Case Report

Hammock mitral valve: A rare cause of congenital mitral regurgitation — A case report

Gopalan Nair Rajesh, Kalathingathodika Sajeer, Anishkumar Nair, Chakanalil Govindan Sajeev, Mangalath Narayanan Krishnan

Additional Professor, Department of Cardiology, Government Medical College, Kozhikode, Kerala 673008, India
Senior Resident, Department of Cardiology, Government Medical College, Kozhikode, Kerala, India
Senior Resident, Department of Cardiology, Government Medical College, Kozhikode, Kerala, India
Professor, Department of Cardiology, Government Medical College, Kozhikode, Kerala, India
Professor and Head, Department of Cardiology, Government Medical College, Kozhikode, Kerala, India

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A B S T R A C T
Hammock valve, also known as anomalous mitral arcade is a rare mechanism for congenital mitral insufficiency. We report a case of a two-week-old neonate who presented with features of heart failure and an apical systolic murmur. Echocardiogram showed severe mitral regurgitation and abnormal mitral valve with direct attachment of mitral leaflets to papillary muscle without intervening chordae tendinae, typical of hammock valve. Heart failure was controlled with ionotropes and diuretics. The literature on the hammock mitral valve is reviewed.

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1. Introduction
Congenital mitral regurgitation due to hammock mitral valve is very rare, and to date, only 13 cases of this anomaly have been reported, most of which were detected before 3 years of age. It is a congenital malformation of the mitral tensor apparatus characterized by enlarged papillary muscles connected to mitral leaflets by a typical fibrous tissue bridge which creates a fibrous continuity between valvular and subvalvular apparatus.

2. Case report
A 2-week-old newborn was referred to our institution for evaluation of respiratory distress. Examination revealed substernal and intercostal retraction, cardiomegaly with left ventricular type of apex beat and prominent systolic murmur at apex. Oxygen saturation was 94% on room air and there was mild hepatomegaly. Chest X-ray showed pulmonary congestion and electrocardiogram showed biventricular dominance with right axis deviation. Transthoracic echo evaluation
showed abnormal mitral valve with direct attachment of mitral leaflets to papillary muscle without intervening chordae tendineae. Colour Doppler echo evaluation showed severe mitral regurgitation (Fig. 1A–C Movie 1, 2). Heart failure was managed with inotropes and diuretics.

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3. Discussion

Anomalous mitral arcade or hammock mitral valve is a rare congenital malformation of the mitral valve and of its tensor apparatus that was first described in 1967 by Layman Edwards. The term hammock mitral valve was first coined by in 1976 by Carpentier, because of the unique appearance of mitral valve apparatus from its left atrial aspect as viewed during cardiac surgery. This anomaly is characterized by enlarged and elongated papillary muscles connected to each other and to the free edge of the anterior mitral leaflet by a bridge of fibrous tissue. The absence of chordae tendineae between the fibrous bridge and anterior leaflet results in a fibrous continuity that interferes with the valvular motion and prevents normal apposition of leaflets. It is suggested that this malformation is the result of an arrest in the developmental stage before attenuation and lengthening of the collagenized chordae tendineae. The features of anomalous mitral valve arcade are (1) A near normal mitral valve orifice; (2) short, thick, and poorly differentiated chordae tendineae with direct attachment of the papillary muscles to the anterior mitral leaflet; (3) narrow interchordal spaces and (4) relatively well developed chordae attached to the posterior papillary muscle. The papillary muscles which are large and elongated are connected to each other and to the free edge of the anterior mitral leaflet by a bridge of fibrous tissue. The natural history of the malformation is characterized by progressive worsening of valvular regurgitation, stenosis or both.

Pulmonary vascular resistance changes throughout the gestation and after birth. In neonate, rise in pulmonary venous pressure is preceded by greater increase in pulmonary arterial pressure. This results in increase of calculated resistance across pulmonary vascular bed. Thus post capillary pulmonary hypertension as consequence of severe mitral regurgitation due to hammock mitral valve may increase pulmonary vascular resistance. In cases of mitral regurgitation presenting with heart failure, changes in left atrial pressure which occurs independent of alteration in pulmonary vascular resistance, should also be considered. The majority of the reported cases are in the paediatric age group and there are only a few reports of anomalous mitral arcade in adults. A patient who reaches adulthood usually undergoes mitral valve repair or replacement. In Hammock mitral valve, repair can be performed using annuloplasty, commissurotomy, modified Gerbode–Hetzer plication plasty, modified Paneth–Hetzer posterior annulus shortening techniques and papillary muscle splitting according to the presenting morphology. The clinical course varies depending on the severity of mitral stenosis and regurgitation.

Conflicts of interest

All authors have none to declare.

References