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PARTIAL ANOMALOUS PULMONARY VENOUS CONNECTION TO THE SUPERIOR VENA CAVA WITH ATRIAL SEPTAL DEFECT

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ÖZET

ATRIAL SEPTAL DEFEKTLİ BİR OLGUDA VENA KAVA SUPERİORA PULMONER VENÖZ BAĞLANMADA PARSİAL ANOMALİ

Kliniğimizde atrial septal defektli ve vena kava superiora pulmoner venöz bağlanmada parsiyel anomalisi olan bir olguya kateterizasyonla tanı kondu. Bu anatomik özellik toplumda sanılandan daha sık olabilir. Hastalık farklı kardiyak ve pulmoner anomalilerle birlikte bulunabilir.

Anahtar kelimeler : *Parsial anomali pulmoner venüz bağlanma, atrial spetal defekt*

SUMMARY

A case of partial anomalous pulmonary venous connection to the superior vena cava with atrial septal defect was diagnosed by catheterization in our clinic. This distinct anatomic entity is perhaps more commonly prevalent that has been previously believed. It may be associated with other cardiac and pulmonary anomalies.

Key words*: Partial anomalous pulmonary venous connection, atrial septal defect*

INTRODUCTION

Partial anomalous pulmonary venous connection to the right side of the heart is a frequent anomaly that has been found in 0.7 % of routine autopsies (l). The first diagnosis during life by angiocardiography was described at 1949 by Dotter and colleagues (2). The association of partial anomalous pulmonary venous connection (PAPVC) and an atrial septal defect (ASD) has been recognized for more than a century. About 10 % to 15 % of all patients with an ASD will have anomalous pulmonary venous connection (3). Rarely does PAPVC occur without ASD (4). We report here a patient with PAPVC to the vena cava superior and an associated atrial septal defect.

CASE REPORT

A 25-year-old man was referred for evaluation of increasing palpitation and shortness of breath on exertion. Physical examination revealed a grade 2/6 systolic ejection murmur heard maximally at the upper left sternal border, and loud fixed splitting of the second heart sound. In the twelve lead electrocardiogram (ECG) was seen right axis deviation, right ventricular hypertrophy and incomplete right bundle branch block. A chest roentgenogram showed moderate cardiomegaly, mild pulmonary plethora, and a prominent pulmonic vessels. Two dimensional echocardiography outlined a 10-mm sinus venousus type atrial septal defect with left to right

shunting, right atrial and right ventricular enlargement. Selective pulmonary angiograms demonstrated the right superior pulmonary veins entering the vena cava superior vein. There was seen flow of contrast material into enlarged right superior vena cava and right atrium. The right pulmonary veins was opened the superior vena cava. The right and left pulmonary artery angiograms were performed. Contrast injection of the right pulmonary artery was showed prominent pulmonary venous return via superior vena cava and right atrium. The left atrial contrast injection was showed left-to-right shunt about 10 mm in diameter.

The surgical therapy was advised.

DISCUSSION

The natural history of patients with PAPVC is similar to that of the patients with a large ASD of fossa ovalis (5). Chronic congestive heart failure or severe pulmonary hypertension from pulmonary vascular disease usually does not occur until the fourth or fifth decade of life. Younger patients are usually asymptomatic, whereas most of the older patients complain of dyspnea on exertion or exercise intolerance (3). To diagnose PAPVC to superior vena cava is difficult to make by transthoracic or transeosophageal echoardigraphy (6). However, Danilowicz and Kronzon (7) reported that contrast echocardigraphy is useful in diagnosing the presence of PAPVC, particularly when it occurs in association with ASD.

Cardiac catheterization and angiography are often useful in defining the anatomy of PAPVC (8, 9). Increased oxygen saturations in the low superior vena cava gives presumptive evidence, but identification of the specific anatomic details is best accomplished by sineangiography after right pulmonary artery contrast injection. The typical connection of the right upper and middle pulmonary veins to the superior vena cava can often be seen. Surgical treatment of this malformation has become safe (10).

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