Case Report

COR TRIATRIATUM IN A 60-YEAR-OLD WOMAN BECAME SYMPTOMATIC AFTER PAPILLARY CORDAE RUPTURE

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ABSTRACT

We describe a case of cor triatriatum in a 60-year-old woman. The diagnosis was initially made by transesophageal echocardiography after the presentation of the patient with pulmonary edema caused by papillary cordae rupture. The patient made an uneventful recovery after membrane resection and mitral valve replacement.

Key Words : Cor triatriatum, Papillary cordae rupture, Mitral valve replacement

INTRODUCTION

Cor triatriatum (CT) is an uncommon congenital cardiac anomaly in which the left or right atriums are divided into two chambers. Two general types were described as Cor Triatriatum Sinister and Dexter according to affected atrium location. The common type is " Cor Triatriatum Sinister ". The pulmonary veins drain into the accessory chamber that communicates through the accessory membrane orifice to true left atrium with left atrial appendage (1, 2). Three types of cor triatriatum were described as diaphragmatic, hourglass and tubular (2). Thilenius offered another complicated classification but this classification is not commonly used (3). It is rarely seen in adulthood and concomitant cardiac abnormalities are previously reported (4 - 7). CT rarely remains asymptomatic until adulthood. It may also mask the symptoms of concomitant cardiac pathologies such as mitral insufficiency. Here, we present an adult CT which remained asymptomatic till the rupture of the papillary cordae.

CASE REPORT

A 60-year-old patient had been admitted to another hospital because of acute pulmonary edema, in a deteriorating condition. The chest X-ray shows pulmonary congestion and biventricular hypertrophy. The electrocardiogram shows right axis deviation and biventricular hypertrophy. Transesophageal echocardiography showed cor triatriatum with a wide opening and severe mitral regurgitation due to the papillary cordae rupture (Figs. 1, 2.). The patient was entubated and treated medically for 6 days. The medical history of the patient revealed a 6 year period of exertional dyspnea and fatigue but no medical record of the previous examinations were

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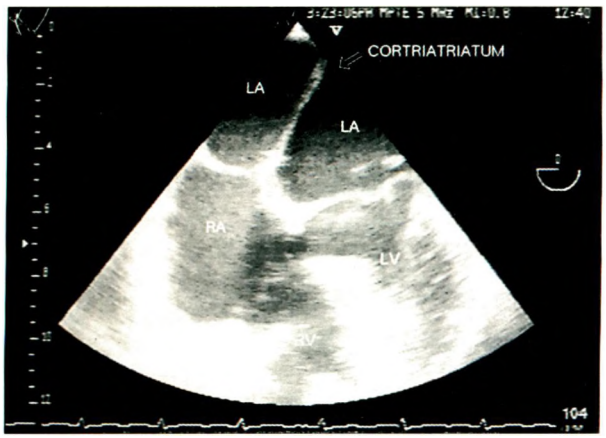

Fig.1: Two chambers and the membrane in parasternal long axis view.

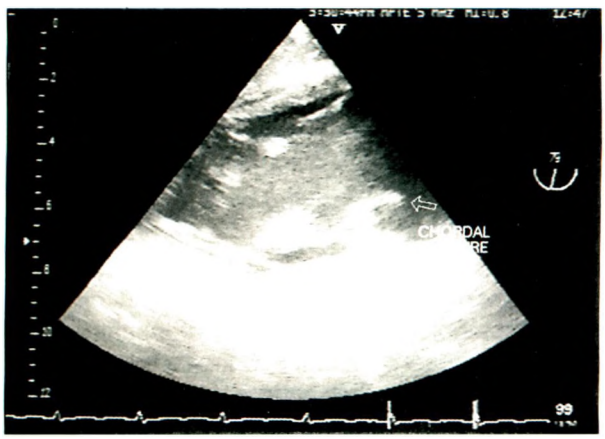

Fig.2: Papillary cordae rupture in apical four chamber view.

available. The patient was transferred to our hospital for cardiac catheterization and coronary angiography which showed severe mitral insufficiency with a 40 mm Hg pulmonary artery pressure and normal coronary artery anatomy. The patient was scheduled for early surgery.

The usual bicaval cannulation for cardipulmonary bypass was carried out with blood cardioplegia and 28°C hypothermia. The right atrium and interatrial septum was opened. The inspection showed a well formed accessory chamber and membrane with a 3x3 cm opening (diaphragmatic type) to the left atrium. The upper chamber receives the pulmonary veins.The mitral valve had a severe myxamatous structure and dilated annulus with a complete rupture of the posterior papillary muscle. The membrane was resected and mitral valve replacement with bileaflet mechanical prosthesis (Carbomedic No.27, US) was performed. The patient made an uneventful recovery and was discharged on the 6th postoperative day.

DISCUSSION

CT is a rare anomaly in adults mostly becoming symptomatic after the exceeding flow through the orifice of the membrane or hemodynamic changes due to the concomitant cardiac anomaly which is also an extremely rare condition. The exception of survival until adulthood as presented in this case was due to a wide opening differs from the usual knowledge of literature inspite of concomitant mitral valve pathology. The development of investigative procedures such as transesophageal echocardiography increased the accuracy of the diagnosis. Elective surgery is the only option for the treatment of CT in patients presenting symptoms of pulmonary venous obstruction.

Cor triatriatum in adulthood has been generally diagnosed late in the 4th and 5th decades. The most concomitant abnormalities are mitral insufficiency, secundum atrial septal defect and the unroofed coronary sinus with left persistent superior vena cava (4, 5).

The severity and the onset of symptoms in CT depends on the size of the opening between the accessory chamber and the true left atrium. When the opening is small CT manifests as an obstructive lesion with severe pulmonary edema and pulmonary hypertension that requires early surgical intervention. Only a few patients in whom the opening is wide enough for survival into adulthood and surgery may be delayed. The diagnosis is made incidentally in asymptomatic patients and mostly with transesophageal echocardiography in symptomatic patients (4, 6). In our patient the opening was wide enough for survival into the adulthood. The patient had a history of mild exertional dyspnea and limited physical activity probably due to myxomatous mitral valve disease for six years and compansated with a wide opening of CT. The membrane itself probably restricted the regurgitant flow to the pulmonary vasculature till the incidental papillary cordea rupture resulted in pulmonary edema. The orifice of the membrane

was large enough to avoid pulmonary hypertension for the excess pulmonary flow due to mitral regurgitation.

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