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Two Congenital Left-to-Right Shunt Anomalies in a Septuagenarian: ARare Occurrence

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CASE REPORT

Two Congenital Left-to-Right Shunt Anomalies in a Septuagenarian: A Rare Occurrence

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ABSTRACT

Atrial septal defect (ASD) and patent ductus arteriosus (PDA) are common congenital anomalies presenting in childhood. Life expectancy of an uncorrected PDA is shortened to half; and ASD of a significant size has increased morbidity and mortality. Their co-existence in an elderly patient with first presentation at 70 years of age is a rarity. We present the case of a 70-year woman with one-week history of dyspnea with high blood pressure and signs of heart failure. She was found to have a PDA and an ASD with left-to-right shunt. She was managed conservatively. She was offered cardiac catheterization, but she refused. This is the first documented case in local literature with two such congenital heart defects presenting in a septuagenarian. In a country where average life expectancy is in the 60's, the survival of the patient with two heart defects, beyond-average survival age, is interesting.

Key Words: *Septal defect. Atrial. Ductus arteriosus. Patent. Heart failure.*

INTRODUCTION

Atrial septal defect (ASD) is a cardiac anomaly that causes shunting of blood between the pulmonary and systemic circulation. It frequently remains undiagnosed in children, with 96% probability of survival into adulthood.[1] Ductus arteriosus is a normal vascular structure that connects descending aorta to the pulmonary artery in the embryonic period, becoming abnormal if it remains patent after first few weeks of life patent ductus arteriosus (PDA). It accounts for upto 14% of all congenital heart defects.[2]

The co-existence of two congenital anomalies in an elderly patient has not been described in local literature. To the best of the authors' knowledge, they report the first case of a 70-year woman from Karachi with co-exisiting PDA and ASD.

CASE REPORT

A 70-year woman presented to the emergency room (ER) at the Aga Khan University Hospital, with dyspnea at rest, orthopnea and paroxysmal nocturnal dyspnea for one week. She gave a 4-year history of hypertension, but was not taking medications. Her past history revealed that she had some dyspnea on severe exertion since childhood, limiting extremes of physical activity. She had 4 children delivered via spontaneous vaginal delivery at a local clinic.

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On examination, she had a blood pressure (BP) of 180/100 mmHg, heart rate of 112 bpm, oxygen saturation of 96% on room air and a temperature of 37.2°C. She had bilateral pitting pedal edema, a fixed split of the second heart sound (S2) with a loud pulmonary component. A continuous "machinery" murmur was best heard at expiration in the second left, parasternal, intercostal space with a palpable thrill. Chest auscultation revealed bilateral rales and wheeze.

ECG revealed sinus tachycardia with no ST/T wave changes. Chest X-ray (Figure 1) revealed gross cardiomegaly with pulmonary edema. A transthoracic echo (Figure 2) revealed severe biatrial and mild right ventricular enlargement. The left ventricular systolic function was reduced, with a visually estimated ejection fraction of approximately 30% and moderate to severe global hypokinesia. Aortic and mitral valves were thickened and calcified with mild mitral regurgitation.

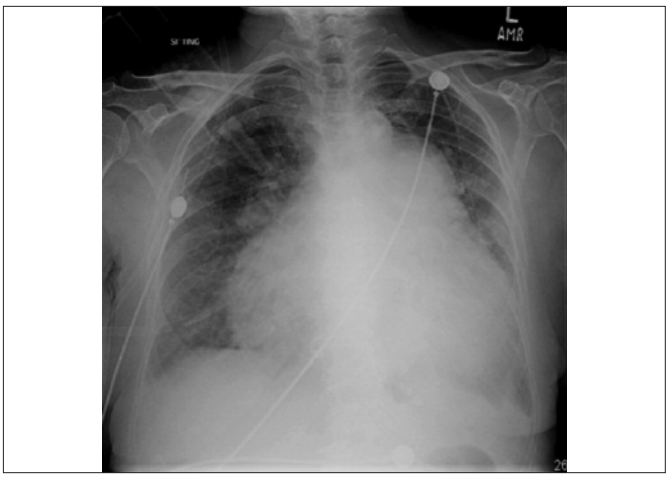

Figure 1: Chest Xray: Sitting, AP view showing centrally placed trachea with an intact bony cage and visualized bones appearing slightly osteopenic. There is gross cardiomegaly with prominent hilar congestion. Gastric bubble is visualized.

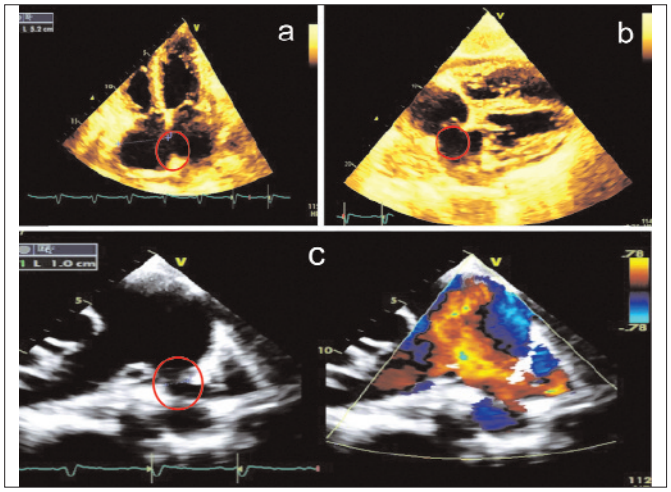

Figure 2: Echo images **(a,b)** show atrial septal defect as marked with the red circle; echo image **(c)** shows without and with colour doppler images of patent ductus arteriosus as marked with the red circle.

Moderate tricuspid regurgitation was present with a peak pressure gradient of 55 mmHg. The pulmonary arteries were dilated with an estimated pulmonary artery systolic pressure of 70 mmHg. Inferior vena cava was dilated with loss of inspiratory collapse. There was a PDA with a left-to-right shunt and a secundum ASD (14 mm in diameter, with additional fenestrations) with a left-to-right shunt. There was mild circumferential pericardial effusion without signs of cardiac tamponade.

She was admitted with a cardiac decompensation due to congenital heart disease and elevated BP. She was started on glyceryl trinitrate (GTN) infusion and intravenous furosemide. The GTN infusion was overlapped with perindopril and amlodipine and later the infusion was stopped. The response to treatment was adequate, after which her chest cleared. The intravenous furosemide was switched to oral and carvedilol and aspirin were added. Cardiac catheterization was advised but she refused further invasive workup.

Over the course of admission, she became asymptomatic and her BP settled within normal range. She was discharged on fluid and salt restriction, regular medical therapy and cardiac rehabilitation plan, and was to be followed in outpatient clinic.

DISCUSSION

PDA and ASD in this age are rare. It is important to have knowledge of congenital anomalies while thinking common things in this age group. Coming to correct diagnosis, one can treat patients' hypertension effectively and make their lives comfortable. Literature review has shown a few documented cases of PDA in elderly patients worldwide. The two oldest cases of patients with PDA in literature have been of a 92-year old Japanese woman with infective endocarditis,[3] and a 90-year old man from USA.[4] In Pakistan, a case of a 27-year old woman with PDA successfully treated in her second trimester of pregnancy is reported.[5] Another study in Pakistan presented data for 208 cases with ages ranging from 19 to 73 years with congenital cardiac disease.[6] However, separate age groups for different defects were not reported.

Prior studies show that the mortality rate of uncorrected PDA in adults is around 1.8% per year,[7] and survival of patients with unrepaired ASD into 90th decade.[8] However, ASD of significant size with right-sided heart dilatation are associated with morbidity and mortality and should be considered for surgical closure regardless of age.[9]

The average life expectancy in Pakistan is 67 years for females and even lesser for males.[10] In such a country, our patient's survival into the 70th decade with two uncorrected anomalies, beyond average life expectancy with minimal symptoms, is indeed interesting. She was offered cardiac catheterization for further management of her PDA and ASD, but she refused because of her faith in the Divine that she lived long-enough with two rare anomalies; and believed she would live longer without further intervention. This was similar to the 72-year old Polish lady with PDA and PFO who was offered interventional occlusions but refused.[11]

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