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Case Report

A conception with congenitally complicated connection: a rare case of pregnancy with total anomalous pulmonary venous connection

Priyadarsene P., Mahalakshmi M.*, Sangeetha R.

Department of Obstetrics and Gynecology, Madras Medical College, Chennai, Tamil Nadu, India

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***Correspondence:**
Dr. Mahalakshmi M.,
E-mail: drmahalakshmi1007@gmail.com

ABSTRACT

Total anomalous pulmonary venous connection (TAPVC) is a congenital cyanotic heart disease characterized by abnormal communication of pulmonary veins into the right atrium rather than into the left atrium leading to mixture of oxygenated and deoxygenated blood. Uncorrected TAPVCs do not survive till adulthood and pregnancy in patients with TAPVC is extremely rare. In patients with TAPVC, the heart cannot withstand the hemodynamic changes associated with pregnancy and hence termination of pregnancy in the first trimester is often required. Here we reported an extremely rare case of pregnancy in a patient with uncorrected TAPVC, terminated successfully in our institute without any complication.

Keywords: TAPVC, Congenital cyanotic heart disease

INTRODUCTION

The ever-evolving medical innovation has changed the trend of pregnancies with heart diseases in the direction of increasing cases of pregnancies with congenital heart diseases uncovering more new challenges in managing patients with heart diseases who would not otherwise survive into adulthood and rare so, become pregnant.

Total anomalous pulmonary venous connection (TAPVC) is a congenital cyanotic heart disease characterized by abnormal communication of pulmonary veins into the right atrium rather than into the left atrium leading to mixture of oxygenated and deoxygenated blood. The reported incidence ranges from 0.4 to 2%, which represents approximately seven persons in 1 lakh population.[1]

The probability of survival into adulthood is less than 7% making pregnancy with TAPVC an utmost rarity unless the condition is corrected in childhood.[2] However, in patients with TAPVC, the heart cannot withstand the hemodynamic changes associated with pregnancy and hence termination of pregnancy in the first trimester is often required.

CASE REPORT

A 30 years primigravida, a known case of supracardiac TAPVC with severe pulmonary hypertension visited our OPD with positive urine pregnancy test at 5 weeks of gestation.

Our patient had breathlessness on exertion (NYHA CLASS II) from childhood and recurrent respiratory tract infection for which she was evaluated and diagnosed at the age of 17 to have supracardiac type TAPVC with severe pulmonary hypertension. She was since then started on oral sildenafil for pulmonary hypertension. Owing to socioeconomic condition the patient did not have regular cardiac follow up and was irregularly compliant with the medication.

She was thin built with a body mass index of 17.8, comfortable at rest, not dyspneic or tachypneic, afebrile with pan digital clubbing and central cyanosis. She was not pale and had no pedal edema. Her vitals were pulse rate of 90/min, blood pressure of 90/70 mmhg in the right arm and a saturation of 82% in right upper limb. Her jugular pulsations were normal. Her lungs were clear on auscultation.

Chest radiography revealed cardiomegaly with widening of the superior mediastinum and a classic snowman appearance, consistent with supracardiac TAPVC. ECG showed a normal sinus rhythm of 88/min with right axis deviation. Hemoglobin was 21.3, PCV 63 in alignment with the cyanotic disease and her renal function tests, liver function tests, thyroid profile and coagulation profile were within normal limits.

Viability scan showed a single live intrauterine gestation corresponding to 6 weeks. Echocardiography confirmed supracardiac type TAPVC, with an atrial septal defect with right to left shunting, a dilated right ventricle with normal systolic function, severe pulmonary hypertension, RVH >12 mm, TAPSE 13 mm. The risk was stratified as modified WHO class IV of pregnancy risk classification of women with preexisting cardiovascular disease and pregnancy was advised to be terminated.

Extensive counselling of the mother and her relatives on grave prognosis of continuation of pregnancy was done and consent for termination of pregnancy was obtained.

Under antibiotic coverage, medical termination of pregnancy was initiated with administration of 400 mg oral mifepristone followed by 800 µg of misoprostol vaginally after 36 hours. Repeat doses of misoprostol (400 µg, 6th hourly) were administered until expulsion of products. However, due to persistent retained products of conception and risk of sepsis, suction evacuation was done under IV sedation after three repeat doses of misoprostol. The patient remained hemodynamically stable throughout the hospital stay and was discharged with advice to regular follow ups with cardiology team. The dangers of future pregnancy were cascaded and detailed counselling regarding use of proper contraceptive measures with a view of avoiding future pregnancies was given.

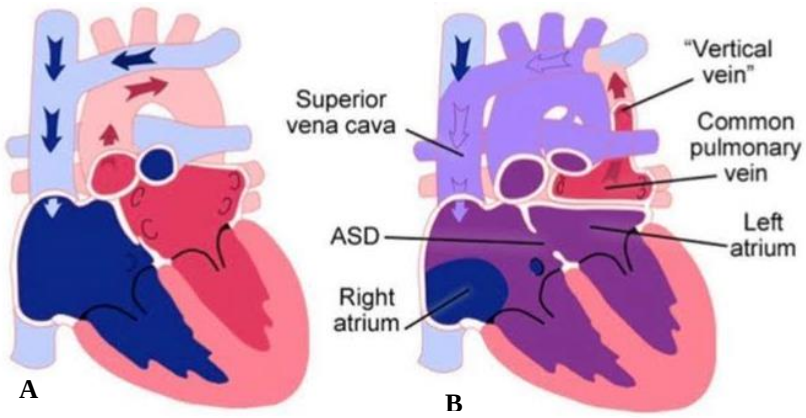

Figure 1: (A) Normal heart; and (B) total anomalous pulmonary venous connection.

DISCUSSION

The TAPVC connection is a rare congenital anomaly of the heart in which there is an absence of direct communication between the pulmonary veins and the left atrium, resulting in a mixture of oxygenated and deoxygenated blood in the right atrium. It is often associated with atrial septal defect or with transposition of great arteries, pulmonary atresia, truncus arteriosus, or single ventricle. Uncorrected TAPVCs do not survive till adulthood. Pregnancy in patients with TAPVC is extremely rare and often an indication for termination of pregnancy since hemodynamic changes in pregnancy, such as increased cardiac output and heart rate are not tolerated in patients with TAPVC. The unusual presentation of our patient is that there were only a few mild symptoms until her second decade of life and this shall be attributed to the large ASD and absence of pulmonary obstruction. However, presence of severe pulmonary hypertension puts her under WHO risk category class IV which precludes extremely high maternal mortality with more than 27% risk of cardiac events during pregnancy thereby contraindicating pregnancy promoting the need for termination of pregnancy and the same was followed through with this patient.

The safety of termination of pregnancy by medical or surgical methods in a case of TAPVC has not been elucidated given the paucity of cases. But there has been a similar report of termination of pregnancy in a case of TAPVC by medical method by Bangal et al.[3] However, both medical and surgical method proved safe in our patient.There has been one successful caesarean section delivery in a case of TAPVC delivering a 1.05 kg baby with good Apgar at 29 weeks of gestation by Kandasamy

et al where the patient with TAPVC had a large ASD and pulmonary stenosis thereby improving systemic circulation and minimal systemic symptoms and a successful pregnancy outcome.[4]

CONCLUSION

TAPVC with pregnancy becomes a very critical condition adding much to the morbidity and mortality of the pregnant mother and early termination of pregnancy is the present treatment option. However in TAPVC with near favourable cardiac physiology, a successful pregnancy outcome can be envisaged.

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