Intrapericardial foregut cyst associated with intrauterine death

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Abstract

Intrapericardial developmental foregut cysts are rare and are most frequently incidental findings at necropsy in adults. A 29 year old Asian woman delivered a 24 week stillborn fetus seven days after diagnosis of intrauterine death caused by rupture of a foregut cyst. Multiple cysts occupied the wall of the right atrium and its rupture caused haemopericardium and cardiac tamponade.

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A 29 year old Asian woman (gravida 3, para 3) delivered a 24 week stillborn fetus seven days after diagnosis of intrauterine death. The pregnancy was complicated by gestational diabetes mellitus and hypothyroidism. The fetus was normally developed, weighed 513 g, with a crown-rump length of 23 cm.¹ There was moderate maceration, but no external abnormalities were identified.

At necropsy, the pericardium was distended with 3 ml of clotted blood, with a rupture site visible in the wall of the right atrium. There were no anomalies of the gross anatomy of the heart (3.1 g) or great vessels.

Microscopically, the atrial wall was of variable thickness, with recent and organising haematoma at the rupture site (fig 1). Multiple cysts up to 3 mm in diameter were identified in the atrial wall. These were lined by a columnar epithelium

that was ciliated (fig 2) and showed focal pseudostratification. No inflammatory infiltrate was present. The cysts contained cell debris that was partly calcified. Concentrically arranged smooth muscle, but no cartilage or seromucinous glands, was present beneath the epithelial lining. No neuroectodermal structures such as skin, skin appendages or neuronal tissue were identified. The atrial septum and ventricles were normal. The remainder of the necropsy was unremarkable and the placenta (166 g) showed changes consistent with intrauterine fetal death. Cytogenetic analysis performed on amniotic fluid following diagnosis of intrauterine fetal death showed a normal female karyotype (46, XX).

Discussion

Developmental foregut cysts within the pericardium are rare, with approximately 35 cases reported.² Although most frequently an incidental necropsy finding in adults, some cases have presented with symptoms of pericarditis.^{3–4} They are frequently described as intrapericardial bronchogenic cysts (IPBC) among other terms.² One third of patients with such cysts are infants, who are almost always symptomatic with respiratory distress and cyanosis, the size and location of the cyst causing vascular compromise.^{5–6} IPBC are usually not associated with any cardiac anomalies.⁷ Microscopically, they should be distinguished from the more common teratomas that contain tissues of neuroectodermal derivation such as skin, teeth or glial tissue. As little distinction was made between IPBC and teratomas in the earlier literature on the subject, some of the deaths ascribed to cardiac teratoma may have been due to IPBC.⁸ For example, Jellen and Fisher⁹ reported a case of neonatal death secondary to what they called an intrapericardial teratoma, although the histology suggests that this may have been a bronchogenic cyst.

Cystic tumour of the atrioventricular node has a characteristic location, and as an endodermal derivative may also show ciliated epithelium.² Ciliated epithelial cysts have been reported on the tricuspid valve¹⁰ and in the left ventricle.¹¹ Despite the absence of other bronchial tissues such as cartilage, their histogenesis is similar. Before the obliteration of the dorsal mesocardium between the 7th and 16th somatic stages, the foregut is in close proximity to the developing heart.¹² Sequestration of foregut tissue may occur before fusion of the paired coelomic cavities that become the pericardium, and result in intrapericardial or intramyocardial cystic rests. Those in the myocardium are

Figure 1 Right atrium, with lumen (asterisk) and thinned wall that has ruptured (centre right). Pericardial haematoma is shown on the right, and an intact cyst in the atrial wall on the left of the picture (haematoxylin and eosin; original magnification $\times 25$).

Figure 2 Cyst wall showing ciliated columnar epithelium and circular smooth muscle (haematoxylin and eosin; original magnification $\times 520$).

typically located in the posterior wall of the left ventricle.¹¹ Differences in timing of such misplacement probably account for the varying location and composition of foregut cysts.

Haemopericardium leading to intrauterine death is a rare event. Emboli from placental vessels causing acute myocardial infarction with resultant right ventricular rupture was described by Wolf *et al.*¹³ Other cases have followed a motor vehicle accident¹⁴ and amniocentesis.¹⁵ The case described here is unique in that it is the first in which an intrapericardial foregut cyst was

the cause of intrauterine death. We have not been able to find an account of rupture of the heart secondary to this lesion, but given the histogenesis and frequent presentation in early childhood, it is not altogether surprising that a foregut cyst may cause intrauterine death.

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Gastric outflow obstruction caused by gall stones and leading to death by complex metabolic derangement

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Abstract

A 67 year old woman was admitted with a three week history of vomiting, having become increasingly confused for three days. Investigations revealed deranged serum biochemistry consistent with a combination of a diabetic non-ketotic hyperosmolar state and a metabolic alkalosis consistent with gastric outflow obstruction. She was treated with intravenous saline, intravenous insulin, and subcutaneous heparin, but did not improve clinically and

had an asystolic cardiac arrest the following day; she was transferred to the intensive care unit and despite treatment with inotropes she died 40 hours after admission. Necropsy revealed that the stomach was massively dilated with gas and stomach contents, and contained many small black faceted gall stones. In addition a large non-faceted brown-yellow gall stone was wedged in the pyloric antrum causing total obstruction. The patient had died from a complex metabolic derangement including