

Aortopulmonary window- A rare presentation in post-partum female.

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Abstract

Aortopulmonary window (APW) is a rare anomaly characterized by anomalous communication between aorta and pulmonary artery with separate aortic and pulmonary origins. This accounts for approximately 0.1 % of all congenital cardiac anomalies. Clinical presentation is in infancy and early childhood with left to right shunt. If not corrected, large defects are usually fatal in early childhood. We report a 25 year old patient with Eisenmenger syndrome with large unrepaired APW, who successfully completed her first pregnancy.

Keywords: Aortopulmonary Window, Congenital Heart Anomalies, Eisenmenger syndrome

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Introduction:

During early embryonic development, the aorta and pulmonary arteries separate by growth of a spiral septum dividing the common trunk into the aorta and the pulmonary artery. This spiral septum is created by fusion of a truncal septum growing cephalad from the semilunar valves and the aortopulmonary spiral septum growing caudally from the pulmonary bifurcation. Incomplete development of these septae results in aortopulmonary septal defect [1]. This accounts for approximately 0.1 % of all congenital cardiac anomalies. Clinical presentation is in infancy and early childhood with left to right shunt. If not corrected, large defects are usually fatal in early childhood.

Case Report

A 25 yrs old postpartum female, presented to medical emergency with cyanosis and clubbing with features suggestive of puerperal sepsis on 7th postpartum day, for which she was admitted and managed accordingly. Subsequently, patient revealed occasional episodes of cyanosis and palpitation on exertion during last 3 years. Otherwise her pregnancy was uneventful. Clubbing was present and jugular venous pressure was raised.

On cardiac auscultation heart sounds were feeble at cardiac apex and loud P2 heard on left 2nd parasternal space

with prominent heart sounds in right parasternal region. No definite murmur was audible. She had no significant abdominal organomegaly. X-ray chest revealed tracheal deviation and mediastinal shift to right. Electrocardiogram showed P pulmonale and features of right ventricular hypertrophy and right axis deviation. 2D-echocardiography revealed severe pulmonary artery hypertension with dilated right atrium and ventricle and paradoxical motion of interventricular septum. No intracardiac shunt was seen.

On CECT chest there was evidence of large abnormal communication between main pulmonary trunk and ascending aorta with small sized right lung. On computed tomography (CT) angiography the aortopulmonary defect was again seen, from just above the aortic and pulmonary origin, forming a common trunk and then continuing as left main pulmonary artery and ascending aorta. The right pulmonary artery was not identified.

The tricuspid aortic and pulmonary valves were normally seen. Origin of right coronary artery was normal, while the left coronary artery was seen originating about 1.5 cms higher, just at the level of shunt, shown in Figure 1 (a-d).

Catheter angiography could not be performed due to patients' unwillingness.

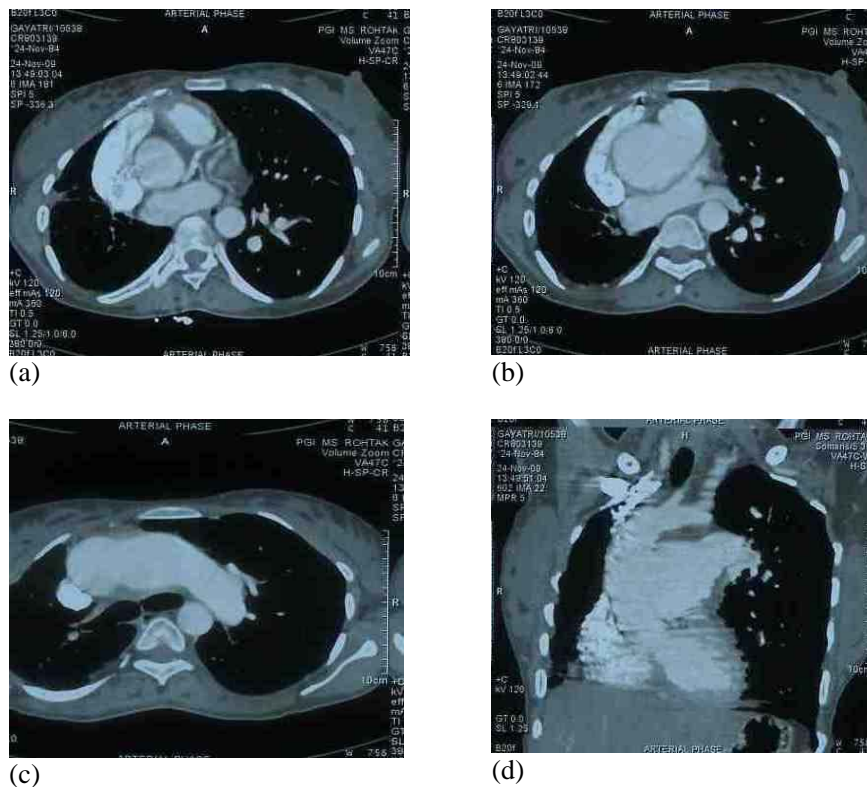


Figure 1.

- There is separate origin of the aorta and pulmonary artery with lower parts seen separate from each other. The left coronary artery is seen arising high up from aorta.
- There is abnormal communication of the ascending aorta and pulmonary arteries with a large common channel.
- Upper extent of the lesion is seen with division into arch of aorta and left main pulmonary artery.
- Coronal reformat better displays the aortopulmonary defect, seen dividing into arch of aorta and left main pulmonary artery.

Discussion

During early embryonic development, the aorta and pulmonary arteries separate by growth of a spiral septum dividing the common trunk into the aorta and the pulmonary artery. This spiral septum is created by fusion of a truncal septum growing cephalad from the semilunar valves and the aortopulmonary spiral septum growing caudally from the pulmonary bifurcation. Incomplete development of these septa results in aortopulmonary septal defect. Aortopulmonary window is described as a defect in the aortopulmonary septum with separate origins of aorta and pulmonary arteries and normal semilunar valves, separating this condition from truncus arteriosus. DiGeorge syndrome, associated with truncus arteriosus is not significantly associated with APW, suggesting separate developmental origins of these morphologically very similar conditions [2]. This accounts for approximately 0.1 % of all congenital cardiac anomalies [3]. Of the two classifications systems proposed by Mori in 1978 [4], and

by Richardson, in 1979 [5], Mori's classification is more used and describes three types of defects. Type I, or proximal: the communication is usually rounded and located above the semilunar valves, between the ascending aorta and the pulmonary trunk. Type II, or distal: the defect, in a spiral curve, involves the pulmonary bifurcation at the level of the right pulmonary artery (RPA). In this type of defect, there can be hypoplasia or even interruption of aortic arch due to the significant deviated flow from the aorta towards the RPA during fetal life. Type III is characterized by total absence of the aortopulmonary septum resulting from the combination of proximal and distal defects. Type I (proximal) defects occur in proximal part of aortopulmonary septum between aorta and main pulmonary artery. Type II (distal) in distal part of the aortopulmonary septum, extending into the RPA. Type III is a combination of the type I and II. Type I defects are the commonest and type III are the rarest [6].

About 50% of cases of APW are associated with other defects, and this makes its diagnosis more difficult. Ac-

According to the literature, the most frequently associated anomalies are: aortic arch interruption (15-20%), especially type A; patent ductus arteriosus (11%); ventricular septal defect (8%); right aortic arch (7%); anomalies of the coronary arteries (8%); tetralogy of fallot (5%); subaortic stenosis (3%); bicuspid aortic valve (3%) [6]. Development of the coronary arteries also occurs during the separation of the great vessels, which explains anomalous position of the coronary arteries. The associated cardiovascular conditions seen in our case are hypoplastic right pulmonary artery with hypoplasia of right lung and relatively higher origin of left coronary artery.

Recently echocardiography, CT angiography and magnetic resonance imaging have also been used and found sufficient for diagnosis [7,8].

The majority of patients present in infancy with congestive cardiac failure. Untreated patients usually die in early childhood. Adults usually present with dyspnea on exertion, occasional palpitations and episodes of cyanosis with history of recurrent respiratory tract infections. Usually a systolic murmur is heard with evidence of pulmonary artery hypertension. Our patient completed her pregnancy uneventfully.

Definitive treatment is surgical. Few patients surviving into adulthood usually have a very high pulmonary vascular resistance making them inoperable. Aggarwal et al., [9] have reported a few successfully operated adult cases. According to them surgical repair can still be performed in patients with Eisenmenger syndrome, if there is significant reversibility of pulmonary vascular resistance index on administration of oxygen.

Disclosures

The authors have no financial conflict of interest.

Abbreviations used: APW, Aortopulmonary Window; CT, Computed Tomography; RPA, Right Pulmonary Artery;

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