Letter to Editor

Cor Triatrium Presenting as Mitral Stenosis and Severe Anemia

Dear Editor,

Cor triatriatum is a rare congenital cardiac malformation. It has an estimated incidence of 0.1% of all the congenital heart diseases.[1] Cor triatriatum means triatrial heart, i.e., a heart with three atria. In cor triatriatum, the atrium is divided into two parts by a fold of tissue, a membrane, or a fibromuscular band.[2] In pediatric population, this anomaly may be associated with major congenital cardiac lesions, while in adults cor triatriatum is frequently an isolated finding.

We report the case of a 28-year-old man complaining of breathlessness for past 6 months which was insidious in onset, slowly progressive (NYHA Grade III dyspnea), aggravated on exertion, and relieved at rest. Also, there was history of cough for past 15 days which was dry, intermittent, aggravated on exertion, and relieved on taking rest. The patient also had severe dimorphic anemia. Rest of his medical history was unremarkable.

On examination, pulse rate and blood pressure were within normal limits, and respiratory rate was 26 breaths/min. Severe pallor was present. There was a mid-diastolic murmur heard after the opening snap at the apex. There was loud P_2. Infra-axillary pleural rub was present on the left side. Hematological evaluation revealed Hb 4.4 gm%, and peripheral blood smear showed predominantly microcytic hypochromic RBCs with few macrocytes. Chest X-ray showed straightening of left upper border of heart, prominent main pulmonary arteries, dilatation of upper lobe pulmonary veins, and Kerley B lines in lower and mid-lung fields. 2D echocardiography clearly showed the membrane dividing the left atria into two chambers [Figure 1]. Transesophageal echocardiography (TEE) revealed a membrane in left atrium (LA) just above the LA appendage on one side and attached to interatrial septum (IAS) on the other side, with gradient of maximum 11 and mean 7 mm of Hg across the membrane. Minimal mitral regurgitation and mild tricuspid regurgitation of Grade II/IV and mild pulmonary hypertension were also present. Right ventricular systolic pressure (RVSP) was 44 mm of Hg. Left ventricular ejection fraction (LVEF) was 60%. All cardiac valves were normal, IAS was intact and no Atrial septal defect (ASD) was seen. These features were suggestive of cor triatrium [Figure 2]. The patient became asymptomatic after correction of anemia. Further, patient has been advised surgical correction of cor triatrium.

First reported in 1868, cor triatriatum, a heart with three atria (triatrial heart), is a congenital anomaly in which the LA (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into two parts by a fold of tissue, a membrane, or a fibromuscular band.[1-3] It may be entirely intact (imperforate) or contain one or more openings (fenestrations) ranging from small, restrictive type to large and widely open. In pediatric population, this anomaly may be associated with major congenital cardiac lesions. In adults, cor triatriatum is frequently an isolated finding.[4] The incidence of cor triatriatum is 0.1–0.4%.[1,3]

Cor triatriatum sinistrum can be misdiagnosed as mitral stenosis (as in our patient).[5] In adults, cor triatriatum sinistrum can be asymptomatic. Clinical manifestations depend upon the size of the opening in the septum and the presence of associated congenital cardiac defects.[6] In adults, clinical manifestations are often delayed due to the presence of a large opening. Symptoms include exertional dyspnea, effort intolerance, and easy fatigability, hemoptysis, orthopnea, palpitations, and signs of systemic

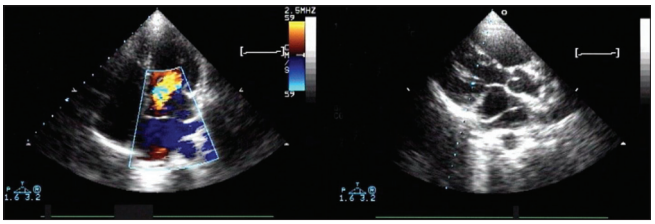

Figure 1: 2D echocardiography clearly shows the membrane dividing the left atria into two parts

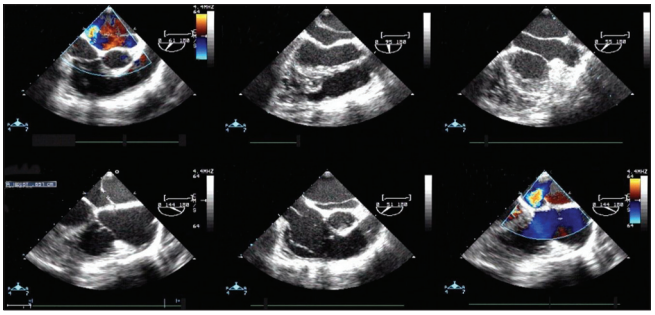

Figure 2: These features in all these images of TEE are suggestive of cor triatrium

embolism/thromboembolism (especially in those with atrial fibrillation and/or restrictive orifice).[5] Findings on examination can be murmur, pulmonary congestion, tachypnea, respiratory distress, pulmonary rales, pleural effusion, hypoxemia, pulmonary hypertension, loud P_2, right ventricular failure, elevated jugular venous pressure, right ventricular heave, tricuspid regurgitation, systolic murmur along sternal border with respiratory variations in intensity, right upper quadrant abdominal tenderness due to liver congestion, hepatomegaly, jaundice, ascites, peripheral edema, sinus tachycardia or rapid irregular heart rate, frequent premature atrial complexes, and findings related to associated cyanotic or acyanotic congenital cardiac defects.[5,6] Chest radiograph can show cardiomegaly, pulmonary congestion, prominent pulmonary arteries, or pleural effusion.[5] Echocardiography is the most commonly used imaging technique for the diagnosis of cor triatriatum,[2,3,5,6] although TEE is frequently needed to precisely define the anatomy of the membrane, its relation to other structures, and the pulmonary venous drainage pattern.[2,3,5,6]

Medical care for patients with cor triatriatum includes stabilizing hemodynamics and controlling ventricular rate in patients with atrial fibrillation. Surgical resection of the accessory membrane has been successful in symptomatic patients with cor triatriatum.[1,4] Transition from asymptomatic to symptomatic in the adult occurs mainly because of fibrosis and calcification of the orifice in the accessory membrane or development of mitral regurgitation and/or atrial fibrillation. Severe anemia resulted in cardiac overload in our patient. It was a precipitating factor for symptoms related to cor triatrium in adulthood in this case.

Cor triatrium is easily surgically correctable when hemodynamically significant. TEE is a minimally invasive and highly sensitive diagnostic tool for confirming the diagnosis of cor triatrium.[2,3,5,6]

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