CASE REPORTS

Pulmonary artery rupture in pregnancy complicating patent ductus arteriosus

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Abstract

Fatal haemopericardium in a 27 year old pregnant woman was caused by rupture of a dissecting aneurysm of the pulmonary artery. She had an uncorrected patent ductus arteriosus and severe pulmonary hypertension. The wall of the pulmonary artery showed atherosclerosis and cystic medionecrosis.

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Case report

A 27 year old West Indian woman with a patent ductus arteriosus had been clinically well and took no medication. Cardiac catheter studies when she was 13 years old had shown pulmonary hypertension with pressures of 70/40 mm Hg. There was reversal of the shunt during exercise (right-to-left), indicating that she was unsuitable for surgery and therefore the patent ductus had remained uncorrected. On examination, the apex beat was displaced

laterally to the anterior axillary line and there were signs of pulmonary hypertension—loud pulmonary heart sound (P_2) and a palpable pulmonary artery. There was a long early diastolic murmur of pulmonary regurgitation and an ejection systolic murmur. An electrocardiogram showed evidence of right ventricular hypertrophy.

Her first pregnancy, when she was 25 years old, had been uneventful; however, there had been clinical signs of increasing pulmonary hypertension. At 38 weeks' gestation a normal female infant was delivered through the vagina. Immediately post partum the mother developed a pyrexia, which was thought to be endocarditis, and this settled on antibiotics. She was strongly advised to avoid further pregnancy.

The second pregnancy, two years later, was progressing well and she was admitted to hospital for bed rest. She died unexpectedly in bed at 33 weeks' gestation.

Necropsy and histological findings

The body was thin but well nourished (height 163 cm, weight 54.6 kg). There was a large haemopericardium caused by rupture of a dissecting aneurysm of the main pulmonary artery into the pericardial sac at the base of the heart. The main pulmonary trunk was dilated to 5 cm in diameter and the dissection originated in a raised atheromatous plaque on the anterior aspect of the pulmonary artery 3 cm above the pulmonary valve (figs 1 and 2). There was a large patent ductus arteriosus (diameter 2 cm) leading to an aorta of normal calibre. The right ventricle was hypertrophied (100 g). The left ventricle including the septum weighed 160 g. There was no other gross cardiac malformation.

Histological examination of the main pulmonary artery showed a striking accumulation of alcianophilic mucopolysaccharide in the media (cystic medionecrosis) (fig 3) with dissection between the inner and outer halves. There were also small amounts of medial alcianophilic mucin in the aorta. In the lungs there were widespread severe hypertensive changes with plexiform lesions (plexogenic pulmonary arteriopathy).

No other anatomical or histological abnormality of significance was seen and the fetus was normal.

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Figure 1 Necropsy appearance of the heart and pulmonary artery, showing dilatation of the main pulmonary artery and right ventricular hypertrophy. The probe marks the site of pulmonary artery rupture.

Figure 2 Necropsy appearance of the opened main pulmonary artery, showing atherosclerosis. The arterial dissection is seen in the cut edge of the pulmonary trunk (arrow).

Discussion

Pulmonary artery aneurysm is a rare condition and is invariably associated with pulmonary hypertension. In a necropsy review of 111 cases,¹ pulmonary hypertension due to cardiac malformation was present in 66%. Patent ductus arteriosus was the commonest single lesion, occurring in combination with 22% of aneurysms.

When Coleman *et al*² reviewed published reports they found only six cases of ruptured pulmonary artery aneurysm associated with patent, ductus arteriosus alone, in addition to their own. There have been reports of two more cases.^{3,4} We identified only one report of rupture in pregnancy,⁵ however, this was associated with pulmonary infundibular stenosis as well as patent ductus arteriosus. There is also a report of a maternal death occurring 17 hours post partum owing to pulmonary artery dissection associated with a patent ductus alone,⁶ presumably precipitated by labour. Our case is the only report we found of rupture during pregnancy associated with a patent ductus as the only anatomical abnormality.

Cystic medionecrosis is usually associated with dissection of the pulmonary artery³ as is atheroma and our case is no exception. Indeed both are known to be related to severe pulmonary hypertension.

This case emphasises the possibility of arterial dissection in pregnancy, particularly in the presence of pre-existing cardiovascular disease. This association has been well described previously, in particular by Guthrie and MacLean⁷ who showed that nine of 57 non-aortic arterial dissections in women occurred either during pregnancy or shortly after delivery.

It is of interest to speculate on the cause of the apparently increased incidence of both systemic and pulmonary arterial dissections in pregnancy. It has been suggested that the increase in body water in pregnancy is in part taken up by connective tissue mucopolysaccharides.⁸ Robertson pointed out there is no reason why mucopolysaccharides in the vessel

Figure 3 Histological section showing pools of mucopolysaccharide in the media of the pulmonary artery (haematoxylin and eosin; original magnification, $\times 500$).

wall should not be involved in this process⁹; indeed there is some histopathological evidence that they are.¹⁰ We suggest that the likeliest course of events in our case was cystic medionecrosis and atheroma, followed by dissection and rupture. These were consequent upon the increased cardiac output and exaggeration of mucopolysaccharide deposition associated with pregnancy.

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