatus slightly modified. We noted the presence of a voluminous subannular aneurysm whose antero-posterior diameter was 45 mm, and with an area of 32 cm² containing a spontaneous contrast of grade 2+, but without intra-aneurysmal thrombus. The overall kinetics of the walls of the left ventricle was homogeneous and correct. Moderate tricuspid regurgitation was noted with a severe pulmonary hypertension estimated at 62 mmHg, for a right atrial pressure of 10 mmHg.

Laboratory tests found a hypochromic microcytic anemia with a hemoglobin level of 9.3 g/dL and an anti-streptolysin O level of 1600 IU/L, and raised inflammatory markers with a C-reactive protein of 48 mg/L and a fibrinogenemia of 5.72 g/L. HBsAg test was negative. The tuberculin skin test was negative as well as the search for acid-fast *Bacilli* in sputum. Serology for HIV-1 and HIV-2 were negative. Blood cultures however could not be obtained. The medical treatment given was in the form of furosemide 80 mg daily, spironolactone 50 mg daily, captopril 50 mg daily associated with treatment with a low molecular weight heparin.

The evolution was marked by the appearance of mainly persistent nocturnal chest pains associated with fever and tachycardia with examination findings of muffled heart sounds and signs of bilateral pleural effusion. Blood count found a further decrease in hemoglobin level to 8.3 g/dL associated with a thrombocytosis of 490,000/mm³. Follow-up echocardiography (Fig. 3a and b) demonstrated the occurrence of a pericardial effusion predominant facing the lateral wall of the left ventricle measuring 15 mm without signs of compression or haemodynamic compromise. The indication for surgical treatment of the aneurysm was made but the patient died before surgery.

3. Discussion

Mitral subannular aneurysm was described for the first time in Nigeria [1] in 1962. This condition was later described in many African series and in the world [2–5,7]. It is a disease whose cause remains unclear, with rare and varied etiologies. Because of its predilection in black people, genetic origin has often been suggested [2–4]. Combination with tuberculosis, rheumatic fever, infection with *Chlamydia pneumoniae* or Takayasu's arteritis have also been reported [2,6–8]. This suggests the role of infection and inflammation in the pathogenesis

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