Case Study

A large Aortopulmonary window with a ventricular septal defect: A rare combination presenting at the age of 16.

ABSTRACT

Aortopulmonary window is a rare congenital anomaly with a communication between ascending aorta and main pulmonary artery. It may be associated with other cardiac malformations like aortic arch anomalies, ventricular septal defect, tetralogy of fallot etc. Survival beyond infancy is rare and early surgical intervention is important to prevent development of irreversible pulmonary hypertension. We present a rare case of larger Aortopulmonary window along with a large ventricular septal defect asymptomatic till the age of 16 years.

Keywords: ***Aortopulmonary window, Ventricular septal defect, Pulmonary hypertension.***

1. INTRODUCTION

Aortopulmonary window (APW) is a rare congenital anomaly and represents 0.2 to 0.4 % of all congenital heart diseases.[1] There is an abnormal communication between the ascending aorta and the main pulmonary artery in presence of two separate semilunar valves. APW may be associated with variety of other congenital malformations like interruption of aortic arch, Tetralogy of fallot, ventricular septal defect (VSD) or coronary artery anomalies.

2. CASE REPORT

A 16 years old girl was referred to our hospital with complaints of multiple episodes of syncope and dyspnea on exertion (NYHA class III) since 6 months. She had minimal symptoms in childhood but never sought medical attention and successfully completed her high school. The recent onset syncopal episodes were associated with physical exertion. There was no history of cyanosis, squatting episodes, hemoptysis or recurrent chest infections in childhood. There was no history of similar or any other major illness in the family. On examination cyanosis along with grade I clubbing was noted. Saturation in room air was 87%. There was grade III parasternal heave along with a single S2. ECG showed significant right ventricular hypertrophy. Transthoracic Echo was suggestive of a large ventricular septal defect (22mm) along with severe pulmonary hypertension. (Figure 1) A cardiac catheterization was subsequently done which revealed a large perimembranous ventricular septal defect with equalization of pressures in both ventricles (Figure 2) along with an anomalous left superior venacava draining into right atrium. Ascending aortogram showed simultaneous opacification of main pulmonary artery and its branches (aortopulmonary window Type I) (Figure 3). Mean pulmonary artery pressure was 82mmHg. Oximetry study was conducted which showed Qp: Qs ratio of 0.5. The pulmonary vascular resistance was 29 woods units. After 100% oxygen there was no reduction in pressures and PVR. In view of no significant reversibility the patient was deemed inoperable and has been advised medical management. She is currently stable at follow-up. Computerized tomography (CT) was done, which showed a large Aortopulmonary window (type I defect) with both right and left branch pulmonary arteries arising from main pulmonary artery. Three dimensional reconstruction with volume rendering images have been shown. (Figure 4)

3. DISCUSSION

APW represents anomalous division of a common aortopulmonary trunk during embryogenesis. Aortopulmonary window in association with other congenital anomalies like a VSD presenting in adolescence is uncommon. The large communication between aorta and main pulmonary artery seldom closes spontaneously or reduces in size with time. Elliot first described it in 1830. [2]

Mori's classification is widely accepted one which divides APW into three types.[3] Type 1: A proximal defect midway between the aortic valve and the bifurcation of pulmonary artery (most common). Type II: A distal defect between the anomalous right pulmonary artery and the ascending aorta and Type III: a large confluent defect usually with absence of aortopulmonary septum. The large left to right shunt present at birth results in congestive heart failure with pulmonary hypertension within first month of life.

The condition is fatal in most cases if untreated in infancy or early childhood hence early surgical intervention is important to prevent early development of irreversible pulmonary vascular disease. Although rare such late presentation in adolescence or adulthood is possible in a developing country like ours because of various socioeconomic factors. In a study of 20 patients from Mumbai, India, 35% of patients with APW were older than 15 years.[4] The neonates usually present with tachypnea, sweating and failure to thrive. Clinically a bounding arterial pulse along with a forceful apical impulse (due to LV volume overload) can be felt. A pulmonary ejection click may be heard sometimes. A systolic murmur is audible over left second or third intercostal space. Few, who escape infancy rapidly develop suprasystemic pulmonary vascular resistance and resemble a VSD or PDA with eisenmenger's syndrome. Auscultatory signs of severe pulmonary hypertension will predominate and no murmur is generated across the defect. Chest X-ray and ECG findings are typical of any condition causing left ventricular volume overload and PAH.

Echocardiography is the preferred modality for diagnosis. Color flow imaging identifies abnormal continuous forward flow from aorta into pulmonary artery. Doppler can be useful in differentiating APW from a patent ductus arteriosus, (PDA) with demonstration of diastolic flow reversal as seen in PDA.Cardiac catheterization is usually performed 1) to confirm the diagnosis 2) to study other associated congenital anomalies 3) perform Vasoreactivity testing for operability. Advances in investigative and surgical modalities have led to favorable outcomes with low risk.

Gross first reported successful surgical closure of APW in 1952.[5] Several surgical techniques like trans pulmonary or a trans aortic patch closure have been described subsequently with variable success. Transcatheter device closure of simple defects with good margins is also possible Such successful Transcatheter closure of APW has been occasionally reported. [6] Aggarwal et al from India have described successful surgical closure of APW in six patients with mean age of 21 years.[7] Postoperative mortality depends on the age of the patient at operation, extent of pulmonary vascular disease, and presence of other cardiac defects. The emergence of irreversible pulmonary vascular resistance although precludes any intervention and has adverse prognosis.

A large aortopulmonary window coexisting with a large ventricular septal defect with irreversible pulmonary vascular resistance presenting at the age of 16 years makes this case a rare occurrence.

4. CONCLUSION

Aortopulmonary window is a rare congenital anomaly which clinically resembles a large nonrestrictive VSD or PDA. High mortality in infancy along with the rapid and early onset of irreversible pulmonary vascular disease underlines the importance of early surgical correction.

CONSENT

"All authors declare that 'written informed consent was obtained from the father of the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal."

ETHICAL APPROVAL

As the report is a case presentation formal ethics approval is not applicable. The report has been conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

REFERENCES

1. Richardson JV, Doty DB, Rossi NP, Ehrenhaft JL. The spectrum of anomalies of aortopulmonary septation. J Thorac Cardiovasc Surg 1979; 78:21–7.
2. Elliotson J. Case of malformation of the pulmonary artery and aorta. Lancet 1830; 1:247-51.
3. Mori K, Ando M, Jakao A, Ishikawa S, Imai Y. Distal type of aortopulmonary window: report of 4 cases. Br Heart J 1978; 40:681-689.
4. Pinto, Robin & Bhagwat, Ajit & Loya, Yuridia & Sharma, Satyavan. (1994). Profile of aortopulmonary window in India—A study of twenty cases. Cardiology in the Young. 4. 142 - 145. 10.1017/S1047951100002080.
5. Gross RE. Surgical closure of an aortic septal defect. Circulation 1952; 5:858-63.
6. Trehan V, Nigam A, Tyagi S. Percutaneous closure of nonrestrictive aortopulmonary window in three infants. Catheter Cardiovasc Interv 2008; 71:405–11.
7. Aggarwal, S. K., Mishra, J. , Sai, V. , Iyer, V. R. and Panicker, B. (2008), Aortopulmonary Window in Adults: Diagnosis and Treatment of Late - presenting Patients. Congenital Heart Disease, 3: 341-346. doi:10.1111/j.1747-0803.2008.00210.x.

Figures :

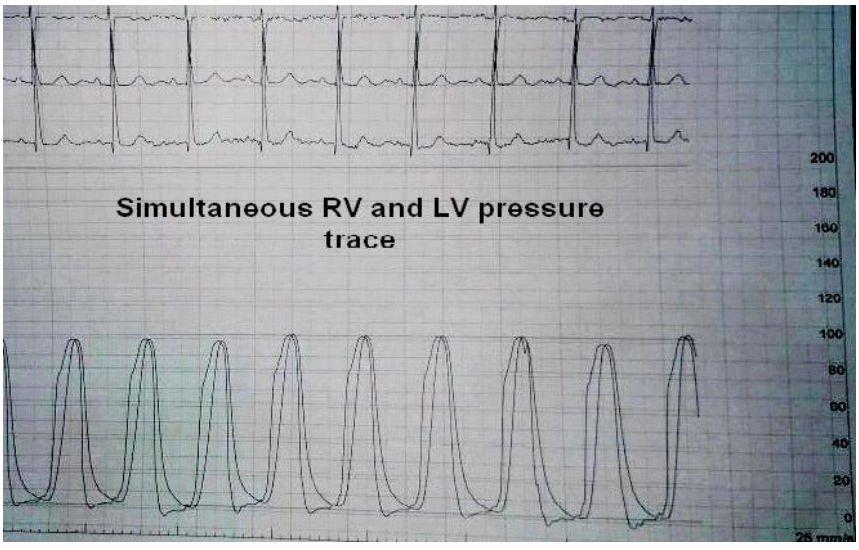

Figure 1- Simultaneous RV & LV pressure tracing suggestive of equalisation of ventricular pressures.

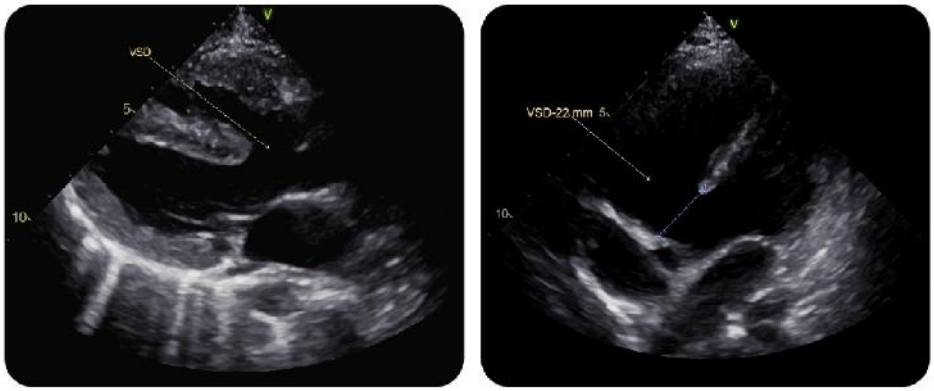

Figure 2- Echo - parasternal long axis view and apical 4 chamber view showing large VSD.

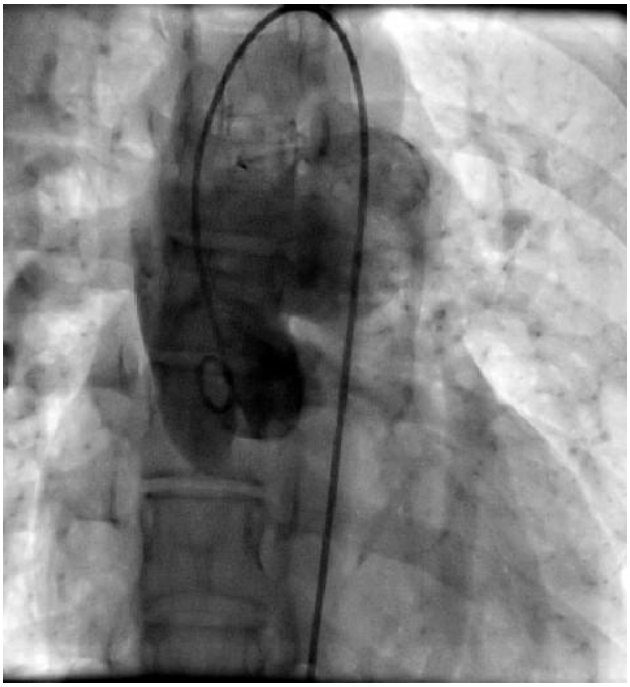

Figure 3- Aortogram showing large communication between ascending aorta and main pulmonary artery.

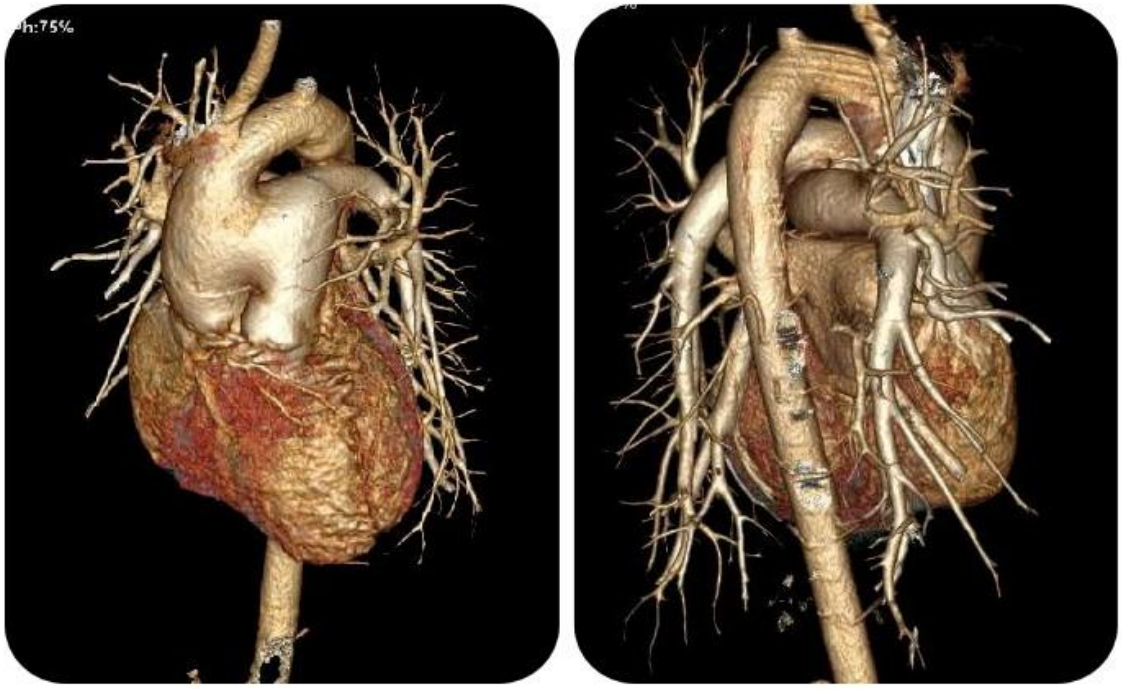

Figure 4-Volume rendered CT images clearly depicting a Aortopulmonary window (Type I defect)

144