Case Report

42 Years Old Male with Single Atrium

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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. We report here such a case of SA who presents his symptoms in different way of its natural course of presentation.

Introduction

Single atrium (SA) is a rare variety of interatrial communication, occasionally associated with abnormalities of the venae cava and coronary sinus, is thought to be one form of endocardial cushion defect and is first described by young and Robinson 1907.

Right sided portion of common chamber has anatomic features of right and left sided portion has anatomic features of left atrium, receives blood from pulmonary vein.

Despite complete absence of atrial septum appreciable venoarterial mixing is exceptional. Systemic $O_2$ saturation $>90\%$ are not uncommon. Majority of patients are symptomatic during first year of life with dyspnea on effort, fatigue respiratory tract infection, mild cyanosis, physical underdevelopment. Symptoms of single atrium resembles that of nonrestrictive atrial septal defect but are earlier in onset and more pronounced although occasional patients are relatively well into late childhood or early adolescence.

Cyanosis sometimes absent or insignificant when the patient is at rest but almost always present after excersise.

We describe here such a case of Single atrium (SA) who presents his symptomatology at adult age.

Case Report

A 42 yrs. old man was referred to our hospital with gradual onset of fatigue exertional dyspnea, palpitation for the last 2 years. (which was classified as NYHA functional class grade II). Clinical deterioration occurred over 5-6 month leading to features of failure. On clinical examination patient was dyspneic(NYHA class III), prominent neck vein, significant pedal edema enlarged tender hepatomegaly, no icterus, this patient remain asymptomatic until adulthood.

Routine blood examination was within normal limit.

The chest radiograph revealed gross Cardiomegaly with LA enlargement prominence of right descending pulmonary artery Fig. 1.

Fig. 1 : X-ray chest

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The ECG revealed an atrial rhythm and incomplete RBBB.

Echocardiogram revealed absence of atrial septum with pulmonary hypertension with an intact intraventricular septum. Figs. 2a and 2b.

Cardiac catheterization was advanced into the left atrium and revealed that O₂ was stepped up in the right atrium, the left to right shunt ratio was 90%, the systolic right ventricular pressure was 35 mmHg and pulmonary arterial pressure was 28 mmHg.

**Discussion**

Previously, Single atrium (SA), a rare congenital anomaly was thought to be a form of endocardial cushion defect.

According to Campbell and Nissen, the anomaly can be classified into three groups i) persistent ostium primum ii) A partial atrioventricular (AV) canal iii) and a complete AV canal.

Leavy and associates however, reported a case of complete absence of the atrial septum, indicating that this condition may exist alone as a specific entity without an endocardial cushion defect. They recommended that the term single atrium should be used to denote the condition characterized by 1) complete absence of the atrial septum, 2) absence of the malformation of AV valve and 3) 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.

Here in our case there is complete absence of atrial septum, without any endocardial cushion defect, malformation of atrioventricular (AV) valve or IV communication who presents his symptoms at adult age not like that of CA which usually appear in early childhood or occasionally into late childhood and early adolescence.

According to Leavy associates our patient of single atrium without having AV valve malformation or IV communication who came with feature of heart failure in adulthood.

So our interest of presenting this case are: 1. It is a rare variety of congenital anomaly. 2. Late presentation of clinical manifestation beyond its natural course. 3. Can masquerade sign and symptoms of nonrestrictive ASD. 4. Can be a cause of congestive heart failure in adult age.

**Abbreviation**

RV – Right Ventricle, PT – Pulmonary trunk ; RA-Right atrium; LV-Left ventricle; cA-Common atrium, LPA – Left pulmonary artery.

**References**

2. Young AH, Rooison A. Some malformation as the human heart. M Chron 1907/1908;47:96.