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Current Cardiology Reviews, **2020,** *16,* **73-76**

CASE REPORT

Large Unrepaired Aortopulmonary Window Presenting in Adulthood

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ARTICLE HISTORY

Received: March 05, 2019
Revised: April 29, 2019
Accepted: April 30, 2019

DOI:
10.2174/1573403X15666190513105231

Abstract: ***Background*:** Aortopulmonary window is an uncommon congenital heart disease, with untreated cases not surviving beyond childhood. However, very rarely it can present in adult patients with features of pulmonary hypertension. Clinically these patients cannot be differentiated from other more common conditions with left to right shunt. Transthoracic echocardiography if performed meticulously, can depict the defect in aortopulmonary septum.

***Results*:** We report a case of large unrepaired aortopulmonary window in a 23 years old patient, diagnosed on transthoracic echocardiography.

Keywords: Echocardiography, congenital heart disease, pulmonary artery hypertension, computed tomography, aortopulmonary window, transthoracic echocardiography.

1. INTRODUCTION

The Aortopulmonary (AP) window or aortopulmonary septal defect is a rare congenital heart disease accounting for 0.2-0.4 % of all congenital heart diseases [1]. It is characterized by an abnormal communication between the ascending aorta and the pulmonary trunk in the presence of two semilunar valves, resulting from the anomalous division of aortopulmonary trunk during embryogenesis. Most cases present in infancy with heart failure and if left untreated, do not survive beyond infancy and early childhood [2]. Patients with a small defect can present in adulthood with symptoms of Pulmonary Arterial Hypertension (PAH) [3]. We report a case of large AP window with presentation in adulthood.

2. CASE REPORT

A 23-year-old male was presented to the out-patient department of our institute with complaints of dyspnoea (NYHA class –II) for last one year. No history of orthopnoea or paroxysmal nocturnal dyspnoea was present. The patient also gave a history of regular palpitations on exertion for the last five years for which he did not seek any medical consultation. There was no history of frequent chest infections in childhood. No history of cyanosis, abdominal fullness, or pedal edema was present. On examination, his blood pressure was 142/56 mmHg, pulse rate was 92bpm and respiratory rate was 16 /minute. Oxygen saturation on room air was 92%. On cardiovascular examination, there was grade 3 parasternal heave with palpable P2. On auscultation, P2 was loud. There was a grade 3/6 systolic murmur over the left parasternal area in 2nd and 3rd intercostal space and another pansystolic murmur over lower left parasternal area i increasing with inspiration suggestive of tricuspid regurgitation.

Electrocardiography (ECG) showed normal sinus rhythm with features of biventricular hypertrophy. Chest x-ray posteroanterior view (Fig. **1**) showed cardiomegaly with down and out apex (suggesting left ventricular pattern of enlargement). Main pulmonary artery (MPA) was enlarged with dilated right descending pulmonary artery (measuring ~17mm). In addition, there was a paucity of vascular markings in outer one-third of lung fields bilaterally. On transthoracic echocardiographic examination, there was Left Ventricular (LV) enlargement (LVEd: 79mm) with normal LV systolic function (LV ejection fraction: 62%, measured using modified Quinones equation by M-mode) and mild tricuspid regurgitation (TR gradient: 92mmHg). There was turbulence in MPA with the presence of AP window (defect size: 29 mm) in parasternal short axis view (Fig. **2A)**. Colour Doppler showed the presence of left to right shunt (Fig. **2B**) with a peak gradient of 45 mmHg. No other associated cardiac anomaly was found. The patient subsequently underwent Computed Tomography (CT) pulmonary angiography, which confirmed the diagnosis of AP window (Fig. **3A**). The defect measured ~32mm anteroposteriorly and ~28mm craniocaudally, and was located midway between semilunar valves and bifurcation of the main pulmonary artery. MPA was dilated, measuring ~38mm. Right, and left pulmonary arteries were also dilated (measuring ~25mm and 24mm, respectively). In addition, first and second order intraparenchymal branches of pulmonary arteries were also dilated with the pruning of distal branches (Fig. **3B**). Patchy areas of mosaic

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1573-403X/20 $65.00+.00

attenuation were also observed in bilateral lungs (Fig. **3C**) suggestive of perfusion related changes.

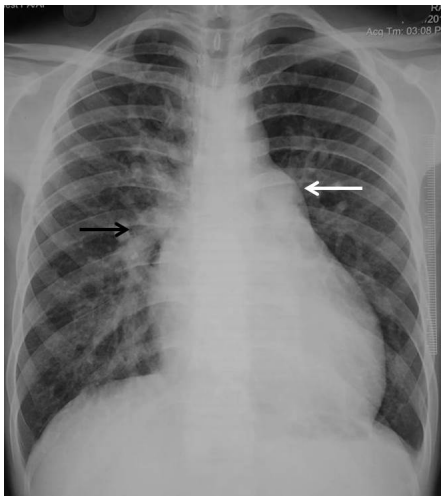

Fig. (1). Chest x-ray PA view showing cardiomegaly with down and out apex. Main pulmonary artery is enlarged (white arrow) with dilated right descending pulmonary artery (black arrow). Also note relative paucity of vascular markings in peripheral lung fields. *(A higher resolution / colour version of this figure is available in the electronic copy of the article).*

A final diagnosis of AP window with severe PAH was made. The patient was advised catheterisation study to rule out irreversible PAH (Eissenmenger syndrome), as it is a contraindication for corrective surgery. However, the patient refused catheterisation study and requested a referral to a higher center due to the non-availability of corrective surgery in our institute. Unfortunately, the patient was lost to follow up after the referral.

3. DISCUSSION

AP window or aortopulmonary septal defect is a rare cardiac anomaly representing the anomalous division of aortopulmonary trunk during embryogenesis [4]. It may occur in isolation or in conjunction with other congenital heart diseases like ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA) and tetralogy of Fallot (TOF) [5]. In the AP window, abnormal communication is present between ascending aorta and pulmonary trunk. Mori *et al*. [6] classified the AP window into three types. Type I defect is located midway between the pulmonary bifurcation and semilunar valves (as in our case). Type II defect is present between the anomalous right pulmonary artery and ascending aorta. Type III defect is a large defect involving the aortopulmonary septum in its entire extent.

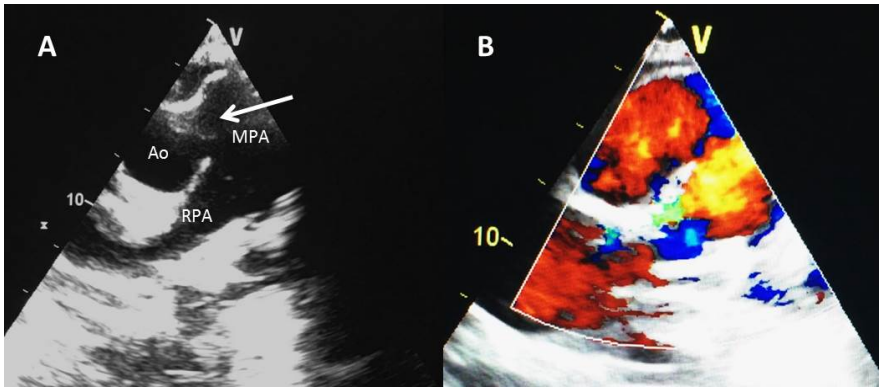

Fig. (2). A: Transthoracic echocardiography, parasternal short axis view demonstrating a defect (white arrow) between aorta (Ao) and main pulmonary artery (MPA). **B**: Color Doppler image showing shunting of blood across the defect. Also note presence of turbulence in main pulmonary artery (evident by bright color signal). *(A higher resolution / colour version of this figure is available in the electronic copy of the article).*

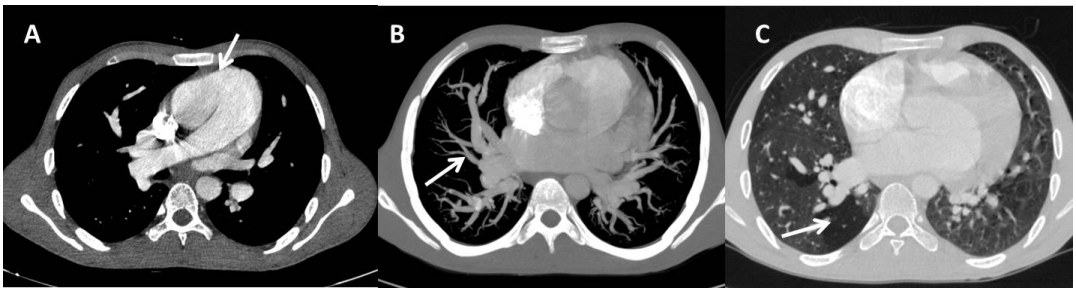

Fig. (3). A: Axial CT chest image, (mediastinal window) showing abnormal communication between ascending aorta and main pulmonary artery (arrow). Also note dilated main pulmonary artery. **B**: Axial maximum intensity projection (MIP) image showing sudden tapering of peripheral intraparenchymal branches of pulmonary artery (arrow). **C**: Axial CT chest image in lung window demonstrating mosaic attenuation (alternating areas of increased and decreased attenuation) in visualized portion of right lung. Note decreased number of vessels in areas with decreased attenuation (white arrow), indicative of oligaemia.

Video: Parasternal short axis view on transthoracic echocardiography demonstrating abnormal communication (white arrow) between ascending aorta (Ao) and main pulmonary artery (MPA). *(A higher resolution / colour version of this figure is available in the electronic copy of the article).*

Most cases of AP window present in infancy with associated high mortality. Neonates present with symptoms of congestive heart failure like sweating, tachypnea and failure to thrive. A forceful apical impulse is present on clinical palpation due to the left ventricular volume overload. On auscultation, a murmur can be heard in second and third intercostal space. Adult patients present with symptoms of breathlessness and palpitations. A history of recurrent chest infections in childhood may be present [2]. A murmur is present on auscultation. Large defects will be presented with the associated PAH. Clinically, AP window cannot be differentiated from other left to right shunts: a large VSD or PDA and persistent truncus arteriosus.

Chest X-ray and ECG demonstrate features of left ventricular overload and PAH. Echocardiography remains an important technique in the diagnosis of AP window. Transthoracic echocardiography usually demonstrates the defect, although distal lesions may be missed. Doppler shows abnormal continuous flow from aorta into the pulmonary artery. On transthoracic echocardiography, a dropout can occur in the region of the aortopulmonary septum in normal patients thereby mimicking AP window. However, evaluating the septum in two or more planes and recognizing "T" artefact at the edge of the defect can help to differentiate normal dropout from actual defect [7]. In a patient with features of left ventricular overload and PAH, without any other shunt lesion, the possibility of AP window should be considered. In doubtful cases, transesophageal echocardiography can be used for diagnosis [2]. Rarely, the defect can be detected on cardiac catheterization only [4]. Cardiac catheterization not only helps in confirming the diagnosis but can also be used to study other associated cardiac anomalies and to assess the operability of the lesion. However, it is an invasive procedure with inherent risks.

CT is a non-invasive modality which can be used in the diagnosis of doubtful cases. However, its role largely remains unexplored in the diagnosis of AP window. CT angiography can accurately depict the size and type of defect [8]. In addition, associated PAH and related lung parenchymal changes can also be evaluated. Magnetic resonance imaging (MRI) can also be used for the diagnosis of AP window [9]. However, it is expensive and not widely available.

Early treatment of AP window is recommended to avoid progression to irreversible PAH. Absence of other anomalies and early surgery favour good outcome. Median survival in untreated AP window has been reported to be 33 years [4]. Surgical techniques include transaortic or transpulmonary patch closure of the defect. Aggarwal *et al*. reported successful closure of the AP window in six adult patients [2]. Percutaneous transcatheter device closure of small defects has also been reported [10]. Development of irreversible pulmonary hypertension is a contraindication to surgery and carries adverse prognosis. Treatment of these patients includes pulmonary vasodilators (endothelin receptor antagonists and phosphodiesterase -5 inhibitors), anticoagulant drugs, diuretics and phlebotomy sessions [11, 12].

To conclude, the AP window is a rare congenital cardiac anomaly and can be rarely present in adult patients. In a patient with signs of left ventricular overload and PAH with the absence of other common shunt lesions like VSD and PDA, a possibility of AP window should be considered and a meticulous search should be done for a defect in aortopulmonary septum on transthoracic echocardiography. Early diagnosis of this rare heart disease is desirable for prompt management before irreversible PAH sets in.

CONCLUSION

Aortopulmonary window or aortopulmonary septal defect is a rare congenital heart disease characterized by abnormal communication between the ascending aorta and the main pulmonary artery in the presence of separate semilunar valves. Most cases are present in infancy and early childhood with very few cases presenting in adulthood.

Clinically it presents with features of left to right shunt and cannot be differentiated from other more common shunt lesions like VSD and PDA.

The defect can be easily visualized on transthoracic echocardiography. Cardiac catheterization and CT angiography can provide a diagnosis in difficult cases.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethical clearance was not required for this study as the investigations were done as a part of routine patient work up.

HUMAN AND ANIMAL RIGHTS

Not applicable.

CONSENT FOR PUBLICATION

Informed written consent was obtained from the patient.

STANDARD OF REPORTING

CARE guidelines and methodology were followed in this study.

FUNDING

None.

CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

ACKNOWLEDGEMENTS

Declared none.

SUPPLEMENTARY MATERIAL

Supplementary video file is available on the publisher's web site along with the published article.

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