

Total Agenesis of the Left Pericardium

J. Glauco Lobo F^o, Ricardo R. Maia Oliveira, Rafael Pontes de Siqueira, Rodrigo M. Landim, João Marcelo A. C. de Albuquerque, Carlos Bellini G. Gomes, Elita Borges, Maria Cláudia Leitão, J. Nogueira Paes Jr

Fortaleza, CE - Brazil

This is the report of a 46-year-old patient with the preoperative diagnosis of an atrial septal defect (ASD) of the ostium secundum type. After sternectomy, partial agenesis of the left pericardium was diagnosed. It is our opinion that, if the radiographic picture is suggestive of this entity, a clinical search for cardiopulmonary anomalies should be performed, because the majority of these associated anomalies can and should be surgically corrected.

Total agenesis of the left pericardium is a rare condition; no more than 200 cases have been reported in the medical literature. There is a male predominance (3:1), and one third of the cases seem to be associated with other cardiopulmonary anomalies, making the present case of the association of total absence of the left pericardium and ASD a very rare entity. Seventy-five percent of the congenital anomalies of the pericardium usually happen on the left side and are related to a premature atrophy of the left cardinal vein (Cuvier's duct) and compromise the circulation to the left pleuropericardial membrane.

Case Report

A 46-year-old male presented with fatigue on minor effort. No relevant clinical or surgical data were found in his medical history.

On physical examination he had signs of right heart failure, with enlargement of the jugular vein, palpable liver below the right costal border and edema of the legs. The examination of the thorax showed left deviation of the *ictus cordis*. A 4/6 systolic murmur caused by an ASD was heard on the left parasternal border. Another systolic murmur, caused by tricuspid regurgitation, was also detected at the fourth intercostal space in the left parasternal region. Cardiac rhythm was normal.

Routine laboratory examinations were normal. The chest X-ray showed global enlargement of the cardiac silhouette (2+/4+) and levoposition of the heart (fig. 1). Electrocardiogram showed normal sinus rhythm, with right

deviation of the axis, signs of enlargement of the right chambers and incomplete right bundle branch block. The echocardiogram showed *situs solitus* in levocardia, severe dilation of the right atrium, ASD of the *ostium secundum* type measuring 24 mm (fig. 2), dilation of the tricuspid ring, enlargement of the right ventricle and paradoxical motion of the ventricular septum. Because of the patient's age and because of the fact that he was a heavy smoker and his pulmonary artery pressure needed to be measured, the patient underwent cardiac catheterization and coronary angiography in order for the physicians to better plan surgery. In addition to the above-mentioned problems, a 90% obstruction was detected at the proximal right coronary artery. The patient was then sent to surgery for correction of the ASD together with coronary artery bypass grafting.

After sternectomy, it was noted that the pericardium was absent (fig. 3). The heart was deviated to the left and there was no adherence between the left ventricle and the pleura. The right coronary artery was significantly calcified. The diaphragm was normal at inspection. Pericardial borders were smooth and, in contiguity with the posterior left pericardium; fibrous-adipose tissue was found that was crossed over by the left phrenic nerve. Right pericardium and the right phrenic nerve were normally positioned. After extracorporeal circulation was started, with the heart still, the ASD was corrected and then revascularization of the right coronary artery was performed using a saphenous magna graft. No per- or postoperative complications occurred, and the patient was discharged seven days after surgery.

Discussion

Congenital pericardial defects are uncommon and frequently asymptomatic, being diagnosed at *post-mortem*². Congenital total absence of the left pericardium represent 57% of all congenital defects of the pericardium. Approximately 78% of all of these defects occur on the left side and have been related to the premature atrophy of the left common cardinal vein or left Cuvier's duct^{2,4}.

The radiographic findings are levoposition of the heart and an elongated left border of the heart¹. There is also an area of hypotransparency between the aortic arch and the pulmonary artery, which is the result of the interposition



Fig. 1 - Chest X-ray in the antero-posterior position.



Fig.2 - Enlarged heart and the ASD. Levoposition of the heart. AD- right atrium; AE- left atrium.



Fig. 3 - Aspect of the heart during surgery showing the absence of the left pericardium.

of the pulmonary tissue at this level. Retrospectively, these findings could be found in the present case.

In this case, the phrenic nerve was not located at its normal position but inside a fibrous-adipose tissue that was connected to the posterior border of the right pericardium. Because the phrenic nerve is usually malpositioned in these cases of congenital anomalies of the pericardium, it is important to carefully identify it during surgery in order to avoid causing accidental lesions to it.

In conclusion, we believe that, in the presence of radiographic suggestions of pericardial abnormalities, a clinical search for cardiopulmonary anomalies should be systematically performed, even if the patient is asymptomatic, because most associated findings can and should be corrected.

References

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