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Case report

Ruptured abdominal aortic aneurysm in a stillborn fetus

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Summary. The structure of a congenital abdominal aortic aneurysm in a stillborn fetus is described. Its cause is attributed to an abnormality of development of the aortic elastic tissues.

Key words: Aneurysm – Abdominal aorta – Arterial dysplasia – Fetus

Congenital aneurysms of the abdominal aorta are rare. This case report describes a stillborn fetus found at necropsy to have a large abdominal aortic aneurysm.

Case history

The mother was a thirty-five-year-old housewife undergoing her fourth pregnancy by two marriages. Two daughters are alive and well. The third pregnancy ended in miscarriage at nine weeks' gestation. The latest pregnancy proceeded normally until 28 weeks when it was thought that there was polyhydramnios but, by one month later, an ultrasound examination showed no evidence of this nor of fetal abnormality. Various haematological and biochemical investigations were made during the pregnancy and these showed no significant deviation from the normal ranges. There was no serological evidence of syphilis and she was immune to rubella. Fetal death occured at 34 weeks' gestation.

Necropsy findings

The body was that of a macerated male fetus of a size consistent with his reported age (Weight 2,666 g; crown-heel length 44 cm; crown-rump length 32 cm; head circumference 30 cm). There was no external evidence of congenital abnormality but bruising in both flanks extended to the umbilical stalk.

Dissection revealed retroperitoneal haemorrhage and an elongated, smooth, nodular mass about 6.5 cm long and 3 cm in diameter lying in the midline (Fig. 1). This was an aneurysm of the abdominal aorta. It extended from above the renal arteries, which appeared normal and emerged from its sides, to the aortic bifurcation, where the dilated, tortuous common iliac arteries arose. On cutting, the aneurysm was seen to arise abruptly from the apparently

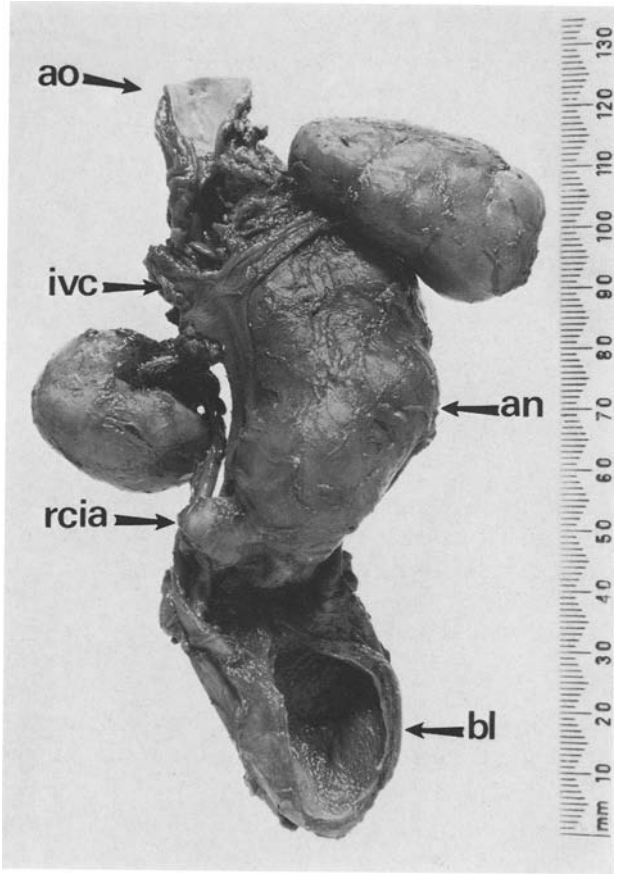

Fig. 1. The aneurysm and adjacent structures viewed anteriorly. The bladder has been opened and rotated inferiorly. (Key:– *ao:* proximal abdominal aorta; *ivc:* inferior vena cava; *an:* aneurysm; *rcia:* right common iliac artery; *bl:* bladder)

normal upper abdominal aorta (Fig. 2). Its internal diameter was 2.5 cm proximally, tapering to slightly less than 1 cm distally, and it was divided by incomplete transverse septa into several large, approximately spherical, chambers filled with laminated thrombus (Fig. 3). The internal walls of the aneurysm and iliac arteries were irregularly corrugated.

The heart (30 g) was considerably heavier than is usual for a fetus of this age (Mean 15 g; S.D. 5 g; Potter and Craig 1976), showing considerable left ventricular hypertrophy but no other abnormality. There was no stenosis of the left ventricular outflow tract nor of the thoracic aorta. The left kidney was rotated against the side of the aneurysm in such a way that the broad, shallow hilum was directed anteromedially. The left ureter was dilated above the point it crossed the left common iliac artery. Dissection of all other organs and of the placenta and umbilical cord showed no significant anatomical abnormality.

Histological findings

Minor intimal and medial changes were widespread and seen in both elastic and muscular arteries throughout the body. Areas of localised intimal thickening by elastic fibres and longitudinally-orientated smooth muscle cells,

Fig. 2. The upper end of the aneurysmal sac showing its abrupt origin from the proximal abdominal aorta and its corrugated internal surface

often associated with fraying, reduplication and fibrosis of the internal elastic lamella or its complete disappearance were seen frequently. Medial longitudinal bundles were not seen in association with these structures, only a minority of which appeared to be related to arterial branch points. Small foci of fragmentation, straightening and fibrosis of the medial elastic lamellae were seen in many elastic arteries. In a few cases the damaged elastic fibres were densely haematoxyphil and reacted with Perls' but not with the von Kóssa stain (Fig. 4). However, electron probe studies revealed the presence of calcium in amounts higher than is physiological (approximately 65–75 mmol/kg of specimen) (T.A. Hall, personal communication).

The adventitia of the lower thoracic aorta in places consisted of a loose, cellular, fibrous connective tissue and numerous blood-filled spaces. This resembled an angiomatous malformation.

The upper abdominal aorta showed both intimal and medial changes of the types described but was otherwise relatively normal in structure. More distally, the medial lamellae became disorganised in arrangement,

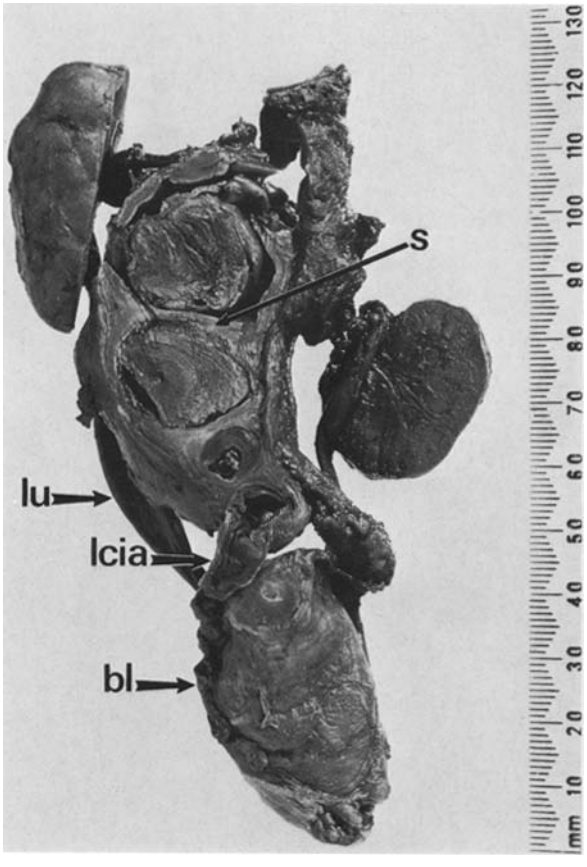

Fig. 3. The opened aneurysmal sac viewed from behind. It is divided into several chambers, filled with laminated thrombus, by transverse septa. (Key:– *s:* septum; *lu:* left ureter; *lcia:* left common iliac artery; *bl:* bladder)

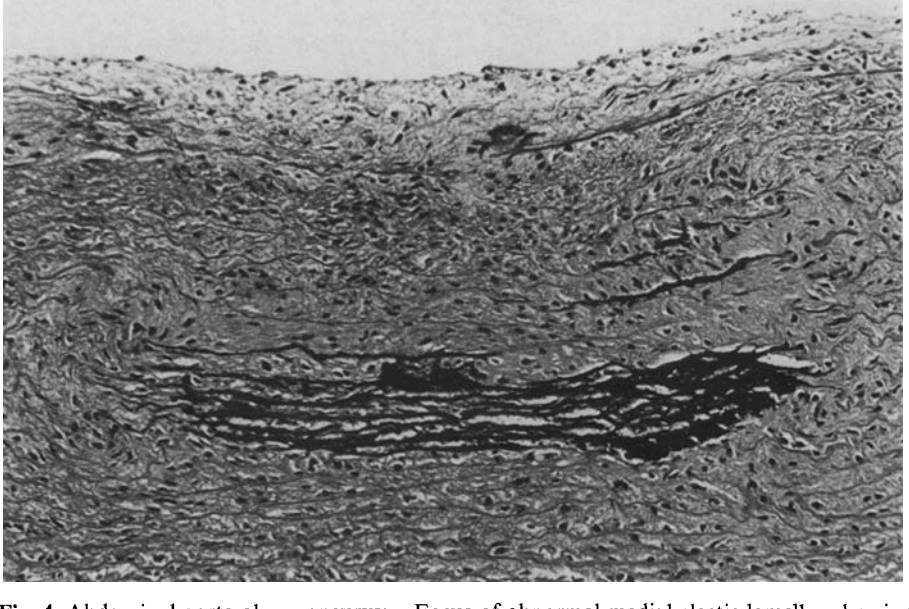

Fig. 4. Abdominal aorta above aneurysm. Focus of abnormal medial elastic lamellae showing deposition of haematoxyphil material. H&E × 160

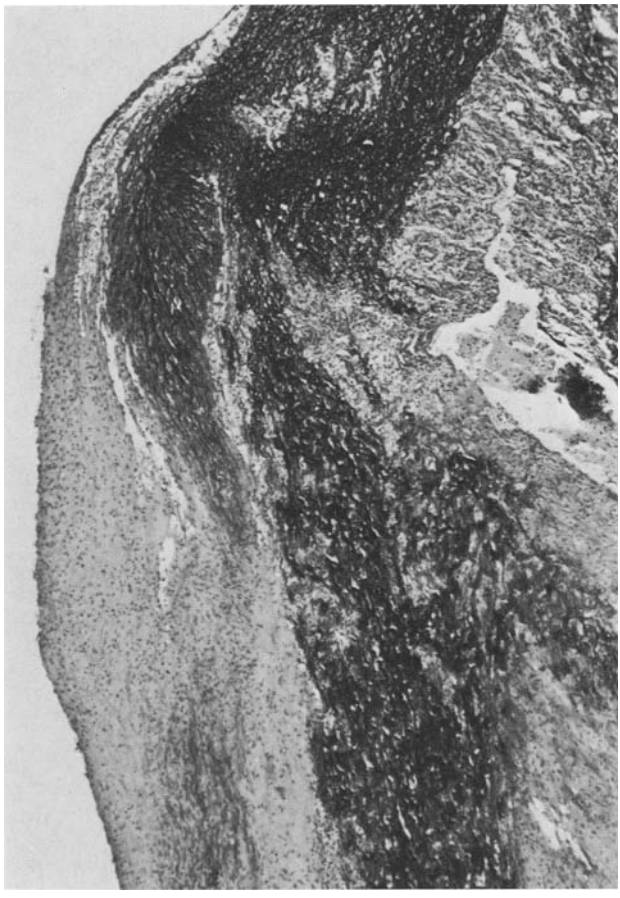

Fig. 5. The proximal end of the aneurysmal sac. The medial elastic fibres become attenuated and rotate outwards and the intima is replaced by a thick myxomatous tissue. Elastic Ponceau S ×64

tending to adopt a radial orientation, and the intima showed an increasingly prominent elastic thickening. As the media twisted outwards, it suddenly became replaced by the aneurysmal sac (Fig. 5) whose structure was most abnormal.

The wall of the aneurysm appeared to be composed of three layers, not always entirely distinct. The innermost layer consisted of a thick, irregular, undulating tissue rich in acid mucopolysaccharide and containing a scanty population of spindle-shaped or branching cells with oval nuclei. There were many fibres of thick, wavy, pale-staining collagen present. A few capillary loops were also noted. The thick middle layer was composed predominantly of collagen bundles lying parallel to the vessel wall (Fig. 6) and a few delicate fibrils of elastic tissue arranged haphazardly. Its cellularity varied considerably; for the most part there were only a few fibroblasts but a few areas showed dense accumulation of plump, spindle-shaped cells with oval nuclei (Fig. 6). In a few places small foci of coarser elastic fibres arranged in parallel bundles resembled the usual structure of elastic arteries.

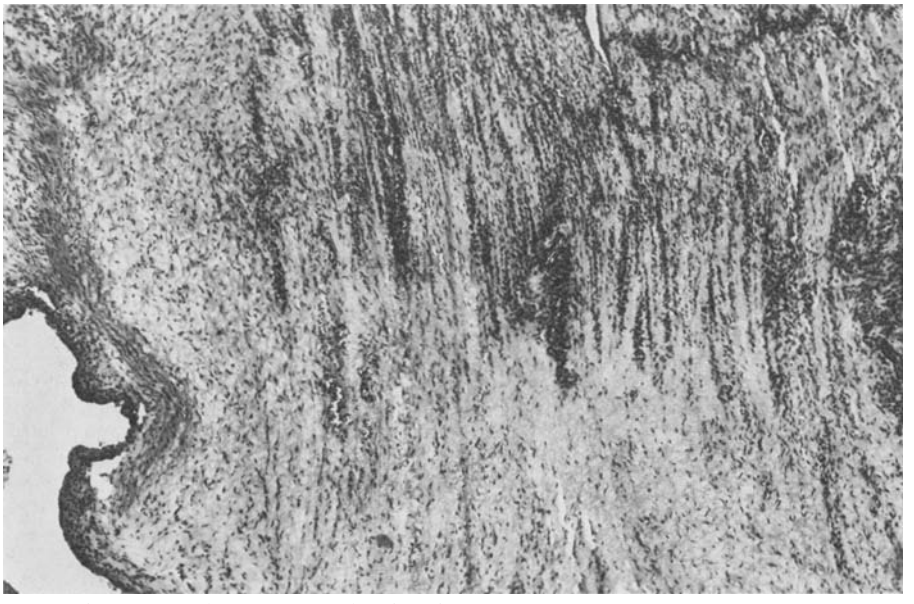

Fig. 6. The wall of the aneurysm showing dense accumulations of spindle-shaped cells and fibrous tissue. Haematoxylin and eosin × 64

The innermost part of the middle layer was rich in acid mucopolysaccharides. The adventitia of the aneurysm consisted of a dense fibro-elastic tissue and proximally, in the angle between the abdominal aorta and the upper end of the aneurysmal sac, there were collections of spindle-shaped cells similar to those of the media. A few adventitial arteries showed cellular intimal thickening.

The septa dividing the aneurysm consisted of an inner core of tissue like that of the middle layer of the wall coated on either side by the loose, myxomatous tissue. Each layer merged smoothly with its counterpart in the wall of the sac. The lumen contained laminated thrombus, calcified in places, but showing no evidence of organisation.

The site of rupture of the aneurysm could not be detected and there was no evidence of arterial dissection. Proximal branches of the aorta, arising from the sac, and the common iliac arteries were similar in structure to the aneurysm.

The left kidney contained a small subcapsular focus of several relatively large arteries situated in young connective tissue. Scattered throughout the interstitium were iron-laden macrophages. The placenta showed non-specific changes consistent with intra-uterine death. Histological examination of other organs showed no changes other than autolysis.

Discussion

The abnormalities recognised in this fetus were a thrombosed, ruptured, abdominal aortic aneurysm with severely dysplastic walls, focal medial arte-

rial lesions and severe left ventricular hypertrophy. There was also a small angiomatous malformation in the adventitia of the thoracic aorta and a tiny, hamartomatous vascular abnormality of the left kidney.

The left ventricular hypertrophy may be accounted for by the increased resistance to flow in the aneurysm and also, possibly, by a reno-vascular hypertension secondary to compromise of the proximal parts of the renal arteries by the aneurysmal sac.

The widespread, patchy intimal changes resembled those described in the coronary arteries of fetuses and infants (Fangman and Hellwig 1947; Levene 1956; Schornagel 1956; Velican and Velican 1976 and 1977) and in fetal peripheral arteries (Robertson 1960). Fangman and Hellwig (1947) and Levene (1956) suggested that such foci are pathological in nature but the other authors believed them to be a physiological response to haemodynamic stresses.

Medial arterial lesions, consisting of degeneration and fibrous replacement of the elastic lamellae, were often associated with the deposition of iron salts and calcification. They appear to be pathological in nature and presumably represent healed foci of arteritis. Whether they bear any relationship to the aneurysm is not clear.

Spontaneous abdominal aortic aneurysms affecting fetuses or infants are of great rarity. Howorth (1967) described a large congenital aneurysm in a 4-day-old girl. It showed scanty histological abnormalities, consisting of moderate variation in thickness of the constituent layers, especially of the intima. Hagood and associates (1976) reported a 22-month-old girl with tuberous sclerosis and an abdominal aortic aneurysm, which they believed to be a consequence of severe hypertension and obstruction of the iliac arteries, which showed fibromuscular dysplasia. Other aneurysms have been described but the accounts are rather limited (Gibson 1946; Phenomenov, cited by Ferrard et al. 1964). Spontaneous congenital aneurysms have also been noted to affect the hepatic, basilar and axillary arteries (Gryboski and Clemett 1967; Doerr 1970; Dormandy and Barkley 1979.

The aneurysm described in this paper showed a bizzare, dysplastic wall affecting predominantly the intimal and medial layers. Similar changes extended into the common iliac arteries and into some of the larger, proximal branches of the abdominal aorta. There was no evidence of infection or of inflammation of the arterial wall and the maternal history was unhelpful. The mother was healthy during the pregnancy and she was not prescribed any medications during its course. Her brother had essential hypertension but otherwise the family is in good health. There is no evidence of inherited disorders of connective tissues. The cause of the aneurysm remains obscure. It is likely to have been due to some abnormality of development of the abdominal aortic elastic tissue. It is interesting to note that the thoracic aorta was unaffected. Some of the aneurysmal changes in this case are similar to those seen in fibromuscular arterial dysplasia. Such alterations may represent a general response of the arterial wall to certain stimuli (Pesonen et al. 1980).

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