

Persistent left superior vena cava in a stillborn fetus – a case report

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SUMMARY

Persistent left superior vena cava is a rare congenital malformation. It represents some form of developmental arrest. It may coexist with right superior vena cava in different patterns and sizes. The knowledge of variations of central venous anatomy and its association with congenital malformation of the heart is important during central venous access device placement. Here we present a case of type A persistent left superior vena cava with hypertrophied right atrium, right ventricle and large *ostium secundum* defect in a stillborn fetus during routine fetal autopsy.

Key words: Left superior vena cava – Anterior cardinal vein

INTRODUCTION

The superior vena cava (SVC) is one among the three major veins that carry venous blood to the right atrium of the heart. It develops from the right anterior cardinal vein and right common cardinal vein (Moore and Persaud, 2008). Normally the left anterior cardinal vein disappears. Persistence of the left anterior cardinal vein is a rare congenital malforma-

tion. Persistent left superior vena cava (PLSVC) represents the left anterior cardinal vein in adults. The left superior vena cava occurs in up to 5% of children with congenital heart disease (Haaga et al., 2009), and 0.3% to 0.5% of the general population (Ömer et al., 2008). We present a case of persistent left superior vena cava.

CASE REPORT

A 27-year-old mother with parity 3 delivered a male term stillborn fetus weighing 3 kg. All previous babies were normal. The stillborn fetus was dissected as a part of routine fetal autopsy for study of congenital anomaly in the department of Anatomy of a tertiary referral healthcare centre in the North Eastern part of India. The study was approved by the institutional ethical committee. Informed consent of the parents of the fetus was taken. A midline incision extending from the suprasternal notch to pubic symphysis was practised. Each and every structure was meticulously observed taking precautions not to damage any of them.

On dissection of the pericardium, a hypertrophied heart was observed. The right atrium and the right ventricle were enlarged (Fig. 1).

The left atrium was small. A relatively small vein was seen extending from the junction of the left internal jugular vein and the left brachiocephalic vein to coronary sinus. The vein was on the posterior aspect of the left atrium, and entered into the left half of the atrioventricular sulcus (Figs. 2, 3). The heart was dissected and each chamber was thoroughly examined.

Coronary sinus was seen opening in the right atrium between the interatrial septum and the tricuspid valve. A large *ostium secundum* defect was observed in the interatrial septum communicating both the atria (Fig. 4). Right SVC and inferior vena cava were normal (Fig. 5). Other systems were found to be normal.

DISCUSSION

SVC which drains blood from the head and neck region develops from the right anterior cardinal vein and the right common cardinal vein. Developmentally anterior cardinal veins are a pair of bilaterally symmetrical veins. The left anterior cardinal vein drains venous blood from the left side of the head and neck region into the left horn of *sinus venosus* via the left common cardinal vein. Normally the lower part of the left anterior cardinal vein degenerates, and bilateral symmetrical arrangement of the veins changes to a unilateral asymmetrical pattern (Campbell and Deuchar, 1954). The left horn of *sinus venosus* persists as the coronary sinus (Moore and Persaud, 2008). Failure of the left anterior cardinal vein to degenerate results in persistence of a left SVC (Cormier et al., 1989). Hence, persistence of left sided SVC represents some form of developmental arrest.

During dissection of the present case the SVC was seen in the normal position. An additional vein was observed on the posterior aspect of the left atrium, extending from the junction of the left internal jugular and the left brachiocephalic vein to coronary sinus. The position and the connections of the left sided vein was similar to the embryological left anterior cardinal vein. Hence the left vein was diagnosed as persistent left superior vena cava with a communication (i.e. left brachiocephalic vein) to the SVC on the right.

PLSVC can occur in several anatomic variations. Nandy and Blair (1965) have classified embryologically the cases of anomalous SVC

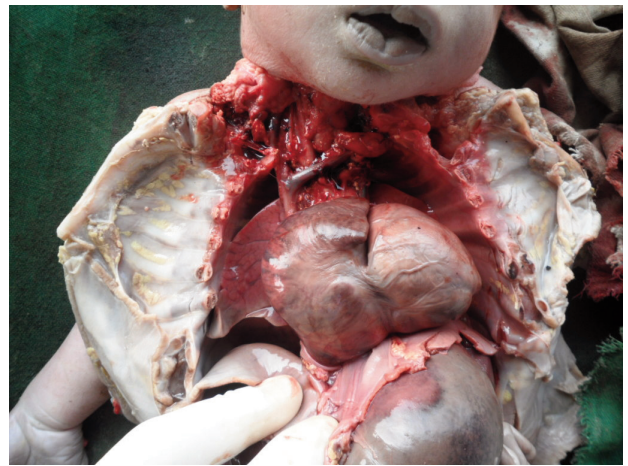


Fig. 1. Anterior view of hypertrophied right atrium and right ventricle with prominent brachiocephalic vein.

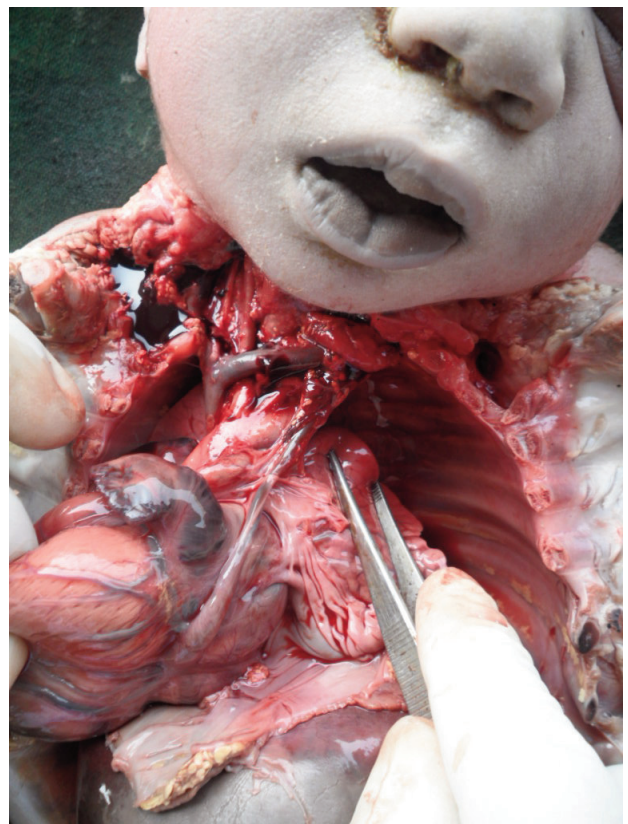


Fig. 2. Posterior aspect of the heart with the left superior vena cava extending from left brachiocephalic vein to coronary sinus (in situ).

into 4 types: A. Persistence of both right and left anterior cardinal veins with persistence of the anastomosis between them. B. Persistence of both anterior cardinal veins without the anastomosis. C. Persistence of the left anterior cardinal vein with the obliteration of the right. D. Persistence of both anterior cardinal veins with similar azygos veins on both sides. The present case is considered to correspond to type A, where an anastomotic channel existed between left and right SVC. Most commonly PLSVC coexists with a right SVC in up to 80



Fig. 3. Posterior aspect of the heart showing a persistent left superior vena cava with arrows pointing towards the left superior vena cava.

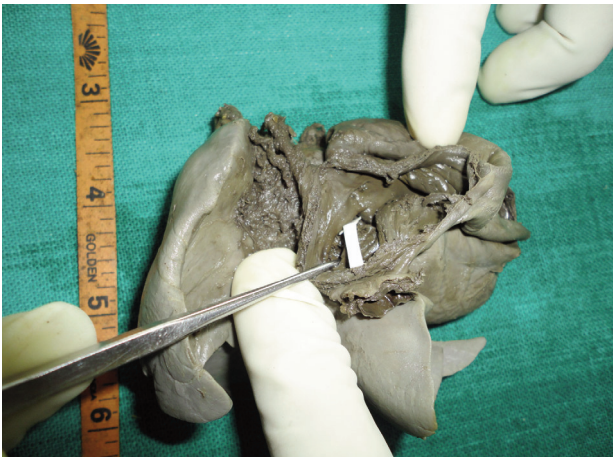


Fig. 4. Photograph showing large interatrial septal defect (*ostium secundum* type) in right superior view with arrow inserted in it.

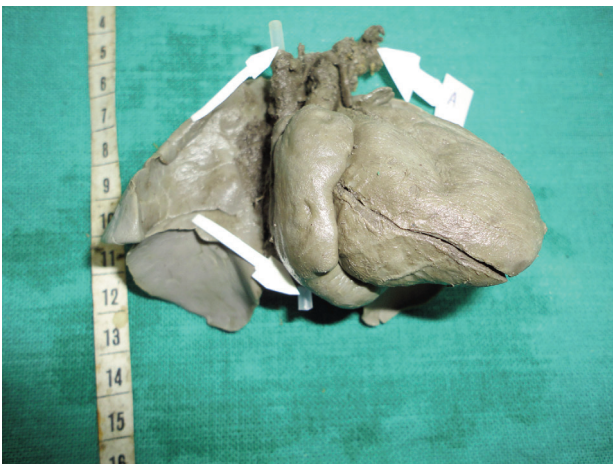


Fig. 5. A normal superior vena cava with arrows pointing towards the superior and inferior vena cava and left subclavian vein can be observed in this anterior view.

to 90% of cases. While in many cases these bilateral SVCs are of relatively equal size, various degrees of differences in size can exist between that of the right SVC and the PLSVC. The PLSVC in this case was smaller than the right SVC (Couvreur and Ghaye, 2009).

Bjerregaard and Laursen (1980) reported that PLSVC is most frequently associated with total anomalous pulmonary venous return (TAPVR) (10%), pulmonary atresia (6%), tricuspid atresia (4.5%), hypoplastic left ventricle and endocardial cushion defects (2.5%), and tetralogy of Fallot (2%) (David et al., 1980). In the present case, anomalies such as TAPVR, pulmonary atresia, tricuspid valve stenosis, and tetralogy of Fallot were absent, but right atrial and right ventricular hypertrophy along with large *foramen secundum* defect were present.

Prevalence of PLSVC is very rare in general population (Ömer et al., 2008). If a patient is suspected to have a PLSVC at the time of attempted central venous access device placement, then an appropriate investigation to fully characterize the central venous anatomy should be undertaken. This is important in order to confirm the presence of PLSVC, to characterize the central venous anatomy of the contralateral right side, to characterize the pattern of cardiac venous return to the right atrium or to the left atrium, and to evaluate the patient for other potential coexisting congenital heart abnormalities (Povoski and Khabiri, 2011).

Conclusion: Although persistent superior vena cava is a rare congenital anomaly, this variation in central venous anatomy and its association with other congenital cardiac malformation should be kept in mind during investigative or surgical procedures on the heart. Routine pathological autopsies may help in assessment of its prevalence and its associations. A case of type A persistent left superior vena cava with right atrium and right ventricular hypertrophy and large *foramen secundum* defect is being presented here.

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