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Single Atrium in 42 year Old Male

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Abstract: Single atrium (SA) is a rare congenital anomaly characterized by absence or virtual absence of atrial septum, vestigial remnant of which occasionally remain. So, here we are reporting a case of single atrium.

Keywords: CHD, ASD, Single atrium

INTRODUCTION

Patient with congenital heart disease may present in adulthood with mild unrepaired lesion, undetected defects (often during pregnancy) or residua or sequelae from surgical repair. As patients with congenital heart disease grow older acquired heart or general health problems impose on the underlying cardiac anomaly. However treatment of congenital heart disease can rarely be considered as being curative and about half of adults with congenital heart disease face the prospect of further surgery or non-surgical intervention, arrhythmia, heart failure, and –if managed inappropriately- premature death. So we are reporting a case of Single atrium in 42yrs old man with complaints of dyspnoea and palpitation.

Complete absence of the atrial septum is rare and is considered to be the least common variety of atrial septal defect [1, 2]. In the literature there would appear to be some nosologic confusion in the use of terms such as cor triloculare-biventriculare, common atrium, or single atrium.

Levy and associates [3] recommended that the term single atrium should be used to denote the condition characterized by: a) complete absence of the atrial septum, b) absence of the malformation of AV valve and c) absence of the Intraventricular (IV) communication. They suggested that the term common atrium (CA) should be used to denote the condition of complete absence of atrial septum, accompanied by malformation of AV valves with or without I.V. communication.

CASE REPORT

A 42 yrs old man, resident of a village in Bihar was presented with gradual onset of Exertional dyspnoea, palpitations for the last 10 yrs. These

symptoms have aggrevated past 1 month, associated with swelling of feet. On clinical examination, patient was dyspnoeic (saturation of 52% with room air), Grade iii clubbing of all 4 limbs, Bilateral pitting pedal edema, Engorged neck veins were noted. On Cardiac Examination, visible pulsations were present over left lower parasternal, Epigastric region. Right ventricular type of apical impulse in 5th intercostal space, lateral to mild clavicular line, left parasternal pulsations, palpable P2 are noted. On Auscultation, p2 was loud. A short systolic murmur in Tricuspid area, Ejection systolic in pulmonary area heard. Abdominal Examination showed tender Hepatomegaly. A provisional Clinical diagnosis of COPD with Cor-pulmonale is made. All blood investigations were normal. Chest x-ray showed gross cardiomegaly with left atrial and right atrial enlargement. The ECG shows right axis deviation with Bundle branch block and Biventricular right Echocardiogram revealed CHD with Hypertrophy. single atrium (absence of atrial septum), with pulmonary hypertension with intact interventricular septum. Patient was referred to cardiac centre for cardiac catheterisation.

DISCUSSION

Ellis et al. [1] consider complete absence of the atrial septum a variety of atrioventricular canal deformity and believe that atrioventricular valve anomalies are always present in this condition. The first report of this condition was made by Lewis et al. [4] who used the term "continuous defect" of the atrial septum. Watkins and Gross [5] described two instances of complete absence of the atrial septum among 43 patients who underwent surgical correction of atrial septal defects by a closed technique. Similar cases were also reported by Probyn-William [6], Cunningham [7], Dubost and Blondeau [8] and Munoz-Armas et al. [9]. In none of their cases was an associated anomaly of the

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atrioventricular valve present. In atrioventricular canal with complete absence of the atrial septum, partial development of the septal elements may have occurred, but the atrioventricular endocardial cushions have failed to grow adequately, and as a consequence, the endocardial cushions and primitive septal structures failed to fuse. The inevitable tension on the septal structures resulting from further growth of the heart, we believe, leads to a progressive increase in the size of the defect between the atrial septal structures and the atrioventricular endocardial cushions. Thus, the principal factor in the causation of this defect would seem to be abnormal development of the A-V endocardial cushions with secondary regression of the atrial septal structures. In such cases, complete absence of the atrial septum will be associated with endocardial cushion defect. In single atrium, the dinical picture does not differ from that of a large atrial septal defect at the level of the fossa ovalis [10].

Patients with common atrium, however, show a decrease in exercise tolerance early in life, increased fatigability, shortness of breath, mild cyanosis or obvious heart failure.

The physical findings in single atrium are typically those of atrial septal defect of the fossa ovalis type. There is prominence of the precordial area, a soft systolic murmur at the pulmonary area and a constant wide splitting of the pulmonary second sound. In common atrium, in addition to the above mentioned physical signs, there is a high pitched systolic murmur at the apex radiating towards the axilla, characteristic of mitral regurgitation. The radiologic findings in both groups are similar to those found in the ordinary type of atrial septal defect, namely: cardiomegaly of variable degree due to enlargement of the right cardiac chambers, with normal left cardiac chambers. Often there is a prominent pulmonary artery segment at the hilar vascular shadow, and plethora of the peripheral branches of the pulmonary vasculature. There is commonly only slight enlargement of the right atrium. From a hernodynamic viewpoint [11], demonstration of complete mixing between systemic venous and oxygenated pulmonary venous blood at the atrial level is rarely found in single atrium.

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