Case reports

Diffuse non-specific aortitis with multiple saccular aneurysms and aorto-enteric fistula

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SUMMARY A rare variant of non-specific aortitis was found at necropsy in a young hypertensive woman. The aorta showed severe, extensive, non-specific aortitis with multiple saccular aneurysms containing thrombi throughout its length except for 2 cm from its bifurcation. A fistulous tract was identified leading from an aortic aneurysm at the level of the renal arteries to the second part of the duodenum. Both renal artery ostia were narrowed. The small intestine contained 2 litres of blood, at necropsy. Bilateral ischaemic atrophy of the kidneys was present.

Non-specific aortitis is not ubiquitous in distribution. This disease was first described as an entity from Japan.1 Non-specific aortitis with multiple saccular aneurysms has been reported only from South Africa.2-4 None of the cases in these series had developed spontaneous aortic fistula. We report a case of severe, extensive non-specific aortitis with multiple saccular aneurysms and spontaneous aortoduodenal fistula.

Case report

CLINICAL HISTORY
A 22 year old woman was found to be hypertensive during a routine check up. Three years later she noticed a lump in the abdomen and since then had suffered from severe intermittent back pain.

On examination all peripheral pulses were felt, there was no brachiofemoral delay, and blood pressure ranged between 170-200/130-140 mm Hg. Prominent suprasternal pulsations, left ventricular heave, loud and split second heart sound with prominent aortic component, and a fourth heart sound were noted. A pulsatile mass measuring 5 cm x 5 cm was present in the epigastrium. Grade two retinopathy was also present.

Her haemoglobin concentration was 11 g/dl, erythrocyte sedimentation rate 54 mm in the first hour, and blood urea concentration 6-7-8-3 mmol/l (40-50 mg/100 ml). Chest radiographs showed a normal size heart, widened aortic arch, and prominent descending thoracic aorta. An electrocardiogram showed sinus rhythm with left ventricular hypertrophy. On the day scheduled for aortography she developed profuse haematemesis and died from circulatory shock.

NECROPSY FINDINGS
Only a 15 cm midline abdominal incision was permitted. The heart weighed 420 g and showed left ventricular hypertension. The aorta and its major branches were normal. The left ventricle was small, the right ventricle was large, and there was atrial septal defect with a systolic thrill. The left atrium was dilated. The left renal artery was stenosed. The right renal artery was normal. The aorta was dilated and there was a large saccular aneurysm in the abdominal aorta. There was a fistula between the aneurysm and the duodenum. The mesenteric vessels were normal. The liver was moderately enlarged with fatty changes. The pancreas was normal. The stomach was normal. The jejunum and ileum were normal. The cecum and appendix were normal. The small intestine contained 2 litres of blood, at necropsy. Bilateral ischaemic atrophy of the kidneys was present.

Fig. 1 Diagram showing the aorta, multiple aneurysms, aortoduodenal fistula (bidirectional arrow), and atrophied right kidney with severely stenotic left renal artery. Thrombi are seen within the aneurysms. Atherosclerotic patches are also present.
Diffuse non-specific aortitis with multiple saccular aneurysms and aorto-enteric fistula

Fig. 2. Morphological appearance of the abdominal aorta, an aneurysm at the level of renal vessels with thrombus, severely atrophied right kidney, and granular left kidney. The liver, spleen, pancreas, and urinary bladder are also shown.

tricular hypertrophy and dilatation. The proximal 2 cm of the left main coronary artery showed fatty streaks.

The aorta was dilated and irregularly tortuous in its entire length and showed multiple saccular aneurysms varying from $1 \times 0.5 \times 0.3$ cm to $7 \times 6 \times 4.5$ cm in size (Fig. 1). The thickness of the wall of the aorta varied from 0.1 cm to 0.3 cm. The intimal surface was pink and was of the consistency of puff pastry throughout, except for the terminal 2 cm near its bifurcation. In addition, there were interspersed atherosclerotic plaques and ulcers in the intima. Most of the aneurysms were filled with fresh as well as partially organised thrombi. The largest one, situated at the level of the renal vessels (measuring $7 \times 6 \times 4.5$ cm) was partially filled with an organised thrombus. A probe could be passed from its floor through a tunnel (0.9 cm in diameter) to the second part of the duodenum situated 2-6 cm distal to the opening of the bile duct. The right renal artery ostium was narrowed, thickened, and roughened and that of the left artery occluded by an organised thrombus. The main branches of the aorta, except the renal arteries, were free of disease.

Multiple sections from the diseased atherosclerotic and relatively normal looking aorta and also from the wall of the aneurysms were stained with haematoxylin and eosin, Masson’s trichrome stain, and phosphotungstic acid haematoxylin and examined. The aorta showed features of non-specific aortitis with pronounced thickening of the intima due to increased fibrocollagenous tissue, smooth muscle proliferation and deposition of basophilic intercellular substance. The media showed variable destruction of muscle and elastic tissue with replacement fibrosis. The outer two thirds of the media showed prominent neovascularisation.

The pelvicalyceal system and ureters were normal.

Discussion

Non-specific aortitis with multiple saccular aneurysms has been reported from Africa.2-4 This type is uncommon in India, and to the best of our knowledge such a case has not been documented at necropsy. A few cases of non-specific aortitis with multiple aneurysm formation, however, have been reported in a contrast study (S Bhargava, personal communication). The South African variety of aortitis is diffuse and the aneurysms are characteristically multiple and saccular compared with the other varieties of aortic aneurysms reported in various series,5-6 in which they are fusiform and not multiple.

An aortoenteric fistula was found in the present case. This is unusual in non-specific aortitis. On the other hand fistulous tracts are not uncommon in the fusiform abdominal aneurysms described in various series, and about 31% cases rupture into the duodenum.7-10 Although multiple saccular aneurysms have been reported in cases of aortitis seen in South Africa, in none of these cases has an aortoenteric fistula been reported.

The aetiology of the multiple aneurysms in this case was determined by exclusion. Syphilitic origin was unlikely since aneurysms in this disease are most commonly found in the ascending aorta, although the abdominal aorta is the affected site in about 10% of cases. Multiple aneurysms are unusual in syphilis. Multiple aneurysms may also result from severe and extensive atherosclerosis, which is an unlikely cause in this case for several reasons. Firstly, this patient was a young woman. Secondly, at necropsy the process of atherosclerosis was only patchy, and thirdly, in the non-atherosclerotic areas of the aorta also there was pronounced thickening of all the layers—namely, the intima, media, and adventitia. It thus appears from the histological features that in this case the mul-
Multiple aneurysms resulted from a severe, diffuse, nonspecific aortitis.

References