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Case Report

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A CASE OF UNUSUAL SYSTEMIC AND PULMONARY VENOUS DRAINAGE

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ABSTRACT

Variation in systemic and pulmonary vein anomalies anatomic pattern and clinical presentation can present a diagnostic dilemma which can have significant implications on its course and outcome. This highlights the significance of its early recognition and accurate diagnosis with the help of various imaging modalities including transcatheter angiography for planning timely referral and appropriate intervention. We present a unique case of both systemic and pulmonary venous anomaly in a single patient.

INTRODUCTION

Systemic venous anomalies have a wide spectrum from asymptomatic anatomic variation to complex abnormalities leading to cyanosis or complicating surgical repair of congenital heart disease (CHD)[1]. Clinically significant abnormalities of systemic veins are rare in situs solitus or inversus (non isomeric) but exceed 90% in situs ambiguous (isomeric)[2,3]. Total anomalous pulmonary venous drainage (TAPVR) is a rare condition and makes 1-2% of all congenital heart lesions[4]. TAPVR association with lesion like common AV canal, conotruncal anomalies, and systemic venous anomalies, mitral and pulmonary atresia characterize its association with heterotaxy patients[1]. To our knowledge no case of both systemic and pulmonary veins anomalies to this extent in single patient so far been reported.

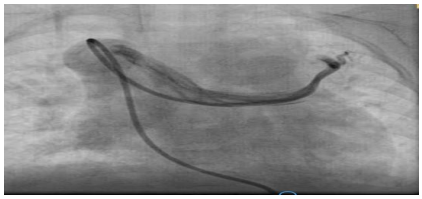

Figure-1: Left sided pul veins coming in common venous confluence and draining via ascending vein on left side in RSVC.

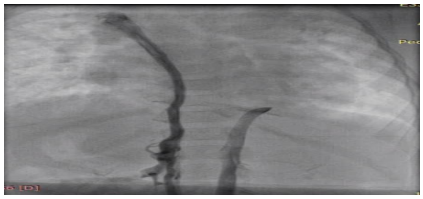

Figure-2: Interrupted suprahepatic bilateral IVC with Azygous continuation of Right sided IVC to RSVC.

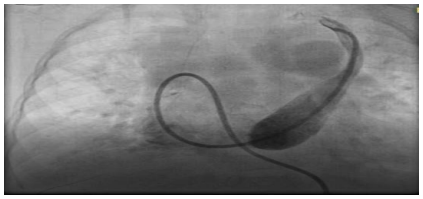

Figure-3: LSVC draining in RA via coronary sinus.

CASE REPORT

We present a case of one year old boy who was referred from a peripheral hospital for cardiac assessment to rule out CHD because of h/o increase work of breathing since birth and intermittent cyanosis specially on crying. Child also had h/o recurrent respiratory tract infections since birth although born full term with weight within normal centiles had so far shown poor weight. On examination he was weighing <2SD for age and sex, saturating 88% in air, well perfused, but tachypneic with mild S/C recession, had clear chest, 2/6 systolic murmur at LSE and no hepatomegaly.

Echocardiogram revealed situs ambiguous, levocardia, interrupted inferior vena cava (IVC), dilated coronary sinus with Left superior vena cava (LSVC), all Pulmonary veins draining via an ascending vein to right superior vena cava (RSVC), dilated RA + RV and Rt to Lt shunting across unrestricted ASD.

Cardiac angiogram was planned to delineate the extent of pulmonary and systemic venous anomalies which confirmed drainage of all four pulmonary veins in a common confluence and then draining in RSVC via ascending vein on left

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side (fig-1), it showed bilateral infra-hepatic IVC (fig-2). Lt sided IVC was found draining in hepatic sinusoids on Rt side and ultimately draining in right atrium (RA) via hepatic veins, Rt sided IVC was draining in RSVC via Azygous continuation (fig-3). LSVC was draining in coronary sinus coming in RA (fig-3).

DISCUSSION

Combination of pulmonary and systemic venous anomalies in heterotaxic or isomeric patient is not an uncommon feature but presence of full blown pulmonary and systemic venous anomalies in a single patient is extremely rare. Although interrupted IVC is characteristic of LAI and bilateral Superior vena cava (SVC) along with partial anomalous venous drainage (PAPVR) is common in these patients, incidence of TAPVR is rare. In contrast our patient has both. Incidence of LSVC in two large autopsy series was approximately 0.3%[5,6]. Persistent LSVC to coronary sinus result in normal return of systemic venous blood to RA but this anomaly may have significant clinical implications for patients with associated cardiac malformation[1]. In one series incidence of interrupted SVC was 86% in patients of visceral heterotaxy[2]. Interrupted IVC with azygous continuation usually does not result in physiologic abnormality and clinical manifestation but can complicate cardiac catheterization, interventional procedures like radiofrequency catheter ablation and surgical procedures like bidirectional glenn operation (BDG) and Fontan. Bilateral nature of four of five venous systems that contribute to formation of IVC can explain the presence of bilateral IVC in supra and infra hepatic region. Bilateral infra hepatic IVC in isolation can occur in association with either normal or abnormal visceral situs without any hemodynamic disturbance as reported by several before[7]. Although existence of unpaired segment of IVC (hepatic segment) does not permit the formation of truly complete bilateral IVC but in presence of absent ductus venous it is possible to have two venous channels mimicking bilateral IVC[8].

CONCLUSION

Presence of all pulmonary and systemic venous anomalies is a rare combination even in heterotaxic patient and can lead to significant clinical morbidity. Early and accurate diagnosis can help in careful surgical planning timely to reduce morbidity and mortality in these complex patients.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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