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Brief Report

Absent pericardium: at risk for endocarditis?

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Abstract A 16-year-old girl with no previous cardiac problems presented to the casualty with septic shock. Investigations revealed endocarditis involving the mitral valve. At the time of surgery, she was noted to have a large left atrial appendage herniating into the left pleural cavity due to partial absence of the pericardium. Complete or partial absence of the pericardium is an uncommon congenital anomaly. Though various complications and presentations are reported, it has not been identified as a risk factor for endocarditis. We discuss the possible mechanism, and need for prophylaxis against endocarditis, in patients with absence of the pericardium.

Keywords: Left atrial appendage; infection; mitral valve

Congenital absence of the pericardium is a rare clinical entity. In isolation, it encompasses a range of defects, from a small hole to a complete absence of the entire pericardium.[1] Despite its rarity, it has been reported for more than 400 years.[2] We report a rare complication of absent pericardium and discuss its possible mechanism.

Case report

A 16-year-old girl was admitted in casualty with a history of fever, diarrhoea, vomiting and headache. She had been well until the day before admission. Clinical examination showed prolonged refilling of the capillaries. She was resuscitated with fluids and inotropic support, and intravenous antibiotics were commenced having obtained appropriate samples of blood for culture. Her past medical history was unremarkable, apart from a dental extraction four weeks previously. There was no history of intravenous drug abuse. Clinically, she had a pan-systolic murmur at the apex, and hepatosplenomegaly.

The inflammatory markers, notably C-reactive protein, were raised. The chest X-ray showed shouldering of the left atrial appendage (Fig. 1) A 12 lead electrocardiogram was normal. The echocardiogram showed a moderately dilated left atrium and left ventricle. There was a mobile mass attached to the aortic leaflet of the mitral valve that prolapsed into the left atrium and caused severe mitral regurgitation. Ventricular function was normal.

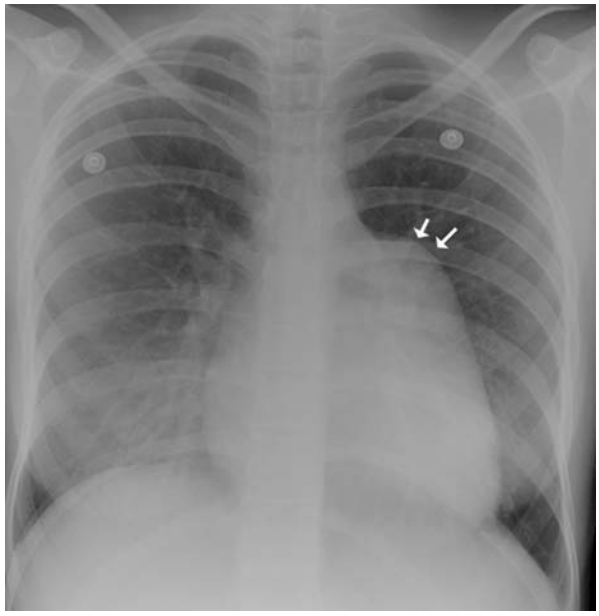

Figure 1.
The chest radiograph, as seen in the antero-posterior projection, shows shouldering of the left atrial appendage (white arrows), a characteristic feature of absence of the pericardium.

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The diagnosis of endocarditis involving the mitral valve was confirmed by transoesophageal echocardiography. She underwent surgery, at which stage she was noted to have partial absence of the pericardium. The left atrial appendage was very large, and herniated into the left pleural cavity. A vegetation was found on the ventricular aspect of the aortic leaflet of the mitral valve. The vegetation was excised, and a sliding plasty was done between the lateral and central scallops of the mural leaflet. As there was still mitral regurgitation, with some prolapse of the central aspect of the aortic leaflet, we inserted an Alfieri stitch, with good result. The large left atrial appendage was ligated. She made a good recovery after the surgery. Antibiotics were continued for four weeks. The cultures from blood and tissues were all negative.

Discussion

Isolated absence of the pericardium in symptomatic patients tends to be described as single case reports. In one-third of cases, however, congenital absence of the pericardium is associated with other cardiac lesions, such as patency of the arterial duct, mitral stenosis, tetralogy of Fallot, and others.[3,4] Congenital enlargement of the left atrial appendage, in contrast, is often difficult to diagnose, and is extremely rare.[5]

The diagnosis of partial or complete absence of the pericardium is usually made incidentally at postmortem examination, during other intrathoracic operations, or from abnormal chest radiograms. The most common clinical presentation is periodic and stabbing chest pain that mimicks coronary arterial disease. Patients with complete absence of the pericardium may present with debilitating symptoms. Other symptoms include dyspnoea and a sensation of "shifting heart".[6]

Physical findings are often unhelpful, but a left ventricular heave may be felt when the deficiency is substantial. The radiographic finding of pulmonary tissue interposing between the pulmonary trunk and the aorta is pathognomic. Resonance imaging can, in some cases, confirm the diagnosis. Surgical procedures employed for patients with congenital absence of the pericardium with debilitating symptoms included left atrial appendectomy, division of adhesions, pericardiectomy, extension of the defect, and pericardioplasty. Elective pericardioplasty may offer symptomatic relief.[6]

As far as we are aware, infective endocarditis has never been reported in the setting of congenital absence of the pericardium. The pericardium consists of an outer fibrous and an inner serous layer. It serves as the potential barrier for spread of infection from adjacent structures. Absence of the pericardium causes traction of the left atrium into the pleural space, which leads to distortion of the left atrial anatomy, and possibly that of the mitral valvar annulus, potentially leading to mitral regurgitation. Indeed, tricuspid insufficiency has recently been found in association with partial absence of the right pericardium.[7] Valvar insufficiency predisposes to a high risk for developing infective endocarditis. It is advisable to protect such patients with antibiotics for potential bacteraemia.

Our experience, therefore, alerts us to a rare but serious combination, namely absence of the pericardium, gross atrial dilation, and endocarditis resulting from mitral valvar distortion in a previously healthy teenage girl. Awareness of these previously unreported combinations is useful for detection, and in prevention of associated serious complications.

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