TOTAL ANOMALOUS PULMONARY VENOUS RETURN IN AN OLDER CHILD PRESENTING AS EFFORT INTOLERANCE : CASE REPORT

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Abstract

Background : Total Anomalous Pulmonary Venous Return (TAPVR) is a rare congenital cyanotic heart disease. Which typically presents during infancy, late diagnosis in older children is unusual. The clinical manifestation of TAPVR depend on the presence or absence of obstruction of the venous channels. If pulmonary venous return is obstructed, severe pulmonary congestion and pulmonary hypertension develops, presents early with cyanosis and respiratory distress. In contrast, those with mild or no obstruction develops mild to moderate degrees of desaturation following fall in pulmonary vascular resistant.

Case Report : We present a case of Total Anomalous Pulmonary Venous Return (TAPVR) with large secundum Atrial Septal Defect (ASD) in a 10 years old boy, presented with mild and nonspecific clinical manifestations might be responsible for the delayed diagnosis. Here is contemporary reminder for taking careful history, focused clinical examination, evaluation by imaging during the assessment of pediatric patients, with prolonged effort intolerance and referral to pediatric cardiologist as required.

Conclusion : Most importantly this case illustrates the useful diagnostic value of CT scan of chest as an aid to Echocardiography to evaluate extracardiac vascular morphology.

Key wards : Total Anomalous Pulmonary Venous Return (TAPVR); Effort intolerance; CT scan of chest.

Introduction

Congenital Heart Disease (CHD) occurs in approximately 0.8% of the live births. TAPVR constitutes 1-2% of all lesions.[1] Few patients with TAPVR survive without surgical correction beyond the first decade.[2] In TAPVR all the pulmonary veins instead of joining the left atrium are connected anomalously to result in the total pulmonary venous blood reaching the right atrium. According to the site of connection it may be supracardiac (50%) cardiac (25%) infracardiac (20%) & mixed (5%). Among them percentage with significant obstruction is highest in infracardiac, about 95-100%.[3]

All forms of TAPVR involve mixing of oxygenated & deoxygenated blood before or at the level of the right atrium. The clinical manifestation of TAPVR depend on the presence or absence of obstruction of the venous channels. If pulmonary venous return is obstructed, severe pulmonary congestion and pulmonary hypertension develops, presents early with cyanosis and respiratory distress. In contrast, those with mild or no obstruction develops mild to moderate degrees of desaturation following fall in pulmonary vascular resistance.

This case report is important because it highlights that TAPVR can present late with mild and indistinguishable clinical manifestations.

Case Report

A 10 year old boy, immunized, 3rd issue of a nonconsanguineous parents, hailing from Cox's Bazar was admitted into Chittagong Medical College Hospital (CMCH) on 8th July, 2020 with the complaints of high grade intermittent fever, respiratory distress & cough for 5 days. His other complaints were exertional dyspnea & progressively worsening vertigo for last 7 months and not growing well since birth. He suffered recurrent pneumonia like symptoms since his early childhood but never got admitted into hospital. Recently he was unable to go outside and play because of marked effort intolerance increasing markedly over days, which was evident to the parents since 7 years of his age. He had no history of convulsion, unconsciousness or syncopal attack. He was delivered at term at home by NVD with uneventful perinatal period. His development was age appropriate. During admission, he was dyspnoeic, temperature was 101° F,

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R/R-35 breaths/min, HR-110 beat/min, BP-100/60mmHg. He was underweight, weight was 18 kg & height was 123cm. Weight for age was -3.5 SD & height for age was -3.1 SD.BMI was 12 kg/m2 , -3.3 SD. Positive systemic findings were, a non-radiating grade II pansystolic murmur at the left lower sternal border vesicular breath sound with bilateral crepitations and baseline investigation findings were neutrophilic leukocytosis, raised ESR & CRP. RT PCR for COVID-19 was negative, CXR shown moderate enlargement of cardiac shadow and increased pulmonary vascular markings (Figure 1).

Initial 2D echo (As probe of color Doppler echo for pediatric population is not available in the admission centre) was done and it could point out pulmonary hypertension (Figure 2). Thereafter CT scan of chest suggested TAPVR, findings were grossly dilated all four pulmonary veins. It was also evident that the left upper lobe pulmonary vein drains into brachiocephalic vein and right upper lobe pulmonary vein drains into the Superior Vena Cava (SVC). Both the venous channels, SVC and brachiocephalic vein were grossly enlarged. Right atrium and right ventricle were dilated and left sided chambers were relatively smaller in size. Aorta was inconspicuous, Pulmonary trunk and main pulmonary artery were mildly dilated (Figure 3). Eventualy Color Doppler Echocardigraph done by Pediatric Cardiologist and impression was Supra cardiac Total Anomalous Pulmonary Venous Return (TAPVR) with large secundum Atrial Septal Defect (ASD) with Moderate pulmonary hypertension (Figure 4).

He is now on diuretics, clinically stable and advised to do a surgery as a definitive treatment.

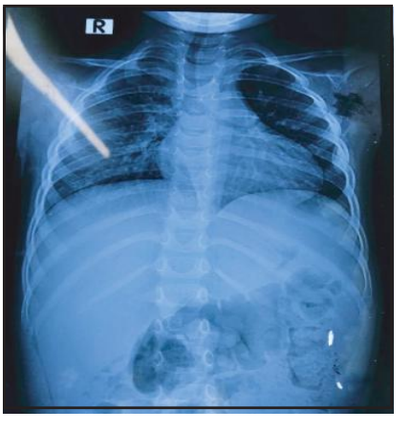

Fig 1 : CXR showing cardiomegaly with increased pulmonary vascular Markings.

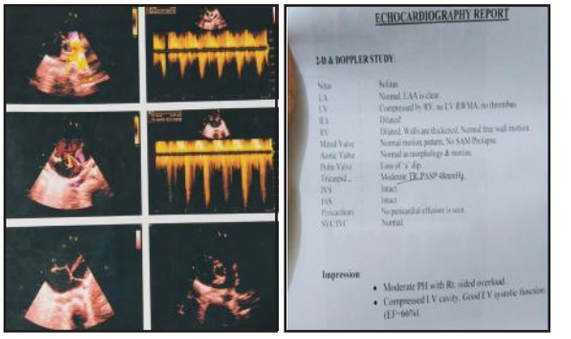

Fig 2 : 2D Echocardiography showing moderate pulmonary HTN.

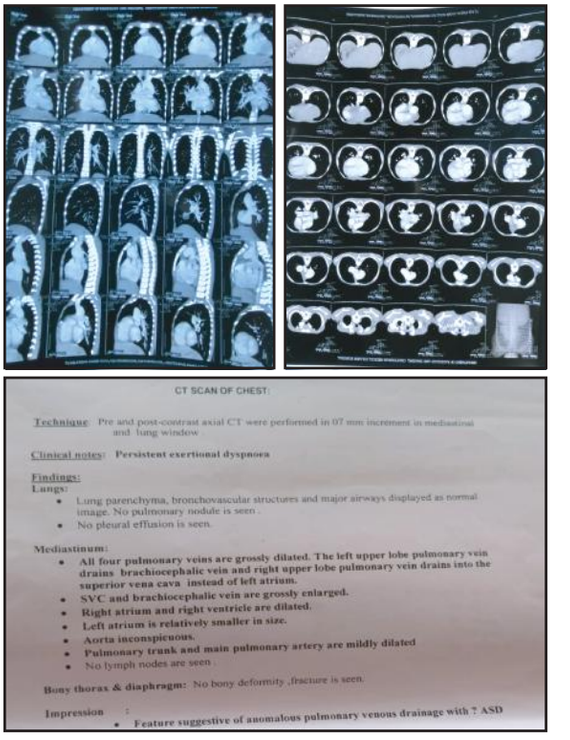

Fig 3 : CT scan of chest showing features suggestive of TAPVR with ASD.

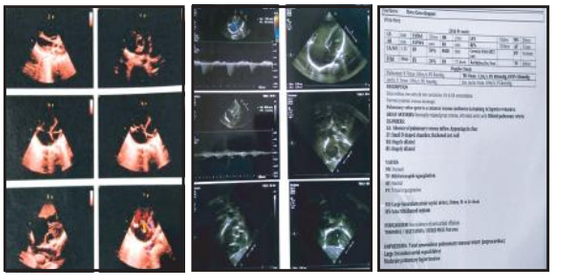

Fig 4 : Colour doppler echocardiogphy showing TAPVR (Supracardiac) large ASD with moderate pulmonary HTN.

Discussion

TAPVR is an uncommon cyanotic heart disease typically diagnosed in the newborn period. TAPVR may be one of several varieties. Pulmonary venous obstruction is possible with any variety, but more common with infra diaphragmatic TAPVR. An atrial shunt is essential for survival. Clinical presentation depends on the presence or absence of obstruction to pulmonary venous return. Patients with obstructed TAPVR are extremely ill soon after birth, with severe cyanosis, and may have low cardiac output and respiratory distress. Patients with TAPVR without pulmonary venous obstruction are often asymptomatic at birth, with difficult to detect cyanosis, but will usually present within the first year of life with tachypnoea, feeding difficulties, failure to thrive or repeated respiratory infections which manifestations match with the reported case.

However, Sachin Talwar in India described TAPVC in 98 patients older than ten years at the time of presentation and Xu-hua Jian in China reported 50% in 12 cases presented with total anomalous pulmonary venous connection.[4,5]

Margaret M Samyn reported a case of late diagnosis in 4.5 year old girl with TAPVR & large ostium secundum ASD, presented with persistent decrease exercise tolerance & fatigue.[6]

While patient in this case report presented with very indistinguish able features during preschool years. But as the age progresses he needed increased frequency of taking care during the last couple of years prior to diagnosis because of extreme fatigue. Parental concern on the extreme fatigue and then eventual recognition of effort intolerance, presence of pulmonary hypertension on 2 D echocardiogram prompted to do CT scan of chest, which warranted further evaluation by experts Pediatric Cardiology.

Echocardiography as the first and safest imaging modality for cardiovascular abnormalities may fail in complete depiction of some complex feature of TAPVC.[7] CT angiography is then a noninvasive and sensitive choice for mapping the PVs without need for invasive cardiac catheterization.[8,9]

Contrast-enhanced Magnetic Resonance (MR) angiography is the radiation-free alternative for CT angiography but needs general anesthesia in younger children.[7]

In a case report, Masaki Ogawa et al suggested that electrocardiography-gated CT and Phase-Contrast Magnetic Resonance Imaging (PC-MRI) are more useful than non–electrocardiography-gated CT for differentiating the vertical vein in TAPVC from persistent left superior vena cava in adult patients.[10]

With the advent of neonatal pulse oximetry testing prior to hospital discharge, rare cyanotic heart diseases like this may be discovered earlier. TAPVR can, as in our case, present in child or adulthood, in the setting of a widely patent atrial septum and no obstruction to pulmonary venous return; here, prognosis is better despite late repair.[11,12]

Limitation

We have limitation in resource personnel on pediatric cardiology.

Conclusion

Total anomalous pulmonary venous return is a rare CHD and mostly presents in the neonatal period. Our case is unusual by the fact that the patient is diagnosed late at his 10 years of age who presented with effort intolerance and repeated respiratory tract infection- 2 nonspecific manifestations. So it can affirm that supracardiac type of total pulmonary venous return can evolve until later part of first decade if the atrial septal defect is large which might have a good outcome after complete repair.

Recommendation

We want to recommend that neonatal screening should be practiced regarding congenital heart diseases in every center. All children with history of repeated respiratory tract infection, effort intolerance should be evaluated with close supervision

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Contribution of authors

RJ-Conception, citing references & final approval.
FC-Drafting, citing references & final approval.
AF-Design, drafting & final approval.
MD-Drafting, citing references & final approval.
MMM-Design, drafting & final approval.

AB-Comception, critical revision & final approval.
NUM-Design, critical revision & final approval.
RS-Conception, critical revision & final approval.

Disclosure

We have no conflicts of interest with the presented materials in this presentation.

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