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NON-COMMUNICATING BILATERAL SUPERIOR VENACAVA – A CASE REPORT AND BRIEF LITERATURE REVIEW

KEYWORDS: superior venacava, coronary sinus, cardinal veins

Dr.Pallavi

Assistant professor, Department of Anatomy Father Muller Medical College Kankanady, Mangalore, Karanataka, India

ABSTRACT *Abnormalities of the central vascular system are always of extreme interest, on account of the light they cast on many developmental problems.Normal anatomy describes the formation of superior venacava by the union of right and left brachiocephalic veins draining the head and neck area. Presence of bilateral superior venacava was encountered during routine dissection in the Department of Anatomy, Kasturba medical college, Manipal. Right and left superior venacavae were formed as a continuation of respective brachiocephalic veins. The left superior venacava drained into the right atrium via the coronary sinus which was enlarged. No communication was observed between the two venacavae indicating that the right and left halves of the head and upper limb drain back to the heart independently. Persistence of the left anterior cardinal vein and regression or absence of the communicating vein would be the possible embryological explanation for this anomaly. Literature reviews suggest that the frequency of its occurrence is 0.3%-1.3% in general population but higher (10-11%) in people with congenital heart diseases. Although it has no physical derangement per se, may complicate or mislead the placement of cardiac catheters or pacemaker leads. Dilated coronary sinus may cause cardiac arrhythmias due to stretching of the AV node and bundle of His. Presence of these anomalies is an indication for the screening of other associated cardiac abnormalities.*

Introduction

Abnormalities of the central vascular system are always of extreme interest; on account of the light they cast on many developmental problems and as from the fact that they usually place a strain upon the central organ of the circulation.[1]The superior vena cava is among the three major veins that carry venous blood to the right atrium of the heart. The others are the inferior vena cava and the coronary sinus. It develops as a result of morphological changes that occur in the right anterior and common cardinal venous system, and during which developmental anomalies of the vena cava can occur.

Normal anatomy describes the formation of superior venacava by the union of right and left brachiocephalic veins which are in turn formed by the union of corresponding internal jugular and subclavian veins, draining the respective half of the head and neck area and the upper limb. Normally measures 6-8cm long and 2cm wide.

Many anomalies of the superior venacava (SVC) are known, each representing some form of developmental arrest. The basic abnormality is failure of the left anterior cardinal vein to involute normally, resulting in persistence of left SVC. Persistent left superior venacava (PLSVC) is an uncommon vascular anomaly, yet the most common congenital anomaly of thoracic venous system.[2]Since these anomalies are clinically silent, they are often unsuspected and discovered incidentally on radiographic studies done for other reasons.[3]A left SVC may be associated with a wide variety of cardiac defects, including single atrium, VSD, PDA, teratology of Fallot, truncusarteriosus, pulmonary stenosis or atresia, tricuspid atresia, Ebstein malformation, Tausing-Bing complex.

Case report

During routine dissection of thoracic region in the department of Anatomy, Kasturba medical college, Manipal, we encountered the presence of bilateral superior vena cava. Both right and left superior vena cavae were formed as a continuation of brachiocephalic vein of the corresponding side. The left superior venacava descended in front of the arch of aorta and drained into the right atrium via the coronary sinus. Communication was not observed between the two venacavae indicating that the right and left halves of the head, neck and the upper limb drained back to the heart independently.Both of them were of relatively equal caliber and length, measuring about 8.6cm on the right side and 8.2cm on the left (fig.1). The coronary sinus was considerably enlarged, measuring 3×5.5cm to occupy almost completely the posterior part of the left half of the atrioventricular sulcus (fig.2).The opening of coronary sinus was also enlarged, which was seen in between the right atrioventricular and inferior venacaval openings. The arch of azygos vein opened into the right SVC and normal tributaries of oblique part of the left brachiocephalic vein like the left superior intercostal vein opened into the left SVC.

Fig.1: Anterior view of the heart in situ showing bilateral superior vena cavae (blue colour). SVC- superior vena cava, PLSVC- persistent left superior vena cava, AA- arch of aorta.

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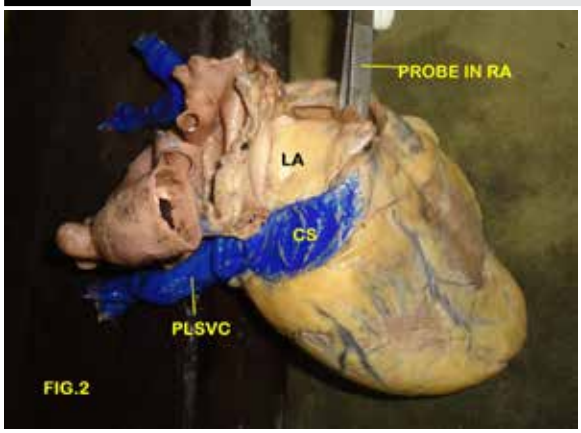

Fig.2: posterior view of heart showing the PLSVC opening into enlarged coronary sinus. CS- coronary sinus, RA- right atrium, LA- left atrium.

Discussion

The first in-depth review on the topic of the great anterior veins of the thoracic region, including PLSVC, in man and mammals, was published in 1850 by John Marshall. Since that time, a plethora of, and too numerous to cite, papers have been published on various aspects of the PLSVC. Reported prevalence of double SVC is 0.1%-0.3% in the general population and 2%-9% of patients with congenital cardiac anomalies. Among them, a coexisting right SVC is present in about 85%.[4]Since the vast majority of cases of this congenital venous anomaly are asymptomatic, its true incidence in the general population may actually be difficult to accurately establish.Suspicion of left superior vena cava may arise on the postero-anterior chest X-ray, where it may appear as widening of the aortic shadow, paramediastinal bulging, paramedian stripe, or a low-density line along the upper left margin of the heart. Transthoracic echocardiogram (TTE) is the mainstay tool for diagnosing PLSVC which also helps to identify other cardiac anomalies that might be associated. A CT scan with contrast seems to offer voluminous information in delineating the anatomy.[5]Several studies have reported the association of the double superior venacava with other variations.Persistent left superior vena cava (PLSVC) with anomalous left hepatic vein drainage into the right atrium was reported by Bhatti S et al., (2007)[5]. Anomalous connection of superior vena cava to the left atrium presenting as epilepsy was reported by Singh S et al., (2008)[6]. Gesase A (2009)[7] reported a case of Double Superior Vena Cava Presenting with Anomalous Jugular and Azygous Venous Systems. Povoski et al., (2011)[8] published an article on Persistent left superior vena cava: Review of the literature, clinical implications and relevance subsequent to his experience of difficulty in central venous access device placement and venography in a cancer patient. Early in 1980 Hardy et al.,[9] had reported that failure to cannulate the PLSVC in the absence of the left innominate vein will result in cerebral congestion.

From a practical standpoint anomalies of the superior venacava can be divided into three classes.[10]

Type I- (a)Double superior venacava with large anastomosis, (b) Double superior venacava with small anastomosis

Type II- Double superior venacava with no anastomosis

Type III- (a) Left superior venacava with no trace of right vessel, (b) Persistent left venacava; unclassified.

Recently, there have been reports raising concerns about significant ventricular inflow tract obstruction as a result of the enlarged coronary sinus. The marked enlargement of the coronary sinus overlays immediately superior to and partially occluding the mitral valve, with consequent obstruction to left ventricular inflow. An enlarged coronary sinus has also been associated with thrombus formation and rupture.

Development of superior vena cava involves the morphological changes that take place in the anterior cardinal venous system that drain the cranial part of the early embryo. An oblique communication that establishes between the right and left anterior cardinal veins results in the obliteration of terminal part of the left anterior cardinal vein. Simple failure of obliteration of the left anterior cardinal vein results in the persistence of the left superior vena cava. Absence of communication between the two superior vena cavae may due the obliteration of the communicating vessel or due to the failure of its development.

Conclusion

A PLSVC does not, in itself produce any physiological derangement. However, it has important clinical implications in certain situations wherein it may complicate placement of cardiac catheters or pacemaker leads. Awareness of this anomaly may reduce confusion about the position of a catheter /lead that appears to have strayed.[11]Incidental finding of PLSVC is of potential importance to surgeons, interventional radiologists, and other physicians actively involved in central venous access device placement in cancer patients.[8]Lack of knowledge can pose serious complications. Transthoracic echocardiography – including agitated saline infusion to the antecubital vein is a robust tool for the accurate diagnosis of this congenital thoracic venous malformation.[5]

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