Indian Journal of
Applied Radiology

Open Science Publications

Volume 5, Issue 1 - 2019

www.opensciencepublications.com

Complex Congenital Cardiac Anomalies and Complex Tapvc-A Case Report

Case Report

Vasudha N[1]*, Pramod N[2] and Jeetendra P[3]

[1]*Department of Anatomy, University Kasaba Bawada, India*

[2]*Department of Radiodiagnosis, D.Y.Patil Medical College, India*

[3]*Department of Radiodiagnosis, D.Y. Patil Medical College, India*

***Corresponding author:** Vasudha N, Professor Department of Anatomy, D. Y. Patil Medical College, DY Patil Education Society; Deemed to be University Kasaba Bawada, Kolhapur, Tel: 9665730990; E-mail: dr.vasudhanikam@gmail.com

Article Information: **Submission:** 07/05/2019; **Accepted:** 19/09/2019; **Published:** 23/09/2019

Abstract

Complex cardiac anomalies are the main cause of infant mortality. Early diagnosis and proper mode of treatment is essential in these cases. Screening of the patients detects the significant congenital heart defects. We present a case of complex cardiac anomalies with Atrial Septal Defect (ASD), Total Anomalous Pulmonary Venous Connection (TAPVC), dilated right atrial and right ventricular chambers, Patent Ductus Arteriosus (PDA), Persistent Left Sided Superior Vena Cava (PLSVC).

Keywords: Total anomalous pulmonary venous return; Diagnostic imaging; Congenital heart defects; Echocardiography, Patent ductus arteriosus; Persistent left superior vena cava

Introduction

Congenital heart disease affects almost 1 in 100 newborn babies worldwide [1]. Congenital heart defects remain the most common congenital anomalies in live births and are the main cause of infant mortality in the developed world. Congenital heart disease is defect in heart and major blood vessels including structural, chromosomal, genetic, biomechanical defects and malformations [2]. Patients with CHD frequently suffer from broad spectrum of subsequent neurological deficits, including motor, cognitive, behavioural, social and attention abnormalities [3]. Congenital cardiovascular malformations present some of the most interesting and difficult challenges in medicine and the repair of heart defects requires advanced technological interventions and they are among the most costly defects to manage [4].

Congenital heart defects are anatomically, clinically, epidemiological and developmentally heterogenous. There are eight major malformations, and these are

1. Conotruncal
2. Atrioventricular Septal Defect (AVSD)
3. Total Anomalous Pulmonary Venous Return (TAPVR)
4. Left Ventricular Outflow Tract Obstruction (LVOTO)
5. Right Ventricular Outflow Tract Obstruction (RVOTO)
6. Septal
7. Heterotaxy
8. Complex [5].

Here we report a case of 6 months old baby girl with complex cardia anomalies.

Case Report

A 6 months old baby girl was referred for screening by paediatrician. On examination, the patient had dyspnoea, fever, and mild cyanosis. There was history of recurrent chest infection, low oxygen saturation and frequently required oxygen supplementation. On auscultation, there was continuous machinery murmur. The patient was advised for X-ray chest, 2D echocardiography and CT pulmonary angiography.

X-ray chest revealed pulmonary plethora. There was enlargement of blood vessels in both the lungs. There was right upper lobe consolidation and mild pneumonia was present. There was mild cardiomegaly that was suggestive of septal defect (Figure 1). 2D echocardiography revealed large ASD with abnormal pulmonary venous draining and PDA. There was dilatation of right atrial and right ventricular chambers with severe tricuspid regurgitation due to right heart volume overload.

CT Pulmonary angiography revealed following findings;

1. Large ASD- (Figure 2).
2. Dilated RA and RV.
3. Supracardiac type TAPVC (Figure 3).
4. Dilated SVC (Figure 4).
5. PDA (Figure 5).
6. Left sided persistent superior vena cava draining into coronary sinus (Figure 6).

All these findings are suggestive of complex TAPVC with multiple cardiac anomalies.

Discussion

Patients with CHD frequently suffer from a broad spectrum of neurological deficits including motor, cognitive, behavioural, social and attention abnormalities [3]. In our case, the baby inspite of suffering from such complex cardiac anomalies there was no neurological deficits seen.

The cardiac pathology in patients with TAPVC shows interatrial septal defects, cardiomegaly, prominent pulmonary trunk, increased pulmonary vascularity on x-ray, low oxygen saturation ranging from 80% to 90% on oximeter. There is right atrial enlargement with right ventricular hypertrophy [6]. Clinically patients with TAPVC may have congestive cardiac failure or profound cyanosis due to interatrial communication and patency of pulmonary venous flow [7]. However, in our case, complex cardiac anomalies were present; there was RA and RV dilatation, low oxygen saturation and mild cyanosis was present. There were no signs of congestive cardiac failure.

TAPVC is not a problem during fetal development due to high pulmonary vascular resistance and shunting blood flow through foramen ovale. However, the heart of patient with TAPVC has no direct pulmonary connection to the left atrium; thus, severe pulmonary congestion and pulmonary hypertension can occur after closing the ductus arteriosus [8]. However, in our case pulmonary hypertension was not revealed as the patient had PDA.

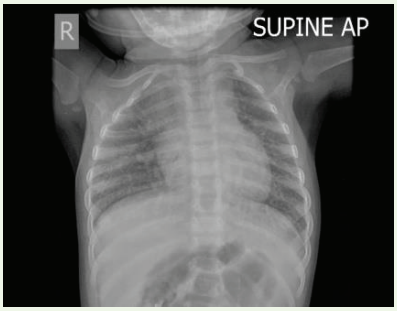

Figure 1: X-ray chest showing mild increased cardiac silhouette with pulmonary plethora.

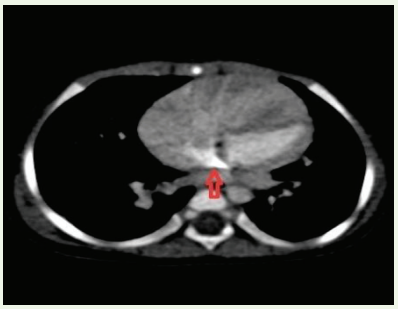

Figure 2: Post contrast axial image showing contrast through left sided svc opacifying the right atrium suggestive of ASD (red arrow).

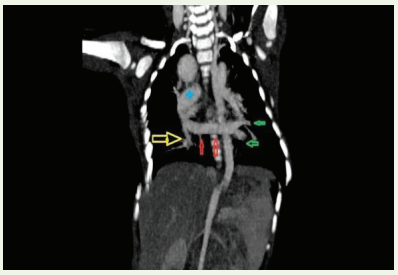

Figure 3: MPR image showing left sided pulmonary veins (green arrows) joining to form a common trunk (red arrows) which crosses the midline and joined by right sided pulmonary vein (yellow arrow). The common trunk (blue star) ascends upwards and ultimately opens at svc and right atrium junction suggestive of TAPVC. Note dilated SVC just above the blue star.

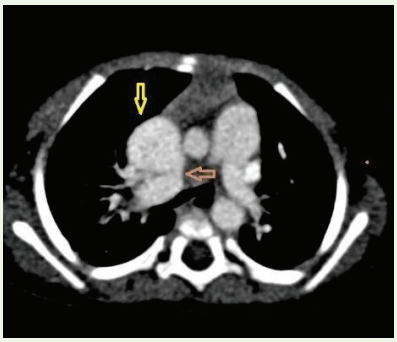

Figure 4: Post contrast axial image showing common channel opening into SVC brown arrow. Yellow arrow demonstrates dilated SVC.

Citation: Vasudha N, Pramod N, Jeetendra P. Complex Congenital Cardiac Anomalies and Complex Tapvc-A Case Report. Indian J Appl Radiol. 2019;5(1): 140.

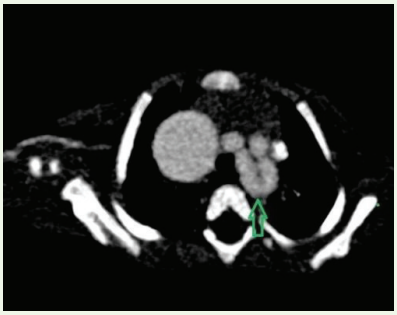

Figure 5: Persistent communication between proximal descending aorta and left pulmonary vein suggestive of PDA (green arrow).

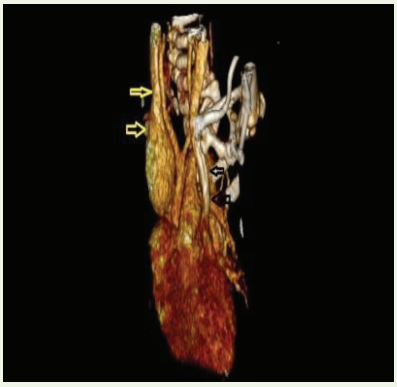

Figure 6: VRT image showing persistent left sided SVC (black arrow) and dilated right svc (orange arrow).

Four types of TAPVC are classified based on the location of pulmonary venous drainage these are:

a) Supracardiac

b) Cardiac

c) Infracardiac

d) Mixed

a) Supracardiac (Type-I TAPVC) is most common, accounting for approximately 44% of cases. In this type, drainage most commonly occurs via a vertical vein to the left brachiocephalic vein. Rarely, supracardiac TAPVC may drain directly to the right SVC, left SVC or azygous system.

Most commonly in supracardiac type the pulmonary veins drain to a confluence posterior to the left atrium. An ascending vertical vein originates from this confluence and travels behind the left atrial appendage and usually, anterior to the left pulmonary artery. However occasionally this vein may travel posterior to the left pulmonary artery and may become trapped in a "vice" between the dilated artery and left bronchus, leading to pulmonary venous obstruction…the vertical vein finally drains into the innominate vein. Occasionally the site of entrance to the innominate vein may also be narrowed, leading to obstruction. The innominate vein and SVC are dilated. The right heart is usually dilated due to the volume overload.

b) Cardiac (Type-II TAPVC) which represents approximately 21% of TAPVC cases, pulmonary veins drains either to the coronary sinus or directly into the right atrium. the pulmonary veins and coronary sinus are significantly dilated and echocardiography shows a characteristic "whale" tail appearance. Obstruction is unusual in this form of TAPVR.

c) Infracardiac (Type-III TAPVC) represents approximately 26% of cases of TAPVC and drains below the diaphragm to a systemic vein- the IVC, a hepatic vein, or the azygous system, or the portal venous system. The draining pulmonary veins in this type of TAPVC may be obstructed, frequently at the level of diaphragm, because of extrinsic narrowing, resulting in pulmonary edema with a normal size cardiac silhouette on chest radiography. In the infra cardiac type of TAPVR, the pulmonary venous confluence is usually posterior to the left atrium and vertically oriented. From here, a descending vein arises and passes through the oesophageal hiatus. Although pulmonary venous obstruction may occur in any type of TAPVC, it is most common in the infra cardiac form being represented in upto 78% of patients.

d) Mixed (Type-IV TAPVC) the final type of TAPVC is diagnosed when the location of pulmonary vein drainage is mixed. In this form pulmonary veins drain to at least two different locations, including a brachiocephalic vein, SVC, azygous vein, coronary sinus, right atrium or below the diaphragm. This type of pulmonary venous drainage accounts for approximately 9% of TAPVC cases. An example includes connection of the right pulmonary veins to the coronary sinus and connection of the left pulmonary veins to the innominate vein [7,9]

Although obstruction can occur in any type of TAPVC, it is most commonly encountered in the infra cardiac type. There are various complications of obstructed TAPVC leading to hypoxemia, pulmonary hypertension and pulmonary vascular obstructive disease [8,10]. However, our case represents supracardiac type of TAPVC with low O_2 saturation, mild cyanosis but no signs of pulmonary hypertension as the patient had PDA.

Embryologically TAPVC results from early atresia or failure of common pulmonary veins to develop, with persistence of at least one connection to the cardinal or the umbilical vitelline venous system. TAPVC is a cause of neonatal cyanosis and can result in rapid death when blood is not shunted from right side of heart to the left side. Isolated TAPVC is diagnosed if the patient has ASD, PDA or both. Complex TAPVC is diagnosed if the patient has other intracardiac lesions in addition to ASD or PDA [10].

Our case represents complex TAPVC supracardiac type as along with ASD, PDA other associated anomalies, which were seen are persistent left sided superior vena cava draining in dilated coronary sinus.

Persistent left sided superior vena cava is one of the most frequent anomalies of systemic venous circulation in 0.3-0.5% of general population and in 4.3% of those with CHD. In most patients with persistent left superior vena cava, a right superior vena cava is also present, and both normally drain into right atrium through dilated coronary sinus [11,12]. In our case there was persistent left superior vena cava draining into dilated coronary sinus as well as dilated right superior vena cava was present.

Citation: Vasudha N, Pramod N, Jeetendra P. Complex Congenital Cardiac Anomalies and Complex Tapvc-A Case Report. Indian J Appl Radiol. 2019;5(1): 140.

Thus, in the patients with CHD and complex TAPVC correct delineation of anatomy and associated cardiovascular anomalies is crucial while planning the treatment. Hence, the baby was referred to cardiologist for further management and treatment.

Conclusion

With improvement in imaging techniques, it is possible to make accurate diagnosis of complex cardiac anomalies. 2D echocardiography and CT pulmonary angiography imaging is effective in the diagnosis of complex TAPVC. This case study is a contribution to the knowledge of complex cardiac anomalies.

Acknowledgement

Eureka Diagnostic Centre; Kolhapur and DY Patil Education Society supported this work; Deemed to be University; D.Y. Patil Medical College; Kolhapur. Thanks to Chancellor, Vice Chancellor, Pro Vice Chancellor, Registrar and Dean of DY Patil Education Society's Deemed to be University and D.Y. Patil Medical College, Kolhapur. Thanks to Dr. Pramod Nagure, Dr Jeetendra Patil and my colleagues of Department of Anatomy for their support.

References

1. Listijino DR, Rubens MB, Rigby ML (2014) Complementary use of imaging modalities in diagnosis of complex congenital heart disease. Dave Asian Heart J 22: 55-59.
2. Singh Y, McGeoch L (2016) Fetal anomaly screening for detection of congenital heart defects. J Neonate Biol 5: 100-115.
3. Morton PD, Ishibashi N, onas RA, Gallo V (2015) Congenital cardiac anomalies and white matter injury. Trends Neurosci 38: 353-363.
4. Lalani SR, Belmont JW (2014) Genetic basis of congenital cardiovascular malformations. Eur J Med Genet 57: 402-413.
5. Botto L D, Mulinare J, Erickson JD (2003) Do Multivitamin or Folic Acid supplements reduce the risk for congenital heart defects? Evidence and gaps. Am J Med Genet A 12: 95-101.
6. Meng-Luen L, Mei-Hwan W, Jou-Kou W, Hung-Chi L (2014) Echocardiographic assesment of total anomalous pulmonary venous connections. Medicine (Baltimore) 100: 729-735.
7. Dilman JR, Yarram SG, Hernandez RJ (2009) Imaging of Pulmonary venous Developmental Anomalies. AJR Am J Roentgenol 192: 1272-1285.
8. Meyer RA (1977) Total anomalous pulmonary venous return. In: Meyer RA (Eds). Paediatric Echocardiography; Philadelphia: Lea and Febiger 113-125.
9. Vyas H, Greenberg SB, Krishmamurthy R (2011) MR Imaging and CT evaluation of congenital pulmonary vein abnormalities in neonates and infants. Radiographics 32: 87-98.
10. Kim HS, Jeong K, Cho HJ, Choi YE, Ma JS, et al. (2014) Total Anomalous Pulmonary Venous Return in Siblings. J Cardiovasc Ultrasound 22: 213-219.
11. Turkvatan A, Tola HT, Ayyidiz P, Ozturk E, Ergul Y, et al. (2017) Total Anomalous Pulmonary Venous Connection in children: Preoperative evaluation with low-dose multidetector computed tomographic angiography. Tex Heart Inst J 44: 120-126.
12. Moorthy N, Kapoor A, Kumar S (2013) Isolated persistent left sided superior vena cava, giant coronary sinus, atrial tachycardia and heart failure in a child. Indian Heart J 65: 603-606.

Citation: Vasudha N, Pramod N, Jeetendra P. Complex Congenital Cardiac Anomalies and Complex Tapvc-A Case Report. Indian J Appl Radiol. 2019;5(1): 140.