International Journal of Contemporary Pediatrics
Mane S. Int J Contemp Pediatr. 2022 Jun;9(6):630-631
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291
DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20221387

Case Report

Rare association of obstructed supracardiac total anomalous pulmonary venous connection with coarctation of aorta: case report

Shrikant Mane*

Department of Pediatrics, Government medical College, Miraj, Sangli India

Received: 10 April 2022
Accepted: 02 May 2022

***Correspondence:**
Dr. Shrikant Mane,
E-mail: shrikantmane17@gmail.com

ABSTRACT

Coexistence of TAPVC (total anomalous pulmonary venous connection) and coarctation of aorta rarely occurs. Newborn weighing 2.4 kg admitted in Sangli civil hospital NICU with respiratory distress with cyanosis. Chest X-ray of baby was suggestive of ground glass appearance of bilateral lung fields and cardiomegaly. SpO_2 of baby was fluctuating between 85 to 90% on nasal CPAP. 2D echo showed baby was having obstructed supracardiac TAPVC with severe discrete post-ductal coarctation of aorta. There was stretched PFO shunting R-L and small PDA shunting R-L. Baby was planned for emergency surgery but couldn't make it to operation theatre and succumbed within 30 hours of life. Till now there are very few case reports of TAPVC with coarctation but no case of obstructed supracardiac TAPVC with coarctation has been reported.

Keywords: Total anomalous pulmonary venous connection, Coarctation of aorta, Congenital heart disease

INTRODUCTION

Only 1.4% cases of TAPVC are reported to be associated with CoA.[1] In this patients lower body blood supply will be through saturated ductal flow. But if there is severe discrete coarctation with small closing PDA and obstructed supracardiac TAPVC, it forms bad subset.

CASE REPORT

Full term 2.4 kg newborn delivered by cesarean section admitted in NICU in view of respiratory distress and cyanosis. On examination baby was having tachycardia, tachypnea with distress and there was central cyanosis. Baby was kept on nasal CPAP with 60% FiO_2 but still not maintaining SpO_2 above 90%. Lower limb pulses were not very well palpable. X-ray showed significant pulmonary venous obstruction (Figure 1).

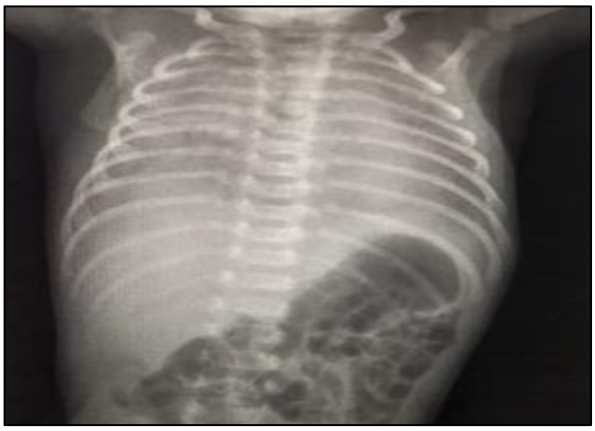

Figure 1: CXR showing cardiomegaly with B/L ground glass appearance.

Echocardiography showed obstructed supracardiac TAPVC (all 4 pulmonary veins draining from common chamber to left vertical vein to innominate to right SVC

to right atrium and the obstruction was at the level of left vertical vein), stretched PFO (R-L), severe discrete post ductal co-arctation with small PDA shunting R-L, dilated right atrium and right ventricle with moderate TR (max PG=60 mm of Hg) (Figure 2 and 3). Patient was intubated and started on PGE1 with dopamine. Baby was planned to shift for emergency TAPVC and coarctation repair on next day to nearby cardiovascular-thoracic surgery centre. Patient was having significant persistent metabolic derangements and lactic acidosis. Patient was having repeated episodes of bradycardia, desaturation inspite of full ventilatory and ionotropic supports. During such one episode baby couldn't be revived and succumbed to death.

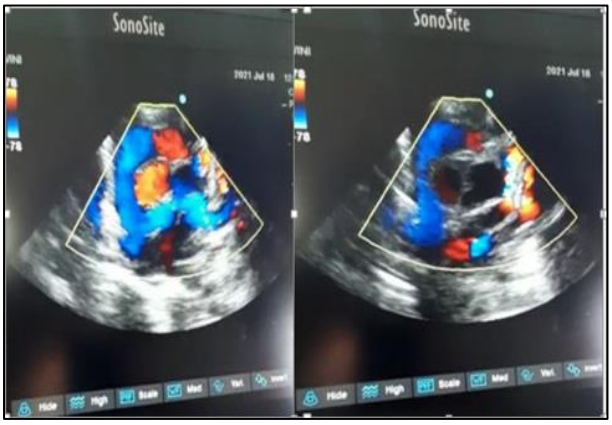

Figure 2: Suprasternal view on ECHO s/o all 4 pulmonary veins draining via obstructed vertical vein to innominate to right SVC.

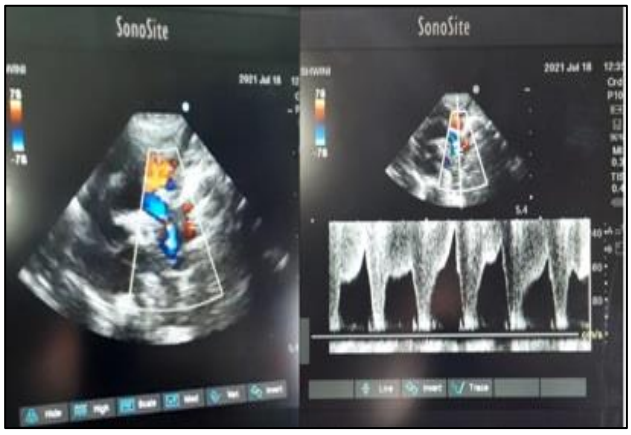

Figure 3: Suprasternal view on ECHO s/o narrowing of post ductal aortic area with flow acceleration across it. Doppler image of same s/o gradient across narrowed area with pan-diastolic spill which suggest severe coarctation.

DISCUSSION

Association of coarctation have been rarely reported along with TAPVC. Only 1.4%, that is, 6 out of 422 patient have been previously reported.[1-3] Repair of infracardiac non obstructive TAPVC with CoA in 7 day old girl reported by Doksoz et al.[1] Leval et al reported a successful one stage repair of 12 day old male baby with diagnosis of non-obstructive supracardiac TAPVC with CoA.[2] Recently Hayashi et al reported successful repair of non-obstructive supracradiac TAPVC with coarctation in 6 day old boy.[4] Generally in TAPVC case already there was low systemic output due to pulmonary venous obstruction and relatively small or restricted inter-atrial communication. But in our case there was obstructive supracardiac TAPVC along with severe discrete coarctation which lead to significant reduction in lower body blood supply and lead to rise in lactate levels and other metabolic de-arrangements, which caused very unstable haemodynamic status of baby inspite of high ionotropic and ventilator supports. We recommend urgent referral to CTVS centre of such baby for surgical treatment.

CONCLUSION

Obstructive TAPVC with severe coarctation patients are bad subset of cyanotic congenital heart diseases. They should be urgently referred to higher CTVS centre or if antenatally diagnosed such case should be delivered in such a centre which is well versed with such critical pediatric cardiac cases.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Doksöz Ö, Güven B, Demirpençe S. Coarctation of the aorta with infracardiac total anomalous pulmonary venous drainage: a rare combination. Ann Thorac Cardiovasc Surg. 2014;20:778-80.
2. de Leval MR, Stark J B-CR. Total anomalous pulmonary venous drainage to superior vena cava associated with preductal coarctation of aorta. Successful correction in a 12-day-old infant. Br Hear J. 1973;35:1098-03.
3. Seale AN, Uemura H, Webber SA. Total anomalous pulmonary venous connection: morphology and outcome from an international population-based study. Circulation. 2010;(122):2718-26.
4. Hayashi H, Sughimoto K, Oka N, Tsuchida Y MK. Coarctation of the aorta with total anomalous pulmonary venous connection: a case report. AME Case Rep. 2021;5:11.

Cite this article as: Mane S. Rare association of obstructed supracardiac TAPVC with coarctation of aorta: case report. Int J Contemp Pediatr 2022;9:630-1.