An Association of Persistent Left Superior Vena Cava with Cribriform Secundum Atrial Septal Defect and A Long Muscular Band in The Right Atrium

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Citation


Abstract

We describe a case of an association of persistent left superior vena cava with cribriform secundum atrial septal defect and a long muscular band in the right atrium.

CASE REPORT

A 28 - year- old male was admitted to our hospital with nonspecific angina pectoris and exertional dyspnea. Transthoracic echocardiogram demonstrated two middle-sized defects in secundum septum with a left-to-right shunt. Cardiac catheterization showed persistent left superior vena cava (PLSVC) and atrial septal defect. Cardiac catheterization pointed out no additional pathology.

He was operated under endotracheal general anesthesia and in supine position. Following a median sternotomy, pericardium was opened longitudinally. After heparinization, extra-corporeal circulation was established between the venae cavae and the ascending aorta. The PLSVC was temporarily occluded with a snare(Figure 1).

A cross clamp was placed on aorta and by antegrade intermittent isothermic blood cardioplegia from aortic root, cardiac arrest was established. Hypothermia was moderate (32ºc). A vent was placed via the right superior pulmonary vein. Standard right atriotomy was made. Cribriform multiple secundum atrial septal defects (ASD) were evaluated regarding their localization, size, other related cardiac structures and possible associated abnormalities. There was a long muscular band in the right atrium (Figures 2 and 3).
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This muscular band was resected. After this step cribriform ASD was made an uniform defect with excision of the septal defect portions (Figure 4).

We performed an e-PTFE patch closure of atrial septal defect as to drain blood from left SVC to right atrium (Figure 5).

Incision of right atriotomy was closed. The post-operative course was uneventful with successful anatomical correction. Postoperative echocardiographic data confirmed complete correction of the lesions.

COMMENTS

Secundum atrial septal defect (ASD) is a common congenital heart disease and accounts for approximately 6% to 10% of all congenital cardiac defects (1, 2). One of the most common congenital anomalies of systemic veins is persistent left superior vena cava (3). Left superior vena cava is developed as a consequence of persistence of the continuation of the
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left anterior and left common cardinal veins with the left horn of sinus venosus when the proximal segment of these veins did not disappear. Association of persistent left superior vena cava with other congenital cardiac diseases is common and frequently encountered during diagnostic studies. While overall incidence is 0.5% to 2%, the incidence of this disease with the association of a congenital cardiac disease reaches 10%. The association of PLSVC and secundum ASD is a congenital anomaly type which is rarely encountered.

In Muñoz study for the research of the frequency of persistent left superior vena cava as well as the associated congenital heart disease, 66 hearts were studied from 1277 necropsies. They were analyzed with the sequential segmental approach. 33 hearts had situs solitus (group I) and 34 hearts had isomeric situs (Group II). The group I showed double superior vena cava, the left one had continuation with the sinus coronary; in 25 of them the left brachiocephalic vein was absent, in 6 this vein was present, 5 with narrow lumen and in one it was dilated. The more frequent congenital heart diseases were ventricular septal defects, truncococcal cardiopathies and anomalies in the atrioventricular connection. The hearts of group II did not have coronary sinus; the venous connections were in the atrial roof. 19 hearts had double superior vena cava and 15 specimens had only the left one. The congenital heart disease in this group were complex with multiple patterns of association.

Owing to the fact that cardiopulmonary by-pass procedure may be problematic in patients with persistent left superior vena cava, this anomaly should be detected before cardiac surgery and required measures should be taken. Preoperative diagnosis is very difficult in some times and may draw the reliable support from echocardiography and catheterization.

The left superior vena cava has surgical significance when congenital heart disease is present. If PLSVC exists, other venous system anomalies should be investigated and cardiopulmonary by-pass procedure should be performed in different ways if necessary.

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