A Case of Partial Congenital Absence of Pericardium Revealed by MRI

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Congenital absence of the pericardium (CAP) is an uncommon finding, and although usually diagnosed incidentally, this condition nevertheless can be symptomatic. We present a case of symptomatic partial pericardial defect in which MRI provided full information for its recognition and definition before surgical correction.

A 63-year-old male smoker with hyperlipidemia was referred for a coronary angiogram following recent typical exertional angina associated with positive echocardiography stress test results showing apical ischemia. The coronary angiogram showed a curious mid-left anterior descending (LAD) distortion associated with an apical left ventricular aneurysm (Figure 1); therefore, we decided to perform an MRI.

We used a 1.5-T MRI scanner with a 6-element phased-array coil. Standard ventricular function examination was performed by acquiring cine loops of standard views with the use of a steady-state gradient-echo technique. Cardiac and pericardial morphology was studied using breath-hold T1-weighted fast-spin echo MRI in the cardiac axial and short-axis planes, and contrast-enhanced images were acquired 10 minutes after IV administration of gadolinium contrast agent. MRI showed the absence of pericardium at the apex and biventricular apical herniation (Figures 2 and 3); these findings led us to diagnosing partial CAP, the constriction of the last part of the mid-LAD that was explained by the rim of the pericardium. Complete definition of the CAP using MRI, LAD compression with apical ventricular herniation, and proven ischemia led to surgical intervention (Figure 4A and 4B). Successful pericardioplasty combined with LAD bypass using the left internal mammary artery was performed. The immediate postoperative MRI showed clearly improved apical movement associated with the successful pericardioplasty result. Control coronary angiogram showed good permeability of the left internal mammary artery and partial disappearance of the LAD distortion (Figure 5). The patient currently is event free after 1-year of follow-up.

Figure 1. Coronary angiogram showing a very tight stenosis (arrow) of the distal part of the mid-LAD artery associated with uncommon distortion of the artery.

Figure 2. Axial T1 MRI of the heart revealing the partial defect of the left pericardium.
CAP is a rare condition ranging from small defects to complete agenesis. Matthaeus Realdus Columbus is credited with the first description in 1559. The first fully documented case was reported by Baille in 1793, and in 1936 Ladd was the first to report the condition in a living subject.\cite{1} CAP is due to premature atrophy of the left common cardiac vein (Cuvier vein), with insufficient blood supply leading to its agenesis. This theory accounts for the majority of the pericardial defects located on the left side,\cite{1,2} the most common being the complete absence of the left side of the pericardium. Complete absence and partial left or right absence are very rare.

CAP can be isolated but also may be part of a more severe or complex cardiac malformation with its own symptoms (patent ductus arteriosus, atrial septal defects, mitral valve stenosis, aortic bicuspid valve, and tetralogy of Fallot) as well as may be associated with extracardiac abnormalities, such as bronchogenic cyst, pulmonary sequestration, and diaphragmatic hernia,\cite{2} or found in other syndromes, such as VATER (vertebrae, anus, trachea, esophagus, and renal) syndrome, Marfan syndrome, and Pallister-Killian syndrome.\cite{3}

The gated cardiac MRI with T1-weighted morphological sequences has good soft tissue contrast resolution that is able to delineate the fine, low-signal intensity of the normal pericardium. Most pericardial defects and their extent can be detected by MRI, except in patients with decreased pericardial fat in whom misdiagnosis can occur.\cite{3} Surgical intervention consisting of the liberation of the fibrous ring and pericardioplasty is required for partial CAP to prevent strangulation of any herniating cardiac...
structures (left atrium, left atrial appendage, left ventricle) through the defect and compression of coronary arteries. Furthermore, symptoms related to CAP (as in our case) indicate surgical correction. However, very small defects and complete CAP with no fatal potential require no intervention. In the case reported here, the associated LAD bypass was decided by the surgeon mainly because there was persistent distortion of the LAD even after opening the pericardium.

Disclosures
None.

References

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