International Journal of Biomedical Research

ISSN: 0976-9633 (Online)
Journal DOI:10.7439/ijbr
CODEN:IJBRFA

Case Report

Rare case of Pregnancy with Total Anomalous Pulmonary Venous Connection (TAPVC)

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital malformation of the heart, in which all four pulmonary veins drain abnormally to the right atrium by way of an abnormal (anomalous) connection. It is a relatively uncommon congenital defect representing approximately 2% of all congenital heart anomalies. It is found in approximately 7 persons in 100,000 populations. It is associated with atrial septal defect, making it compatible to life. Pregnancy associated with uncorrected TAPVC is extremely rare. It is a high risk pregnancy. The outcome depends upon type of TAPVC and cardiac functional capacity. There is high risk of cardiac failure and cardiac arrhythmias during pregnancy. Medical termination of pregnancy is advised at early gestation to prevent complications.

Keywords: Pregnancy, TAPVC, atrial septal defect

1. Introduction

TAPVC is a rare congenital malformation of the heart, in which all four pulmonary veins drain abnormally to the right atrium by way of an abnormal (anomalous) connection. It is a relatively uncommon congenital defect representing approximately 2% of all congenital heart anomalies.[1]It is found in approximately 7 persons in 100,000 populations. It is associated with atrial septal defect; making it compatible to life.[2] Pregnancy associated with uncorrected TAPVC is extremely rare. It is a high risk pregnancy. The outcome depends upon type of TAPVC and cardiac functional capacity. There is high risk of cardiac failure and cardiac arrhythmias during pregnancy. Medical termination of pregnancy is advised at early gestation to prevent complications.

2. Case report

Nineteen year old primigravida, unregistered case, reported to Gynaecology department, as a referred case of heart disease from nearby hospital for termination of pregnancy. She was a diagnosed case of atrial septal defect in childhood and was on penicillin prophylaxis till the age of five years. She belonged to lower socioeconomic class and was educated till 9th class. Neither woman nor her parents were aware about the details of cardiac lesion. She had frequent episodes of respiratory infections, breathlessness and fever in the childhood, for which she used to be hospitalized. There was no history of congenital heart disease in other members of family.

She got married one year back and conceived immediately after marriage. She was comfortable in the first month of pregnancy and had minimal nausea. From second month of pregnancy onwards, she developed breathlessness, cough, palpitations and occasional chest pain. She was unable to carry out her regular household activities. She used to feel fatigue during her regular home chores. Her cough at times increased at night which made it difficult for her to breathe suggestive of paraoxysmal nocturnal dyspnea. She had no antenatal checkups in first trimester of pregnancy. She developed severe breathlessness, cough and palpitations in third month of pregnancy for which she was taken to a cardiologist. She had undergone echo-cardiography and then diagnosed to have "Total Anomalous Pulmonary Venous Connection –Supra-cardiac type with large sized ostium secondum (ASD) with right to left shunt with severe pulmonary hypertension with trivial tricuspid regurgitation with mitral regurgitation". Echocardiography also revealed abnormal pulmonary venous confluence (PVC) of 15 milimeter at the site of entry of superior vena cava with good bi ventricular systolic function with left ventricular ejection fraction of 65 %. She was advised to take tablet Frusilac once a day and tablet Digoxin 0.25 mg per day for five days in a week. Cardiologist gave her advice for termination of pregnancy and corrective cardiac surgery after that.

On examination, she was a thin built woman, apprehensive, afebrile with pulse rate of 100 beats per minute, regular. Her respiratory rate was 23 cycles per minute and blood pressure was 130 /72 mm Hg. Her cardiac examination revealed pan-systolic murmur in pulmonary area. Respiratory system examination revealed occasional rhonchi and crepitations on auscultation. Per abdominal examination showed 16 weeks size of pregnant uterus with live fetus. Obstetric ultrasound confirmed the fetal maturity of 16 weeks with fundal position of placenta, adequate liquor and variable fetal position.

Her ECG findings were suggestive of sinus rhythm with heart rate of 100/min, PR interval of 0.20 sec and right ventricular hypertrophy in V 1- V 6 segment. Her blood profile revealed - Hb- 13.9grams/dl, PCV 40%, TLC 21,400/cumm, Meta bands- 11%, neutrophils -80 %, lymphocytes- 5% monocytes -2%, platelet count -3 lakhs. Peripheral blood smear gave normocytic normochromic picture and neutrophilic leucocytosis , prothrombin time test -10.4 control 10.1, aptt test -40 seconds, control -30 seconds , INR 1.28. Her Blood Urea - 13 mg/dl, S. Creatinine - 0.9 mg/dl , S.Na -137meq/lit , S. K -3.7 meq/lit, S. Bilirubin- Total- 0.8mg/dl , Conjugated- 0.22mg/dl , SGOT -24.7 IU, SGPT -19.9 IU, S. Alkaline Phosphatase 179. Serum Uric acid 2.7 , Blood Proteins - 6.3gm %, Albumin 3.4gm % , Globulin 2.9gm % , Plasma glucose random -74 mg/dl. Serum LDH- 410 IU/L, Blood group and Rh type -O positive.

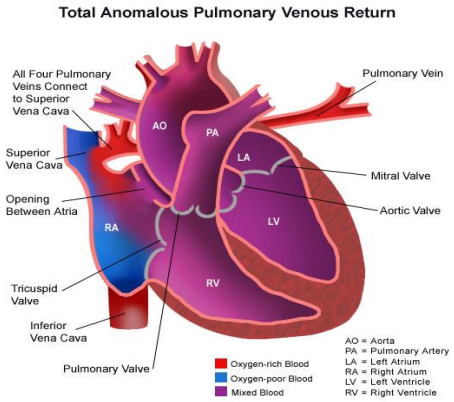
Relatives were explained about the condition of the patient and the risk involved in medical termination of pregnancy. High risk consent was obtained. Physician opinion was sought. She was given prophylactic antibiotics in the form of Inj. Ampicillin 500 mg four times a day, Inj. Gentamycin 80 mg twice a day and Inj. Metrogyl 500 mg three a day by intravenous route. Pregnancy termination was done with extra amniotic ethacridine lactate solution. She was closely monitored for signs of cardiac failure. She was jointly managed by Obstetrician and Physician. She had febrile morbidity and cardiac instability twelve hour after instillation of ethacridine lactate solution. It was managed with administration of injectable paracetamol. She had complete abortion after induction abortion interval of 41 hours.

She was advised to consult cardiologist for corrective surgery and was discharged from hospital after seven days of parenteral antibiotics. She was haemodynamically stable at the time of discharge. Couple was counseled regarding need for taking regular medication and cardiac surgery. Couple was advised to avoid pregnancy till surgical correction of cardiac anomaly. Different contraceptive options were suggested.

3. Discussion

Total anomalous pulmonary venous connection (TAPVR) is a rare form of cyanotic congenital heart disease. It is characterized by abnormal communication of pulmonary veins to the right atrium via the superior vena cava. In this type of TAPVR, the pulmonary veins first come together behind the heart and then drain upwards to an abnormal "vertical vein." This vertical vein joins the innominate vein which connects to the superior vena cava and drains to the right atrium. **(Figure- 1)**

Fig.1- Diagram showing Total Anomalous Pulmonary Venous Connection (TAPVC)



The cause of total anomalous pulmonary venous return (TAPVR) is unknown.[1] It constitutes between 1% and 1.5% of all children with congenital heart disease and can be categorized by the site of drainage into the systemic circulation (supracardiac, 45%; infracardiac, 25%; cardiac, 25%; mixed, 5%). All types of total anomalous pulmonary venous return have to have an associated atrial septal defect (ASD).[2] As none of the pulmonary veins connect to the left side of the heart, blood is shunted from the right atrium across the atrial septal defect. Absence of an atrial septal defect in total anomalous pulmonary venous return is not compatible with survival.

TAPVC usually presents in infancy. The presentation depends on the site of drainage of the pulmonary veins and whether or not the veins are obstructed. The infant may appear very sick and may have the symptoms like bluish colour of the skin (cyanosis), frequent respiratory infections, lethargy, poor feeding, poor growth, rapid breathing. Sometimes, condition may remain asymptomatic in infancy or early childhood.[1]

Clinical signs suggestive of TAPVC are right ventricular heave, Loud S1, fixed split S2,S3 gallop, systolic ejection murmur at left upper sternal border, cardiomegaly. Cardiac catheterization can confirm the diagnosis by showing that the blood vessels are abnormally attached. ECG shows enlargement of the ventricles (ventricular hypertrophy) and right axis deviation. Echocardiogram may show that the pulmonary vessels are attached. MRI of the heart can show the connections between the pulmonary vessels. X-ray of the chest shows a normal to small heart with fluid in the lungs and Snowman sign or figure of 8 configurations` on chest radiograph.[3,4,5,6]

Surgery to repair the abnormal vascular communication is needed as soon as the diagnosis is made in childhood. In surgery, the pulmonary veins are connected to the left atrium and the defect between the right and left atrium is closed. If this condition is not treated, patient can develop heart failure or arrhythmias. Early repair of the defect provides excellent results, if there is no obstruction to the pulmonary veins at the new connection into the heart.[7]

Pregnancy associated with uncorrected TAPVC is extremely rare. Physiological changes during pregnancy, especially the hemodynamic changes in the form of increase in blood volume and heart rate are not tolerated by the abnormal condition (TAPVC) of the heart. The pregnant woman is advised for termination of pregnancy. Present case came to our hospital in second trimester of pregnancy with pulmonary hypertension. Extra amniotic instillation of Ethacridine lactate was considered to be the safest method for pregnancy termination. Patient could tolerate the stress of abortion process and did not show signs of failure. She was discharged from hospital with advice for repair of cardiac lesion before having next pregnancy.

References

1. Webb GD, Smallhorn JF, Therrien J, Redington AN. Congenital heart disease. In: Bonow RO, Mann DL, Zipes DP, Libby P, eds. Braunwald's *Heart Disease: A Textbook of Cardiovascular Medicine*, 9th ed. Philadelphia, Pa: Saunders Elsevier; 2011: chap 65.
2. Darling RC, Rothney WB, Craig JM: Total pulmonary venous drainage into the right side of the heart: report of 17 autopsied cases not associated with other major cardiovascular anomalies. *Lab Invest.*1957; 6:44-55.
3. Sears EH, Aliotta JM, Klinger JR. Partial anomalous pulmonary venous return presenting with adult-onset pulmonary hypertension. *Pulm Circ* 2012; 2:250-5.
4. Ho ML, Bhalla S, Bierhals A, Gutierrez F. MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. *J Thorac Imaging* 2009; 24:89-95.
5. Prasad SK, Soukias N, Hornung T, Khan M, Pennell DJ, Gatzoulis MA,*et al*. Role of magnetic resonance angiography in the diagnosis of major aortopulmonary collateral arteries and partial anomalous pulmonary venous drainage. *Circulation* 2004; 109:207-14.
6. Ammash NM, Seward JB, Warnes CA, Connolly HM, O'Leary PW, Danielson GK (May 1997). "Partial anomalous pulmonary venous connection: diagnosis by transesophageal echocardiography". *J. Am. Coll. Cardiol.* 29 (6): 1351–8. doi:10.1016/S0735-1097(97)82758-1. PMID 9137235.
7. Kanter KR. Surgical Repair of Total Anomalous Pulmonary Venous Connection. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann* 9:40-44 © 2006 Elsevier.