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Brief Report

An epicardial cyst in a child

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Abstract An 8-year-old girl, without any previous medical history, presented with a first short syncope. Physical examination was unremarkable. Transthoracic echocardiography revealed a thin-walled, echo-free cystic structure adjacent to the posterior wall of the left ventricle, and compressing it moderately. Other echocardiographic findings were normal. Both computed tomography and magnetic resonance imaging suggested a simple pericardial cyst, but during surgery we found an epicardial cyst with partial involvement of the circumflex branch of the left coronary artery. Cardiopulmonary bypass was necessary for successful resection of the cyst, leaving behind only the small area in continuity with the coronary artery.

Keywords: Epicardial cyst; successful resection; child; syncope

Cystic structures within the pericardial cavity are rare. They are divided into epicardial and pericardial variants. The latter are the more frequent congenital abnormalities, accounting for 7% of all mediastinal tumors.[1] Such pericardial cysts are usually asymptomatic and unsuspected findings, discovered in the right or left cardiophrenic angles on routine chest radiographs.[2,3] They are mostly unilocular, round or elliptic, with thin and smooth walls, and are filled with clear liquid.

In contrast, there are only three reports of epicardial cysts.[4–6] In 1972, Edwards and Ahmad[4] reported the first successful removal of an epicardial cyst, with two further reports following in 1985 and 1991.[5,6] We present a patient in whom the cyst partially involved the circumflex branch of the left coronary artery.

Case report

An 8-year-old girl was referred to our institution via an external hospital for further examination after a first short syncope. She lost consciousness for about a minute after standing in her school room in the morning. The teacher observed no seizure-like activity, and the girl was able to provide the telephone number of her mother immediately after the syncopal episode in accordance with undisturbed long-term memory. On the other hand, her short-term memory was impaired, and she was confused for a few hours, asking the same questions several times. She had no previous history of cardiac or neurologic disease. Physical examination was unremarkable. Routine laboratory tests on admission showed values within normal range for red and white blood cell count, platelets, blood gases and glucose, lactate, serum electrolytes, total protein, liver enzymes, creatinine and creatine kinase. Chest and skull radiographs were normal, as were the 12-lead surface electrocardiogram, Holter monitoring, and electroencephalography. Cross-sectional transthoracic echocardiography revealed a thin-walled, echo-free cystic structure, measuring 5 by 5 centimetres, adjacent to the posterior wall of the left ventricle, and producing moderate ventricular compression (Fig. 1a). Other echocardiographic findings were normal, as were ultrasonography of liver, spleen and kidneys. Both chest computed tomography (Fig. 1b), and magnetic resonance imaging, were performed to examine the characteristics of the cystic mass in detail. The contour was smooth, and its density homogeneous. The T1-weighted spin-echo image showed the lesion with slightly lesser intensity than the myocardium

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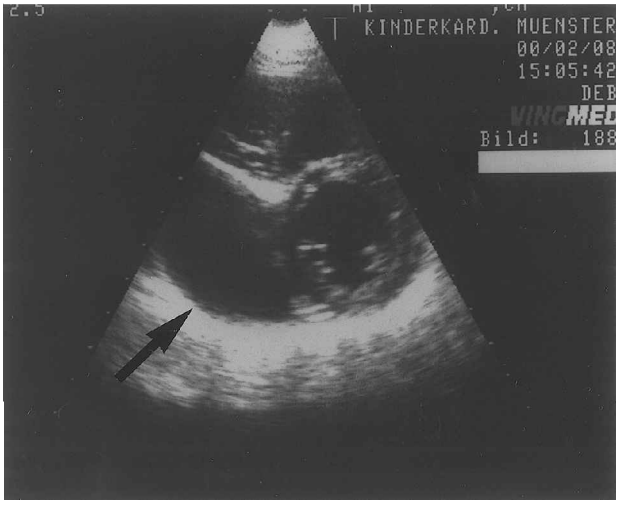

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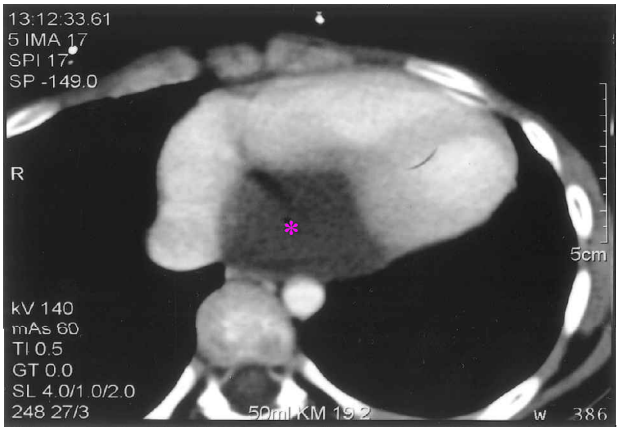

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Figure 1.
Cross-sectional transthoracic echocardiography. The short-axis view at papillary muscles level (a) shows the echo-free cystic structure (↑) adjacent to the posterior wall of the left ventricle. Chest computed tomography after application of 50 ml contrast medium (b) shows the epicardial cyst (*), R = Right.

and distinct from it. High intensity and homogeneity of the lesion on the T2-weighted spin-echo image lead to the diagnosis of a simple pericardial cyst. Surgical removal of the cyst was indicated, and the patient was explored through a median sternotomy. The pericardium appeared normal and was split lengthwise. Exploration surprisingly revealed an epicardial cyst at the posterior wall of the left ventricle, with no adhesion between the cyst and the pericardium. Cardiopulmonary bypass was necessary for excision, particularly because of involvement of the circumflex branch of the left coronary artery and its corresponding vein in the cyst, which were lifted from the myocardium over a short distance (Fig. 2). After aspiration of clear fluid, the free wall of the cyst was excised carefully, leaving behind only the small

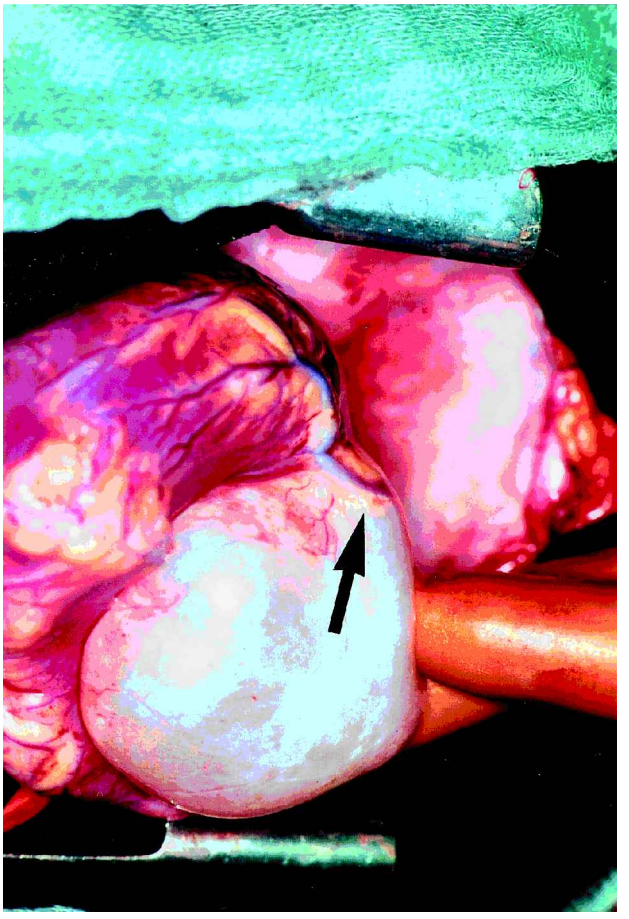

Figure 2.
Exploration behind the heart revealed the epicardial cyst at the posterior wall of the left ventricle, with involvement of circumflex branch of the left coronary artery and its corresponding vein, which were lifted from the myocardium over a short distance (↑).

area in contact with the coronary artery. Resection revealed an area of the myocardium uncovered by epicardium. The remaining edges of the cyst were plicated over the venticular wall, covered with fibrin and collagen. Pathologic examination of the specimen showed a mesothelial-lined cyst overlying a thin layer of fibrous tissue. There were no areas of acute or chronic inflammation. Intraoperative microbiological samples were negative, and cytologic studies revealed no malignant cells. The child recovered without any complication, and she was discharged on the tenth postoperative day. Follow up during the next year was uneventful.

Discussion

To our knowledge, this is the second report of a symptomatic epicardial cyst in a child. Such cysts in the pericardial cavity, being attached to the heart without adhesion to the pericardium, seem to be

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exceedingly rare. The first successful removal was described in 1972,[4] and only two further cases have been reported.[5,6] In our case, the cyst involved the circumflex branch of the left coronary artery, being diagnosed at surgery, as in the previous instance involving a 7-year-old girl.[5]

The epicardium is the visceral layer of the serous pericardium. The pericardial celom is the most precocious of the body cavities, and arises from a series of disconnected mesenchymal lacunas within five weeks of fertilization.[7] The theory of failed coalescence of pericardial lacunas[7] may be an acceptable explanation for the etiology of both epicardial and pericardial cysts, but all in all, little is known about etiology of such cysts inside the pericardial cavity.

Epicardial cysts are unilocular, round or elliptic, with thin and smooth walls, and filled with clear liquid, comparable with pericardial cysts. Epicardial cysts, however, have no adhesion between the cyst and the parietal pericardium, which tends not to be noticed before cardiac surgery. The cysts described previously ranged in diameter from 3 to 9 cm, and contained up to 0.5 litre of clear serous fluid.[4–6]

Patients with either epicardial or pericardial cysts mostly remain asymptomatic.[3] Most cysts, in fact, are unsuspected findings on routine chest radiographs. Symptoms, such as chest pain, dyspnea, and coughing, or serious complications, such as syncope, tend to occur concomitant with enlargement of the cyst.[4–6] This can produce compression of the heart with hemodynamic deteriorations, obstruction to the airways, or erosion of the cardiac wall and vascular structures.[8] Secondary infections may produce an inflammatory process, which can spread from the cyst to the surrounding tissues. One sudden death, related to exercise, occurred after a stress test in a patient with a large pericardial cyst due to necrotic damage of the myocardium involving the conduction system.[9] We did not perform a stress test in our patient. Symptoms of both pericardial and epicardial cysts are probably explained by their size and localization independent from their etiology.

The differential diagnosis includes large right pericardial fat, ventricular aneurysm or diverticulum, tumours of the heart or pericardium, hydatid, teratomatous, or bronchogenic cysts, diaphragmatic tumours and diaphragmatic hernia.[1–3] Chest radiographs, transthoracic echocardiography, computed tomography and magnetic resonance imaging are useful in diagnosing the cystic lesion, although these techniques will not distinguish between epicardial and pericardial cysts.

Surgical treatment of epicardial cysts is advised. Even though the cyst may be asymptomatic, compression on the surrounding organs, erosion of cardiac wall and vascular structures, and rupture into the pericardial cavity with cardiac tamponade must be avoided.[5] Percutaneous aspiration and ethanol injection into the cyst could be an alternative to surgical resection, as in some cases of proven pericardial cysts,[10] but is not indicated in epicardial cysts because the coronary arteries might be involved, as shown in our patient. In our case, the indication for surgery of the cyst was the syncope with prolonged symptoms. Cardiopulmonary bypass was necessary for excision of the cyst, particularly because of the involvement of the circumflex branch of the left coronary artery and its corresponding vein, which were lifted from the myocardium over a short distance.

Our report emphasizes the possible need for cardiopulmonary bypass during removal of cysts within the pericardial cavity, especially in patients in whom preoperative imaging fails to show distinct margins between the cyst and cardiac structures.

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