Fistulous aneurysm of ductus arteriosus*

KENNETH M BOROW, SAMUEL J HESSEL,† LAURENCE J SLOSS

From the Cardiovascular Division, Department of Medicine, the Cardiothoracic Division, Department of Surgery, and the Department of Radiology, Peter Bent Brigham Hospital, and the Department of Cardiology, Children's Hospital Medical Center, and the Department of Pediatrics, Harvard Medical School, Boston, Mass, USA

SUMMARY We present clinical, haemodynamic, and angiographic data on a 48-year-old woman with a fistulous aneurysm of the ductus arteriosus. This is the first reported case of a spontaneous fistulous aneurysm of the ductus arteriosus in an adult in which an antemortem diagnosis was made. This lesion typically presents with shortness of breath, hoarseness caused by recurrent laryngeal nerve paralysis, and chest pain, and poses a critical threat to life because of progressive compression, erosion, and haemorrhage into the oesophagus or tracheobronchial tree. Fistulous aneurysm of the ductus arteriosus should be considered in the differential diagnosis of an enlarging mediastinal mass, especially when a continuous heart murmur is noted. Early definitive diagnosis and surgical repair in adults are essential for survival.

Dissecting aneurysm of the ductus arteriosus is a rare cause of an enlarging mediastinal mass. This lesion is life threatening, and requires emergency surgical intervention. The following report details the non-invasive and angiocardigraphic evaluation of an adult patient with a fistulous ductal aneurysm and discusses possible aetiological mechanisms.

Case report

The patient, a 48-year-old West Indian woman, was well until eight months before admission when she consulted a doctor because of the recent onset of dyspnoea. No heart murmurs were noted, but a chest radiograph (Fig. 1a) showed cardiomegaly, with prominence and calcification in the region of the aortic knob. No treatment was given and there was no progression of her symptoms. Four months before admission, the patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy without complications. Again, several physicians noted no heart murmur. Two weeks before admission she noted progressive exertional dyspnoea. There were no other new symptoms. She had been hypertensive for five years before admission, with blood pressure recordings as high as 170/120 mmHg. Treatment of her hypertension

had been initiated four months before admission.

On admission the patient was afebrile, heart rate was 102 beats/minute, and blood pressure was 140/80 mmHg in both upper extremities. Cardiac examination revealed a left ventricular apical impulse in the sixth left interspace, between the mid-clavicular and anterior axillary lines. A palpable lift was present in the pulmonary area. A grade 3/6 continuous murmur was best heard at the left upper sternal border radiating towards the left shoulder. The second heart sound split physiologically with respiration; the pulmonary component was of normal intensity. The remainder of the physical examination was unremarkable. She had a normal white blood cell count and a haematocrit of 36 per cent. A serological test for syphilis (Hinton) was non-reactive. Sinus tachycardia and borderline evidence of left ventricular hypertrophy were present on the electrocardiogram. The chest x-ray (Fig. 1b), when compared with the film taken eight months previously, showed an increased cardiac size, a pronounced increase in the vascularity of the right lung, with a concomitant diminution in the vascularity of the left lung, and a mass in the region of the aortic knob and left hilum. The trachea was deviated to the right. On tomography, several linear calcifications were noted in the mass.

The pulmonary phase of a radionuclide angiogram, with injection into the right external jugular vein, showed only right lung perfusion; the

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Fig. 1 (a) Chest radiograph eight months before admission showing cardiomegaly, with faint calcifications in the aortic knob and left hilar regions (arrows). There is left hilar prominence. (b) Chest radiograph at time of admission showing further cardiomegaly, a pronounced increase in the right lung vascularity, and a concomitant diminution in the vascularity of the left lung compared with the film eight months before. There is now a left perihilar mass (arrows). The trachea is deviated to the right.

study also suggested a left-to-right shunt. The echocardiogram disclosed a hyperdynamic, volume overloaded left ventricle, with an ejection fraction of 0.81. These findings were felt to be compatible with a moderate left-to-right shunt at the ventricular or pulmonary artery level.

The patient's condition remained stable, and on the third hospital day, cardiac catheterisation was performed. The haemodynamic data revealed mild pulmonary hypertension (right pulmonary artery pressure 50/30, mean 41 mmHg) and mild increase in the filling pressures of both ventricles. Both pulmonary and systemic vascular resistances were moderately increased. There was a pronounced step-up in oxygen saturation in the pulmonary artery. A comparison of pulmonary and systemic blood flows showed left-to-right shunt of 5.1 l/min, with a pulmonary to systemic flow ratio of 3.4:1. A pulmonary arteriogram, with contrast injected into the main pulmonary artery, showed the left pulmonary artery to be totally occluded at its origin; the right pulmonary artery and its branches were normally opacified. Calcifications and a large mass were again observed adjacent to the aortic isthmus. The laevophase of the pulmonary arteriogram showed mild irregularity of the proximal

descending aorta and a communication between the great vessels. Contrast material did not fill the mass in either phase of the study. Injection of radiographic contrast directly into the ascending aorta filled the right pulmonary artery through a ductal communication. The size and position of this communication were demonstrated on a left anterior oblique aortogram (Fig. 2a).

Two days after cardiac catheterisation, haemoptysis developed necessitating emergency surgery. At operation a calcified ductal aneurysm connected the dilated calcified aortic isthmus with the main pulmonary artery. The proximal portion of the left pulmonary artery, together with the ductus and aortic isthmus, formed an aneurysmal sac, 5 cm in diameter and 10 cm in length (Fig. 2b). The left pulmonary artery distal to the aneurysm was obliterated with laminated clot. The mass was adherent to the medial surface of the upper lobe of the left lung, with blood tracking into the parenchyma of the left lung and along the great vessels within the mediastinum. The aneurysm was exposed by resection of the left lung. Control and closure of the aortic origin of the aneurysm was rendered extremely difficult by dilatation, friability, and calcification of the aortic arch and isthmus.

Fig. 2 (a) Left anterior oblique view of the ascending aortic injection showing a large ductus arteriosus (arrows) communicating with the pulmonary artery. (b) Sketch made at time of operation showing the ductus aneurysm. The aneurysmal sac, measuring 5 cm in diameter by 10 cm in length, completely obstructed the distal left pulmonary artery. Ao, aorta; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery.

Postoperatively, the patient was comatose and hypotensive, and died on the third postoperative day. Permission for necropsy was refused. The left lung specimens taken at operation showed mild medial hypertrophy of the muscular pulmonary arteries and variable degrees of arteriolar intimal proliferation. No embolic material was found in the peripheral pulmonary arterial branches. The pulmonary parenchyma showed diffuse haemorrhage and infarction. The ductal aneurysm consisted of a thin fibrotic and calcified arterial wall, with adherent mural thrombus and well-organised clot.

Discussion

INCIDENCE

Since this lesion was first described in 1893, only 11 other reports of adult cases have been published in the medical journals. Our patient is the first known example of a spontaneous fistulous aneurysm of a previously non-patent ductus arteriosus in which the diagnosis was made during life. In this patient, the lesion was defined by echocardiographic, radionuclide angiographic, haemodynamic, and radiographic contrast studies.

AETIOLOGY

The cause of a spontaneous ductal aneurysm in the

adult population is uncertain. Persistence of an infantile ductal aneurysm into adulthood is a possible but unlikely cause.^{1 2} More probably, it is a predisposing abnormality of ductal structure or of the mechanism of ductal closure. In the human fetus, the ductus arteriosus resembles a muscular artery extending between the aorta and the pulmonary artery, both of which have large amounts of elastic tissue in their walls. The media of the normal ductus contains no elastic lamellae comparable to those of the adjoining great arteries. Gittenberger-de Groot³ postulated that the persistence of the ductus arteriosus beyond the age of 3 months results from a primary anatomical defect of the ductal wall, characterised by aberrant distribution of elastic material in the ductal wall, with the presence of subendothelial elastic lamina as its most striking abnormality. This abnormal wall structure was found in varying degrees in patients in which only one end of the ductus arteriosus had anatomically closed. Normally, postnatal occlusion of the persistent ductus arteriosus occurs in stages. Initially, the pulmonary arterial end closes by smooth muscle contraction and intimal proliferation, briefly leaving the blind ductus infundibulum exposed to systemic pressure from the aorta.³ If closure is arrested at this stage, the ductal wall would be continually exposed to systemic arterial

pressure. It is postulated that the combination of systemic hypertension and a ductus diverticulum with an abnormal wall can result in dilatation or aneurysm formation in the ductal remnant. Subsequently, trauma, infection, arteritis, or spontaneous dissection could re-establish a channel from the aorta to the pulmonary artery.²

In our patient, systemic hypertension may have initiated an aneurysm of the ductus diverticulum by increasing the pressure in the patent portion of the ductus long after closure of the pulmonary arterial ostium. The absence of a continuous murmur or shunt vascularity eight months before admission, together with the progressive symptomatic enlargement of the aneurysm suggests the sudden "reopening" of the ductus as a result of spontaneous intramural dissection. Subsequently, compression and thrombotic occlusion of the left pulmonary artery resulted in diversion of all pulmonary blood flow to the right lung.

CLINICAL FEATURES

The presence of an enlarging calcified mediastinal mass adjacent to the left side of the trachea associated with features of tracheal or pulmonary compression (that is dyspnoea, wheezing, haemoptysis, or hoarseness resulting from laryngeal nerve impingement) or chest pain (caused by haemorrhage into or pressure on intrathoracic structures) compose a symptom complex frequently noted in patients with non-patent ductal aneurysms.²⁻⁴ When such a constellation of findings is present, a mediastinal tumour and aortic aneurysm are the primary differential diagnostic possibilities.⁵ Auscul-

tatory evidence of a continuous murmur should then complete the diagnostic constellation of findings pointing to a fistulous aneurysm of the ductus arteriosus. Complications of ductal aneurysms include infection, embolisation, or rupture, the last being the most common.¹

This case shows that the use of non-invasive and angiocardigraphic techniques in adults differentiates, before operation, a fistulous aneurysm of a persistent ductus arteriosus from other enlarging mediastinal masses. Because of the high incidence of fatal complications with this lesion, early definitive diagnosis and surgical repair are imperative.

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Requests for reprints to Dr Kenneth M Borow, Cardiovascular Division, Peter Bent Brigham Hospital, 75 Francis Street, Boston, Massachusetts 02115, USA.