Hepatic Vein Collateral Draining to Left Atrium, an Extremely Rare Anomaly Cause of Hypoxemia, Case Report from Queen Alia Heart Institute

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ABSTRACT

Hepatic veins collateral to the left atrium are considered a very rare anomaly which causes hypoxemia, our patient 14 years old male has single ventricle dextrocardia corrected 12 years ago and developed hepatic vein drainage to left atrium, treated by minimal invasive procedure through ligation of the hepatic veins collateral, after which oxygen saturation changed from 80% to 96% in few seconds.

Key words: Hepatic veins, Hypoxemia, Left atrium

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Introduction

Patients who were born with single ventricle and had undergone surgical correction using bidirectional Glenn shunt and Fontan surgery, are prone to develop abnormal systemic venous collaterals due to high pressure circulation which can lead to hypoxia and cyanosis in these patients. (1,2)

Recently most of such abnormal pathologies are treated by minimal interventional procedures (percutaneous catheterizations).

Case Report

Fourteen years old male with situs inversus, dextocardia, single ventricle, Patent Ductus Arteriosus (PDA), transposition of great vessels (TGV), bilateral superior vena cavae (SVC) without Innominate vein, pulmonary stenosis and common atrium. during first year of life

the patient complained of cyanosis, hypoxia and effort intolerance.

At age of 2 years he underwent bilateral Bidirectional Glenn shunt, on follow up during the following 2 years the patient was doing well and the Glenn shunt was functioning well.

Two years later the patient underwent completion of TCPC (Total cavopulmonary connection) with Right Atrium to left pulmonary artery (LPA) shunting. The patient then was discharged, On follow up 5 years later, TCPC was found functioning well but he has aortic valve (AV) valve regurgitation grade I, despite this the kid had grown up normally and didn't complain of hypoxia and investigations didn't show desaturation.

After 8 years from last operation, the patient started complaining of cyanosis and hypoxia (O₂ saturation around 80-8 % at rest and 70% on

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exercise), cardiac catheterization showed multiple hepato-venous collaterals (Fig. 1, 2).



Fig.1: Catheterization angiogram, shown inferior vena cava and collateral in the left sided liver tissue.



Fig. 2: Cathaterizaion angiogram, shown the progression of the die from the intrahepatic collateral to confluent one above the diaphragm to the left atrium.

Patient underwent trial of Transcatheter embolisation to close these collaterals but the trial failed, so the patient had surgery.

Surgical procedure was done on beating heart off pump, with lower median sternotomy incision (5cm) (Fig. 3), opened by layers and adhesions were released, surgeon start dissection between the diaphragm and the heart until he could identified the collateral which was large in size (2cm), then the collateral was ligated (Fig. 4) and during next few seconds the oxygen saturation was rising from 80% to 96% (Fig. 5) and (Fig. 6). The duration of the procedure was 87 minutes (Fig. 7).

Patient was discharged on the 4th day post operation with no cyanosis and oxygen saturation 96%.



Fig. 3: Lower small median sternotomy incision



Fig. 4: Collateral surrounded by the band before ligation



Fig. 5: Saturation 80% before ligation the collateral



Fig. 6: Saturation 96% after ligation the collateral



Fig. 7: End of operation

Discussion

This case is considered rare for several reasons, first of all the type of collateral which developed after bidirectional glenn shunt and total cavopulmonary connection is rare comparing to other types. According to previous reports the common collaterals are abnormal superior vena caval connection and brachiocephalic vein followed by the left phrenic vein, ⁽³⁾ usually these collaterals happen due to increase in central venous pressure⁽⁴⁾ and such collaterals lead to systemic hypoxia but usually these collaterals develop in early stages after the first surgery, but with our patient the late manifestation is considered a rare occurence too.

The second reason is the method of treatment, as we mentioned above Transcatheter embolisation is the method of choice but in some cases when it fails or there is contraindication surgery will be the solution, the usual surgical procedure is done with on pump, but here we used small incision and ligated the collateral with beating heart.

Finally, the result is satisfactory and follow up is recommended to identify any new collaterals or recanalization.

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