CASE REPORT

A rare case of an isolated aneurysmal supravalvar mitral ring in a young Nigerian adult: a cor-triatriatum mimic

Oyewole A Kushimo1,*,†, Ogochukwu Sokunbi2, Michael Akinkunmi1 and Yeside O Akinbolagbe2

1Cardiology Unit, Department of Medicine, Lagos University Teaching Hospital, PMB 12003, Lagos, Nigeria, 2Department of Paediatrics, Lagos University Teaching Hospital, PMB 12003, Lagos, Nigeria
*Correspondence address. Department of Medicine, Lagos University Teaching Hospital, PMB 12003, Lagos, Nigeria. Tel: +2348054580119; E-mail: wolekushimo@gmail.com

Abstract
Supravalvar mitral ring is a rare congenital abnormality characterized by a ridge of connective tissue located above the mitral valve. It is a cause of congenital mitral stenosis typically presenting in childhood and usually associated with other cardiac abnormalities. We report the rare case of a 24-year-old male presenting with an isolated aneurysmal supravalvar mitral ring. He presented at the emergency room with a 2-week history of worsening heart failure symptoms and antecedent effort intolerance of 4 years duration. He was referred from a primary care facility with an echo diagnosis of cor-triatriatum. Echocardiography done at our centre revealed an isolated aneurysmal supravalvar mitral ring of the intramitral variant. This report highlights the unusual isolated presentation of a supravalvar mitral ring in a young adult and the need to carefully differentiate it from cor-triatriatum, a possible close mimic.

INTRODUCTION
Supravalvar mitral ring, also called a supramitral ring, is a rare condition with an incidence of 1.5% amongst autopsy cases of congenital heart abnormalities [1]. It is characterized by a ridge of connective tissue located on the atrial side of the mitral valve. It may be attached at or above the mitral annulus (supramitral variant) or may be located within the mitral tunnel and adherent to the mitral valve leaflets (intramitral variant) [2]. It is a cause of congenital mitral stenosis accounting for about 12% of cases [3]. It may rarely be isolated but more commonly associated with other congenital abnormalities such as the shone complex [4]. We present an unusual case of a young adult presenting with an isolated aneurysmal supravalvar mitral ring who was referred as a case of cor-triatriatum.

CASE PRESENTATION
A 24-year-old male Nigerian presented to the accident and emergency of our institution with a 2-week history of worsening breathlessness, orthopnoea, paroxysmal nocturnal dyspnoea, cough and bilateral leg swelling. He presented in New York Association (NYHA) class IV. He had an antecedent history of effort intolerance of 4 years duration. He was referred from a primary care facility as a case of cor-triatriatum based on a transthoracic echocardiogram report.

On physical examination, he was dyspnoeic, not pale, afebrile, with bilateral pitting pedal oedema up to his thighs. Pulse rate was 128 bpm, BP 150/110 mmHg, with an elevated JVP, displaced apex beat and a palpable thrill over the apex. On auscultation, heart sounds S1, S2 and S3 were heard with a pan
Figure 1: 2D Apical four chamber transthoracic echo image showing an aneurysmal supravalvar mitral ring attached to the mitral valve leaflets (blue arrow).

Figure 2: 2D Parasternal long axis transthoracic echo image showing the supravalvar mitral ring (blue arrow).

systolic murmur at the mitral and tricuspid areas. Respiratory rate was 32 cycles per minute, with bi-basal crackles and tender hepatomegaly.

Investigations done include an electrocardiogram that showed sinus tachycardia with low voltage complexes in limb leads. Chest X-ray showed cardiomegaly with perihilar shadows. Full blood count was normal. Hypokalaemia (2.6 mmol/L) was noted. Transthoracic echocardiography revealed an aneurysmal supravalvar mitral ring (‘intramitral variant’) adherent to structurally normal mitral valve leaflets (Figs 1 and 2) with moderate to severe mitral regurgitation (Fig. 3), mild left ventricular (LV) inflow obstruction (mean transmitral gradient 6.45 mmHg), multi-chamber dilatation, preserved LV ejection fraction (54%) and a pulmonary artery systolic pressure gradient of 35 mmHg (tricuspid regurgitant velocity 2.64 m/s). Other congenital anomalies such as septal defects, patent ductus arteriosus and aortic coarctation were not observed.

A diagnosis of biventricular heart failure secondary to congenital valvular heart disease (supravalvar mitral ring) was made. He was placed on heart failure medications (Lisinopril, Carvedilol, Torsemide and Spironolactone) and oral potassium supplementation. He made remarkable improvement and was discharged after 12 days on admission in NYHA class II to be followed up in the cardiology clinic. He has been counselled on the need for open heart surgical correction of his cardiac condition.

Figure 3: Colour Doppler transthoracic echo image in parasternal long axis view showing severe mitral regurgitation (black arrow).

DISCUSSION

This is an unusual case of a supravalvar mitral ring in view of the late age of presentation, the isolated finding and its aneurysmal character. Most cases become symptomatic in childhood with a reported mean age at diagnosis of 36 months [2]. The index patient developed significant symptoms in his third decade and this late presentation might explain the aneurysmal expansion of the ring. We found only one similar case of adult presentation reported in the literature concerning an isolated supravalvar mitral ring in a 23-year-old Turkish patient [5].

Supravalvar mitral rings rarely occur in isolation [3, 6]. The few isolated cases reported are usually of the supramitral variant. The intramitral variant usually occurs as a component of the Shone complex that is a constellation of congenital left-sided obstructive lesions namely a supravalvar mitral ring, parachute mitral valve, subvalvular aortic stenosis and aortic coarctation [4]. The index case is an intramitral variant. Hence, our finding of an isolated supravalvar mitral ring of the intramitral variant presents an unusual scenario, which to the best of our knowledge is yet to be reported in the literature. Intramitral rings tend to be attached to the mitral leaflets that can cause valvular dysfunction as noted in our patient.

This patient was referred to our centre with a prior diagnosis of cor-triatriatum, which is a congenital abnormality characterized by a fibrous membrane that divides the left atrium into two compartments [7]. The attachment of this membrane is located superior to the left atrial appendage as opposed to a supravalvar mitral ring that attaches inferiorly. The aneurysmal nature of the mitral ring in the index patient makes it more likely to be misdiagnosed as cor-triatriatum as it gives the impression of a double chamber left atrium. Supravalvar mitral rings can be detected with transthoracic 2D echo in about 73% of cases [8]. However, there are reports of improved detection of supravalvar mitral rings using 3D as compared with 2D echocardiography [9]. The gold standard for pre-operative assessment of mitral valve anatomy is transesophageal 3D echocardiography [10]. The definitive treatment for supravalvar mitral rings is surgical ring resection and possible mitral valve replacement when indicated [2].

In conclusion, supravalvar mitral rings can rarely occur in isolation and present in adulthood. Patients with this abnormality require a thorough evaluation to differentiate them from cor-triatriatum, a possible close mimic.
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CONFLICTS OF INTEREST
None declared.

ETHICAL APPROVAL
None required.

CONSENT
Informed consent was obtained from the patient.

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Oyewole A Kushimo.

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