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Pericardial agenesia: case report from the Fundación Cardiovascular de Colombia.

Clinical Case

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SUMMARY.

Introduction. Pericardial agenesia, a non-frequent disease, usually takes an asymptomatic course. However, it ocasionally causes clinical manifestations that can be life threatening to the patient.

Case report. We present a pericardial agenesia case detected during ductal closure surgery and in which the clinical symptoms were probably secondary to the pericardial defect.

Comments. Although, these kind of procedures are usually indicated by partial defects in which a major risk is sudden death due to herniation and trapping, this surgery is based more on the patient's symptoms than on his anatomic defect variant. ***(Rev Biomed 2005; 16:181-184)***

Key words: Congenital heart disease, pericardial, agenesia.

RESUMEN.

Agenesia del pericardio: reporte de caso de la Fundación Cardiovascular de Colombia.

Introducción. La agenesia de pericardio, una entidad poco frecuente, en la mayoría de los casos presenta un curso asintomático. Sin embargo, ocasionalmente causa manifestaciones clínicas que pueden amenazar la vida del paciente.

Caso clínico. Presentamos un caso de agenesia de pericardio que se detectó incidentalmente durante una cirugía de cierre de ductus y en el cual sus síntomas clínicos se podrían considerar secundarios al defecto.

Comentarios. A pesar que este tipo de procedimientos están usualmente indicados en la corrección de defectos parciales en los cuales hay un mayor riesgo de muerte súbita debido a herniación y encarcelamiento, esta cirugía se realizó con base más en los síntomas del paciente que en su defecto anatómico. ***(Rev Biomed 2005; 16:181-184)***

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Palabras clave: Enfermedad cardiaca congénita, agenesia de pericardio.

INTRODUCTION.

The pericardium is a fibro-serose structure conformed of two layers: visceral and parietal, between which the heart and the large great vessels are found. The visceral pericardial, also called, epicardial, is narrowly linked to the heart surface. The parietal, on the other side, is separated from it by a capilar space that contains pericardic liquid. The pericardium has many functions, such as: the maintenance of the heart`s position in the mediastine, preventing its movement, and avoiding large vessel torsion; reducing the friction between the heart and other adjacent structures during cardiac activity, the avoidance of distension of the cardiac cavities due to volume overcharge; and, probably, the delay of infections diseminating from lungs and pleural cavities to the heart (1). Eighty percent of partial or total pericardial agenesia cases occur in the left heart, 9% are bilateral and all of them are related to the premature atrophy of the common cardinal vein or the Cuvier conduct during the embriological development phase. Additionally, in 30% of the patients, cardiac or pulmonary congenital defects coexist (persistent ductus arteriosus (PDA), atrial septal defect (ASD), mitral stenosis, sequestration, bronchogenic cysts, diaphragmatic defects) (2, 3). Although pericardial congenital defects are usually asymptomatic, they occasionally produce some clinical symptoms that vary from precordial pain to sudden death, especially in partial defects which tend to produce herniation or trapping of the left atrial appendix through the defective zone, torsion of the large arteries or constriction of the coronary artery in the defective ring (2, 4, 5).

Case report.

A four month old child, born by cesarean section due to pregnancy-induced hypertension. Perinatal history of prematurity (29 gestational weeks), weight 1150 g; 4-6-8 apgar, at the first, fifth, and tenth minute, respectively. Because of respiratory distress and hyaline membrane disease, he received two doses of surfactant and required mechanical ventilation for one day and suplementary oxygen for five. After confirmation of clinical stability, he was discharged fifteeen days after admission. At two months of age, he got a flu and presented progressive respiratory difficulty, requiring hospital treatment with antibiotics (Oxaciline and Ceftriaxone) and mechanical ventilation. Radiographic signs of cardiomegaly and echocardiographic anormalities: 7.8 mm atrial septal defect (ASD: ostium secundum) with left to right short-circuit, double pulmonary lesion with slight stenosis and II grade regurgitation, 1.5 mm PDA in the pulmonary extreme with left to right short-circuit and moderate pulmonary hypertension; forced to start anti-congestive management. Then, gastroesophageal reflux disease was needed to be ruled out because of multiple episodes of severe bronchial obstruction and radiological signs of migratory atelectasias and hyperinsuflation during his hospitalization. Due to his negative clinical evolution and the presentation of one episode of apnea, ventilatory support with positive pressure was required. Finally, at the age of four months the patient was transfered to our institution for surgical correction of his cardiovascular defect (PDA).

Admitted with oxygen support, control echocardiogram showed no significant changes. After an episode of crying and bradicardia during his second day of admission, he needed to be intubated. Once stabilized, PDA surgical correction was performed. As an incidental finding during surgery, total absence of the left and partial absence of the right pericardium were found. During the post-surgical period there was the need for mechanical ventilation due to severe and repeated bronchoespasm episodes. Episodes characterized by irritability, bradicardia, and low oxygen saturation were also frequent. Thorax computerized tomography (CT) showed an image compatible with atelectasia in the posterior segment of the left pulmonary lobe, ruling out the probability of pulmonary sequestration. Once stabilized, at 5 months

and twenty days of age, ASD correction and heart fixation was performed: under cardiopulmonary bypass (CPB) and hypothermia (30°C), an auricular incision was done longitudinally to the right atrioventricle groove and 10 mm ostium secundum defect was corrected. Subsequently, the pericardial defect was closed using Marlex mesh, which was attached to the anterior left paraesternal ribs with sutures over the anterior face of the diaphragm. During the post-surgical period the need for mechanical ventilation arose due to bronchoespasm and respiratory difficulty syndrome. Fourteen days after surgery and the after having controlled his arterial pressure, the patient was discharged with home oxygen support and medical treatment (Furosemide, Espironolactone, and inhalators). In medical control, twenty days after discharge, his mother reported a significant reduction in the number and severity of his signs of hemodynamic instability (cyanosis, bradicardia) associated to irritability but persistence of respiratory symptoms (roncus and sibilances). He was finally transferred to specialized neumology consultation for treatment of his respiratory complications.

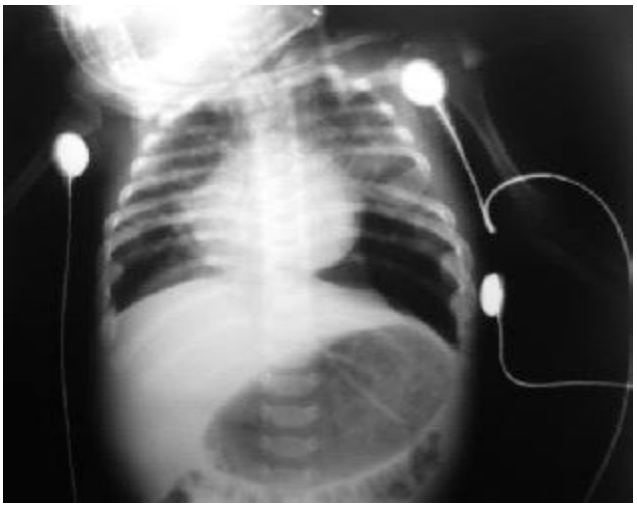

Figure 1.- Thorax X rays before PDA corrective surgical procedure. In the retrospective evaluation of the image, we observed the interposition of the pulmonary parenchima between the heart and the left diaphragm.

COMMENTS.

This patient, in addition to his pulmonary symptoms interpreted as obliterant bronquiolitis sequelae by the Neumology Department, presented episodes of hemodynamic instability and low oxygen saturation that did not disappear after surgical PDA correction. We consider these symptoms and clinical signs of compromise in the venous return system and pulmonary colapse, probably secondary to the torsion of the vascular pedicle (heart herniation on the left side and vena cava angulation). The improvement in the patient´s clinical condittion after the correction of the pericardial defect is in agreement with previous reports in adults who obtained a complete or considerable improvement of their clinical symptoms after this intervention (4).

In this particular case, diagnosis was incidentally done during the PDA corrective surgical procedure. Nevertheless, it is interesting to note that in the retrospective evaluation of the thorax X rays, characterisctic signs of this congenital defect such as the interposition of the pulmonary parenquima between the heart and the left diaphragm were found (figure 1) (4). Additionally, an unusual window for the four cavities and a paradoxical ventricular septal movement were observed in the echocardiogram. Due to the unsuspected pericardial defect before the surgical intervention, no other diagnostic tests such as magnetic resonance imaging (MRI), which has been considered the most sensitive exam due to its superior definition of soft tissues and anatomic structures (6, 7), were performed. However, these kinds of procedures are usually indicated for partial defects in which a major risk of sudden death due to the herniation and trapping exists, this surgery was based more on his symptoms than on his anatomic defect variant (6).

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