Aneurysm of the ductus arteriosus

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SUMMARY Two neonates with aneurysms of the ductus arteriosus were seen. One suffered a fatal spontaneous rupture of the aneurysm, and the second (in which it was an incidental finding) the aneurysm was monitored by echocardiography, and seemed to resolve spontaneously.

Aneurysm of the ductus arteriosus in infants is rare and potentially fatal, but often not recognised. It can present with common symptoms such as cough, hoarseness, and dyspnoea and should be included in the differential diagnosis of all mediastinal masses. The aims of this paper are to highlight the disease and to show the value of echocardiography in establishing the diagnosis. We hope that improved diagnosis will permit appropriate treatment to be instituted and fatal complications avoided.

Case reports

CASE 1

A 3 week old boy who had been previously well presented with a cough and hoarse cry. An upper respiratory tract infection was diagnosed and treated. His symptoms were stable for six days, but

then deteriorated with increasing respiratory distress. On admission to hospital he was ill, cyanosed, and pale. He had a tachycardia, was tachypnoeic and had widespread crepitations in both lungs. A chest radiograph showed an enlarged heart and opacities in the left upper zone. Arterial blood sampling showed that he was hypoxic (oxygen saturation 76%), with a metabolic acidosis (pH 6.97), and a haemoglobin concentration of 98 g/l. Shortly after admission he had a cardiopulmonary arrest; he was resuscitated and transferred to the cardiac intensive care unit. He was moribund with a blood pressure of 30/20 mm Hg and his haemoglobin concentration had fallen to 42 g/l. A repeat chest radiograph showed complete opacification of the left hemithorax with the mediastinum shifted to the right.

Echocardiography was performed using the UM4 (ATL) echo/Doppler equipment. The intracardiac anatomy was normal. There was a moderate pericardial effusion and a large collection of blood in the left hemithorax. On the suprasternal view a thick walled cystic mass 3 cm in diameter was seen. It displaced the left pulmonary artery anteromedially (fig 1a), and the distal aortic arch posterolaterally (fig 1b).

The pericardial effusion was tapped, and 30 ml of

Fig 1 (a) Suprasternal view showing left pulmonary artery (LPA) displaced anteromedially by the thick walled ductal aneurysm (DA). MPA=main pulmonary artery. (b) Posterolateral displacement of the aortic arch (AoA) by the ductal aneurysm (DA), with more than usual separation of the left pulmonary artery (LPA) and distal arch.

Fig 2 (a) Fusiform ductal aneurysm (DA) posterolateral to the left pulmonary artery (LPA) and main pulmonary artery (MPA). (b) Repeat echocardiogram five days later. Note pronounced reduction in the size of the ductal aneurysm (DA) compared with the main pulmonary artery (MPA) and left pulmonary artery (LPA).

haemoserous fluid was drained with a resulting temporary rise in blood pressure and reduction in pulse rate. The improvement could not be maintained, however, despite large infusions of colloid and blood. A left pleural tap drained fresh blood. Because the wall of the cyst was so thick (fig 1a) a bleeding mediastinal tumour was suspected and an emergency left thoracotomy was carried out. After evacuating a large amount of fresh blood clot there was exsanguinating haemorrhage from the region of the ductus arteriosus. The ductus had disintegrated and the aortic and pulmonary ends were open and bleeding. Attempts to control the haemorrhage failed. Histology of biopsy specimens taken from the region of the ductus showed laminated organised thrombus, which supported a diagnosis of ruptured aneurysm of the ductus arteriosus.

CASE 2

A baby boy was born at full term by emergency caesarean section for fetal distress. There was meconium stained liquor, profound bradycardia, and the baby made no respiratory effort. He was slow to respond to resuscitation and required continued intermittent positive pressure ventilation. On physical examination there was a left renal mass (later shown to be due to renal vein thrombosis) and a soft cardiac murmur. Chest radiograph showed bilateral upper zone opacification, worse on the left. Echocardiography showed normal intracardiac anatomy. There was pronounced fusiform dilatation

of the ductus arteriosus (fig 2a) with flow from the aorta to the pulmonary artery shown on Doppler scanning, confirming ductal patency. At its widest diameter, the ductus was three times the size of the distal aortic arch. Daily echocardiograms showed progressive diminution in the size of the aneurysm, associated with closure of the pulmonary end (fig 2b). After a week the aneurysm was only slightly bigger than the distal arch, and there was no demonstrable flow shown on Doppler scanning. His clinical condition progressively improved and the left upper zone changes shown on chest radiography diminished in association with the reduction of the aneurysm size on echocardiography. Continued cardiac and renal surveillance is planned.

Discussion

These cases represent the extremes of a rare disease that is not often recognised. Spontaneous aneurysm of the ductus arteriosus is recognised in two groups: young infants and adults. The infant type is seen most commonly under 2 months of age and usually consists of a fusiform dilatation of the ductus with an open aortic end and a partially or completely closed pulmonary end.¹ It is usually an incidental finding at necropsy.² When symptoms occur they are commonly associated with recurrent laryngeal nerve palsy with cough, hoarse cry, and inspiratory stridor. Rarer presentations include phrenic nerve palsy or thromboembolic complications.^{2,3} Spontaneous

rupture and dissection has been described and was the probable cause of death in five of the 51 cases cited by Cruickshank and Marquis.⁴ It has been assumed that aneurysm of the ductus may resolve spontaneously as thrombus within it becomes organised and fibrosed, but there is no documented evidence for this. In view of the possible fatal complications, therefore, surgical treatment is advocated. Heikkinen and Simila were the first to describe successful elective resection of the aneurysm in two neonates.³

Greater awareness of the condition could have facilitated a preoperative diagnosis in case 1. It was impossible to achieve vascular control at left thoracotomy but the use of cardiopulmonary bypass, hypothermia, and circulatory arrest would certainly have helped in achieving haemostasis. Review of reported work showed only one successfully treated ruptured aneurysm.⁵ We recommend the use of cardiopulmonary bypass in suspected cases of aneurysmal rupture.

Because of our experience with case 1, the diagnosis was easily made with the use of echocardiography in case 2. The presence of a cystic mass in continuity with the left pulmonary artery and the aorta was unmistakable. Provided one maintains a high index of suspicion and includes aneurysm of the ductus in the differential of all mediastinal masses, the diagnosis can accurately and confidently be made on echocardiography. Further investigations, particularly angiography (which may jeopardise an already critically ill infant), are unnecessary.

In the second case the early natural history of the condition was documented. The rapid reduction in the size of the aneurysm led us to question the need for surgical intervention in all cases. A conservative policy of closely monitoring the clinical progress and

echocardiographic appearances was adopted. If the aneurysm had not undergone rapid resolution or had increased in size we would have resected it because of the risks of endocarditis, rupture, and compression of adjacent structures.

We recommend urgent operations using cardiopulmonary bypass if a ruptured aneurysm is suspected. If there is no evidence of rupture, however, we believe a conservative policy of monitoring the progress of the aneurysm by echocardiography is justified. If the aneurysm increases in size or does not undergo rapid resolution we recommend urgent resection.

Addendum

Since submitting this paper we have recognised two further cases. One resolved spontaneously and the other was corrected during cardiac surgery for pulmonary atresia.

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Ciprofloxacin in neonatal *Enterobacter cloacae* septicaemia

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SUMMARY Multiresistant *Enterobacter cloacae* infection in six premature infants was eradicated with intravenous ciprofloxacin (10 mg/kg/day). Bacterial resistance did not develop. Adequate plasma ciprofloxacin concentrations were achieved in all treated patients. No clinical evidence of side effects was observed.

Modi *et al* have documented an outbreak of multi-resistant *Enterobacter cloacae* infection that was observed in this regional intensive care unit between July 1985 and January 1987.¹ The organism was resistant not only to penicillins and aminoglycosides but also to third generation cephalosporins. Later in 1987 a further six infants developed septicaemia due to the same multiresistant strain of *E cloacae* that showed in vitro sensitivity to ciprofloxacin (Ciproxin,