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Case Report

Wong's anomaly – A rare variant of cor triatriatum

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ABSTRACT

Cor triatriatum sinistrum is an extremely rare congenital heart disease. It is even more uncommon in adults, and clinically significant mitral valve lesion complicating cor triatriatum is distinctly rare. Wong et al reported for the first time the rare combination of cor triatriatum sinister associated with severe mitral regurgitation and abnormal tensor apparatus of the mitral valve. We report a similar case and used the term Wong's anomaly for the syndrome, having membranous type of cor triatriatum sinistrum, severe mitral regurgitation and hypoplasia of the papillary muscles and short chordae. Color Doppler Echocardiography showed peculiar 'helmet sign' of mitral regurgitation, wherein the mitral regurgitation color jet fills the distal atrial chamber and abruptly ends in a horizontal plane as it is halted by the intra-atrial membrane.

1. Introduction

Cor triatriatum is a rare congenital anomaly in which the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into 2 compartments by a fold of tissue, a membrane, or a fibromuscular band. It is even more uncommon in adults, and quite exceptional when associated with mitral valve disease.

Herein, we report a case of an adult male who presented with the rare combination of cor triatriatum associated with severe mitral regurgitation (MR) and abnormal tensor apparatus of the mitral valve, who remained relatively asymptomatic till the fourth decade of life. We have used the term Wong's anomaly for the syndrome as this rare constellation of abnormalities which was first reported by Wong et al[1] in 1989.

2. Case report

A 40 year old male patient presented with dyspnea on exertion since childhood, with recent deterioration of the symptom. On examination, vital signs were stable. Pulse was irregularly irregular at a rate of 90 per minute. Precordial examination revealed mild cardiomegaly. First heart sound was varying in intensity, second sound was close split with loud P2. A mid diastolic murmur and a grade 3/6 pansystolic murmur were audible over the apex. ECG showed atrial fibrillation with

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controlled ventricular rate. Chest X ray showed mild cardiomegaly with biatrial enlargement and prominent upper lobe vessels.

Transthoracic Echocardiography showed dilated left atrium (LA) with a thin membrane stretching across the mid part, dividing LA into a proximal and distal chamber (Figs. 1a and 2a, Video1 & 4). A fenestration of diameter 7 mm was noted in the postero inferior part of the membrane with a mean diastolic gradient of 20 mm of Hg and peak gradient of 32 mm of Hg (Video 2). Pulmonary veins were seen draining into the proximal chamber. There was associated severe mitral regurgitation (Fig. 1b, Video 3). The tensor apparatus of the mitral valve appeared abnormal with hypoplastic papillary muscles and shortened and thick chordae. LV function appeared good. The findings were confirmed by a transesophageal echocardiogram (Fig. 2b, Video 5).

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Patient was referred to cardiothoracic surgeon and underwent successful exicision of the membrane with mitral valve repair. Intraoperatively, all pulmonary veins were seen draining into the common chamber which was dilated, with a membrane separating it from the true left atrium. Mitral annulus was grossly dilated with thickened anterior and posterior mitral leaflets. Medial papillary muscle appeared normal; lateral papillary muscle was thickened and hypoplastic. Excision of the cor triatriatum membrane and mitral valve repair with 31 Duran Ancore Ring (Medtronic), reinforced by central Alfieri repair was done, with good results.

3. Discussion

First described in 1868,[2] Cor triatriatum sinistrum is an extremely rare congenital heart disease. The incidence of cor triatriatum has been variously reported as 0.1–0.4%. It is even more uncommon in adults, and quite exceptional when associated with mitral valve disease.

Although mitral valve abnormalities have been reported in postmortem pathologic studies, clinically significant mitral valve lesion complicating cor triatriatum is distinctly rare.[3–6] Occasionally mitral regurgitation has been found at surgery for cortriatriatum,[4,5] although the mechanism was unclear. In the cases of cor triatriatum reported by Marin-Garcia and colleagues,[7] associated mitral valve anomalies were frequent. By contrast, in the series from Van Pragh and Corsini,[3] there was only one case. Papillary muscle hypoplasia with short chordae tendineae limit mobility of the mitral valve leaflets which become fixed along the wall of the left ventricle resulting in incomplete coaptation and MR.[8]

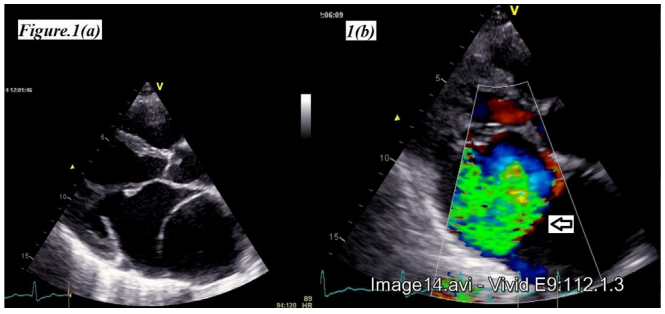

Fig. 1 – (a) Parasternal long axis view of Echocardiogram showing the membrane dividing left atrium into proximal accessory chamber and distal true left atrial chamber. (b). Characteristic 'helmet' shaped mitral regurgitation color doppler jet filling the distal chamber (black arrow).

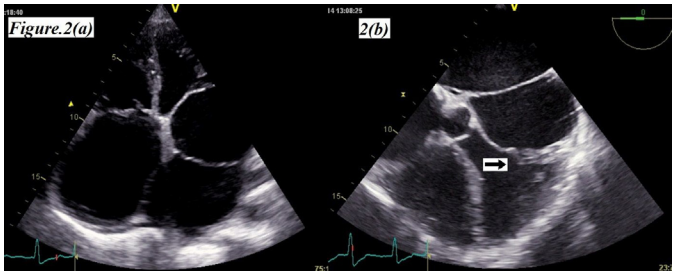

Fig. 2 – (a) Apical 4 chamber view of Transthoracic Echocardiogram (b) Trans Esophageal Echocardiogram showing the true and accessory atrial chambers with hypoplastic papillary muscle and short chordae (black arrow).

Wong et al[1] reported for the first time in 1989 a 49 year old male patient with the rare combination of cor triatriatum sinister associated with severe mitral regurgitation and abnormal tensor apparatus of the mitral valve with hypoplastic papillary muscle. The case reported by us had a similar presentation with membranous type of cor triatriatum sinistrum, severe mitral regurgitation and hypoplasia of the papillary muscles and short chordae. Management differs from isolated cortriatriatum in that in addition to surgical resection of the accessory membrane, patient requires mitral valve repair or replacement for correction of mitral regurgitation.

In this case reported by us, the transthoracic and transesophageal echocardiography clearly demonstrates the membrane that partitions the left atrium as well as the underdeveloped papillary muscles and short chordae tendinae. We also describe the peculiar 'helmet sign' of mitral regurgitation in the color doppler echocardiography, wherein the mitral regurgitation color jet fills the distal atrial chamber and abruptly ends in a horizontal plane as it is halted by the intra-atrial membrane, giving a characteristic hemispheric or 'helmet' shape for the MR jet (Fig. 1b).

The clinical significance of this entity lies in the fact that Cor triatriatum sinistrum is often misdiagnosed as other common cardiac conditions such as mitral stenosis. In the currently reported case by us, there was associated abnormal tensor apparatus of the mitral valve causing severe mitral regurgitation with a characteristic helmet shape of the regurgitant jet in the color doppler echo. There are only very few reports of such a combination in literature.

Conflicts of interest

The authors have none to declare.

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