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Persistent Left Superior Vena Cava: A Rare Association

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Abstract

Persistent left superior vena cava (PLSVC) is the most frequent abnormality in the general population with the frequency of 0.1% to 0.5%. It results from the failure of the involution of the left anterior cardinal vein. Right and Left SVC can coexist together in 80% to 90% of cases. Association of PLSVC with ano rectal malformation (ARM) is very rarely reported. Hence, here is a report of a unique case of PLSVC in a female neonate with ARM

Keywords

cardio vascular anomalies, anorectal malformation, vena cava

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Introduction

The superior vena cava duplication is a rare anomaly with a rate in the general population of 0.3%. Persistent left superior vena cava (PLSVC) is the result of a persistent patency of an embryological vessel (left anterior cardinal vein) that is present during eighth week of early embryonic life. It usually drains into the right atrium through a dilated coronary sinus. However, in some cases, it may drain directly into the left atrium producing a right-to-left shunt. In most of these cases the right superior vena cava may be present and may or may not communicate with the left superior vena cava. Persistent left superior vena cava is generally discovered without any clinical signs and symptoms; diagnosed incidentally by imaging done for other anomalies or lesions.[1-3] This is the first ever case of anorectal malformation associated with isolated persistent left superior vena cava to be managed successfully in our institute and doing well on post operative follow up; is on regular surveillance with us since 4 years.

Case Report

A day 2 female neonate born to non consanguineously married couple, full term vaginal delivery, with birth weight of 2.5 kg, was noticed to have absent anal opening (Figure 1d) with abnormal passage of stools from the vestibule. Otherwise baby was asymptomatic without any history of cyanotic spells. Mother gave a history of intra uterine demise in her 3rd pregnancy: other siblings were normal. On examination; baby had dysmorphism like broad forehead, frontal bossing, hypertelorism, depressed nasal bridge, low set ears with left dysplastic ear having bilateral pre auricular skin tags (Figure 1a-c). Baby was admitted and evaluated. Mother was taught about bowel washes with normal saline once a day; discharged and kept on regular follow up. As a part of routine screening of associated VACTERL, various investigations were done including ultrasonography. With essentially normal ultrasonographic findings on follow up ; 2D echocardiography was done, showed small patent foramen ovale having left to right shunt and left persistent superior vena cava draining into the coronary sinus; which was confirmed on contrast enhanced computed tomography(CECT)(Figure 2). When the baby was 6 months old with a weight of 6 kgs, a primary posterior sagittal anorectoplasty (PSARP) (Figure 1e) under general anesthesia was performed uneventfully in prone position having a mechanical ventilator stand by. Baby withstood the procedure well intra operatively and without requiring post operative ventilatory support. Baby is now 4 years old, asymptomatic and is on regular

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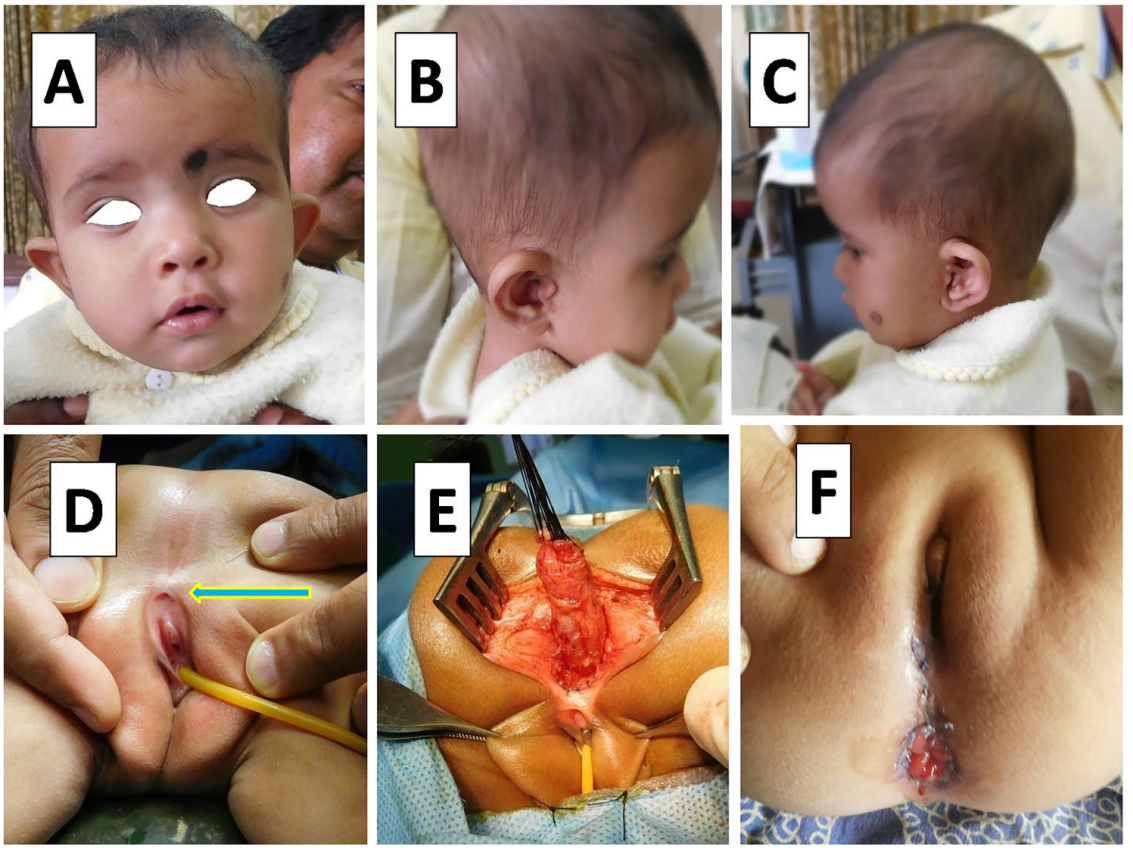

Figure 1. (A) Facial dysmorphism. (B and C) Low set ears with pre auricular tags. (D) Green colored arrow pointing at ano vestibular fistula. (E) Intra operative picture of mobilized rectum. (F) Post operative appearance.

follow up with normal stool pattern through the neo anus (Figure 1f). Detailed genetic study has not been done due to parental financial constraints.

Discussion

The incidence of left superior vena cava in general population is 0.3% to 0.5%. Isolated PLSVC occurs in only 0.09% to 0.13% of patients who have congenital heart disease.[1-6]

A PLSVC is an embryological remnant that represents persistence of the embryonic left anterior cardinal vein and is the most common congenital anomaly involving thoracic central venous return. The PLSVC usually drains into the right atrium through the dilated coronary sinus (92%) with no hemodynamic consequences. In 8% of the cases the PLSVC drains into the left atrium causing right to left shunt, which is usually not large enough to cause cyanosis since it only drains the left upper limb and left side of the head and neck. In the vast majority of cases (82%-90%) a normal (but small) right sided SVC is also present, and a persistent bridging vein (left brachiocephalic vein) is seen in 25%-35% of cases.[1-3]

Almost 40% of the previously reported patients with PLSVC had associated significant cardiovascular anomalies, the most frequent being cardiac septal defects.[3,5,6]

Recto vestibular fistula is the most common ARM in females. VACTERL (vertebral, anorectal, cardiac, tracheo esophageal, renal, and limb) and CHARGE (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities) associations have been described.[3,4,5]

Since its introduction in 1982 by DeVries and Pena;[4] the posterior saggital anorectoplasty (PSARP), in which the recto urogenital connection is divided and the neonatal rectum brought to the perineum; is widely practiced worldwide, in female ARM without the need for primary colostomy, with good functional and cosmetic outcome.

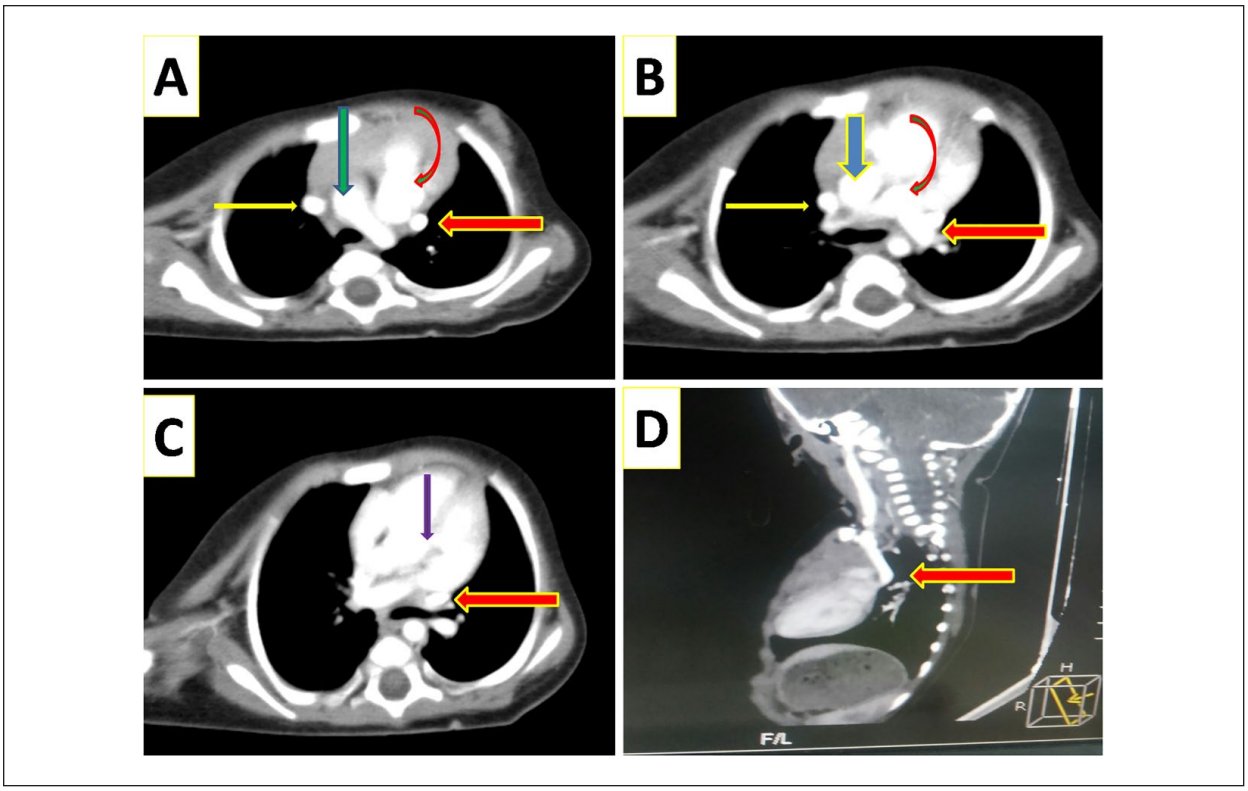

Figure 2. Contrast enhanced computed tomographic images: (A) At the level of arch of aorta. Yellow straight arrow on right-showing right SVC, Green arrow showing arch of aorta, Red curvilinear arrow showing-pulmonary trunk and Red arrow on left shows Left SVC. (B) Yellow arrow on right shows RSVC, Blue arrow shows right atria, Curvilinear red arrow showing pulmonary trunk and Red arrow on left shows LSVC. (C) Red arrow -Left SVC draining into the coronary sinus. (D) Oblique section showing whole course of left SVC draining into coronary sinus.

Teixeitra et al[5] studied 68 neonates of ARM with cardiovascular anomalies: where in 2 neonates had PLSVC with ARM along with other lethal cardiovascular anomalies detected in early neonatal period; who succumbed to their illnesses during the same hospital stay.

Author had a day 2 female neonate with recto vestibular fistula (ARM) having associated PLSVC draining into the coronary sinus; underwent PSARP by pediatric anesthetists and pediatric surgeon in a tertiary care pediatric centre. Hypoxia, hypercarbia and acidosis were avoided intra and post operatively: hence managed successfully and discharged.

As per the literature search, this is the first ever case to be reported in the English literature, the isolated persistent left superior vena cava with ano rectal malformation alive and is on regular follow up with us since 4 years.

Clinical Significance of PLSVC

* PLSVC is an important variant to know in case of central venous catheter positioning including manipulation of the guide wire due to proximity to coronary sinus, causing hemodynamic instability.
* Placement of pacemaker leads and pulmonary artery catheter can be technically challenging, therefore the left subclavian vein is the preferred in these cases.
* Cardiac surgeries involving cannulation of SVC for cardiopulmonary bypass might require isolated LSVC cannulation whenever it is detected.[6]
* Arrhythmias and conduction abnormalities are common in the presence of PLSVC due to a dilated coronary sinus which will stretch the AV nodal tissue predisposing to reentrant tachycardia.
* This case report has been written and reported as per SCARE 2018 criteria.[7]

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Author Contributions

JSA: Operating surgeon as well as the author of this manuscript.

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Ethical Approval and Informed Consent

Written and verbal consent taken from parents.

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Supplemental Material

Supplemental material for this article is available online.

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