A rare association of total anomalous pulmonary venous connection and cor triatriatum

A rara associação de drenagem anômala total de veias pulmonares e cor triatriatum

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Abstract
The association between total anomalous pulmonary venous connection and cor triatriatum is extremely rare. We emphasize the possibility of rupturing the membrane of cor triatriatum in the preoperative cineangiographic study, providing a significant improvement of the clinical features, as well as surgical correction and successful evolution.

Descriptors: Heart defects, congenital, surgery. Heart atrium, abnormalities.

Resumo
A associação entre drenagem anômala total de veias pulmonares e cor triatriatum é extremamente rara. Enfatizamos a possibilidade de rotação da membrana do cor triatriatum no estudo cineangiocardiográfico pré-operatório, proporcionando melhora significativa do quadro clínico, assim como correção cirúrgica e evolução com sucesso.

Descritores: Cardiopatias congénitas, cirurgia. Átrio, cirurgia, anormalidades.

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Article received on June, 2003
Article accepted on July, 2003
INTRODUCTION

The association between total anomalous pulmonary venous connection (TAPVC) and cor triatriatum (CT) is extremely rare [1], and successful corrective surgery has never previously been reported in the national literature.

Generally, TAPVC can be accompanied by patent ductus arteriosus, interventricular shunts, Fallot’s tetralogy, double right ventricle outflow tracts and aortic arch interruption [2]. However, it is necessary to be aware of the possible coexistence of CT, which is characterized by the presence of a membrane that divides the left atrium in proximal and distal chambers, and might have one or more restrictive ostia [3].

We report on this rare association and the possibility of performing a pre-operative cineangiocardiography, thereby rupturing the CT membrane, giving improved hemodynamics, which permits adequate preoperative conditions to perform successful corrective surgery.

CASE REPORT

A male half-caste patient, C.E.V. from Minas Gerais State, Brazil was born prematurely at 32 weeks of gestation. At four days old, he started with vomiting, cyanosis during breastfeeding and persistent tachypnea. He was referred to our department at 1 month and 4 days with 4.2 kg.

Cardiac auscultation presented with rhythmic sounds at two sinus nodes, tachycardia and hyper-phonetic, systo-diastolic murmur +++/6 at the left sternum margin and lungs with vesicular murmur with stertor crackles at their bases. The liver was 3 cm from the right costal edge and the patient suffered from cyanosis of the extremities (saturation of 78%), dehydration and low cardiac output syndrome. The thoracic radiograph evidenced significant pulmonary venous congestion and an increase in the cardiac area, with a cardiothoracic index of 0.78.

The electrocardiogram evidenced significant right ventricular overload suggesting increases in the right atrium and ventricle. The echocardiogram demonstrated TAPVC in the superior vena cava (supracardiac) with pulmonary hypertension, suspecting an association with CT, but it was impossible to give a definite diagnosis due to technical limitations.

The patient was sent for cineangiocardioographic study and for Rashkind’s atrioseptostomy. Although there were difficulties to perform the procedure it was successfully achieved. We believe that rupture of the interatrial septum occurred (Figure 1). The pressure in the pulmonary branch was 42/35/41 mmHg, right ventricle 51/10/15 mmHg, right atrium 12 mmHg and left atrium 7 mmHg. After the procedure, the right and left atria presented with equal pressures of 11 mmHg, hemodynamic improvement (80/40 mmHg to 100/60 mmHg) and oximetry (80% to 93%). The pulmonary branch was dilated, showing confluent pulmonary branches without stenosis. The venous return was abnormal via a common collector tube which emptied in the innominate and superior vena cava through a vertical vein, with a dilated route without signs of obstruction (Figure 2). The left atrium called our attention by presenting a reduced size, without limitations to contrast. No passage of contrast was seen to the right atrium.

The operation was performed two days after the cineangiocardioigram, with deep hypothermia and total cardiac arrest. The time of cardiopulmonary bypass (CPB) was 240 minutes (202 and 38 minutes) and the myocardial ischemia time was 130 minutes, with 9 minutes of total cardiac arrest at 18°C.

The right atrium was opened and an interatrial shunt of a moderate size and with thick edges was found near to the inferior vena cava. Amplifying the interatrial septum...
longitudinally, a fine membrane could be seen that had occluded the mitral valvar orifice, and which presented with a rupture and fissures. This we concluded had occurred during the preoperative hemodynamic procedure. This membrane divided the left atrium in two sections characterizing the presence of CT. After careful resection of this membrane near the mitral valve, it was possible to adequately identify all the portions of the left atrium, which was very small. Thus, we chose an approach by the transverse sinus. On opening the posterior wall of the left atrium, the common pulmonary venous branch (collector tube) was identified, which was opened transversally, thus enabling the anastomosis of the posterior wall of the left atrium with the anterior face of the collector tube. The ligation of the vertical vein was performed near to the innominate vein.

Concluding the main operating phase, the patient was maintained hemodynamically stable during 20 minutes, until he presented with arthrythmia and required CPB support for another 38 minutes to correct metabolic disturbances and optimization of the isotropic drugs.

In the postoperative period, the patient developed with an excess of pulmonary secretion, pneumonia and superficial dehiscence of the wound of the operation, treated with specific antibiotic therapy and was released from hospital on the 19th postoperative day.

Six months after the corrective surgery, the child presented with functional I (NYHA) without medications.

In conclusion, in patients with aforementioned clinical symptoms it is necessary to consider the diagnosis of CT, even if the complimentary examinations do not clearly demonstrate this association. The possibility of the utilization of cineangiographic studies in ruptured membranes is essential for the identification of the clinical condition contributes highly and a complete operative correction optimizes the result.
BIBLIOGRAPHIC REFERENCES


