Sarcoid aortic aneurysm: surgical difficulties

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Sarcoidosis of the abdominal aorta is extremely rare. This is the first report of reconstruction for sarcoid aneurysm of the abdominal aorta. Because the aortic wall is quite friable in this condition, suture ligation of the lumbar arteries and construction of the proximal and distal anastomoses are considerably more difficult than in repair of an atherosclerotic aneurysm.

Sarcoidosis presenting as an abdominal aortic aneurysm has not been previously reported. One case of sarcoidosis involving the abdominal aorta has been described as an incidental post mortem finding in a patient with widespread sarcoidosis involving the lymph nodes, heart, lung, liver, testes and meninges[1].

Because our case is the first aortic reconstruction for sarcoid aneurysm of the abdominal aorta to be reported, we feel it necessary to document the operative surgery and pathology of the condition because they differ significantly from the surgery for an atherosclerotic abdominal aortic aneurysm.

Case report. A 56-year-old male negro presented to hospital because of vague abdominal discomfort of 8 weeks duration. He had no other symptoms and no previous significant illness. On examination, he looked well, and the only significant clinical finding was a 6 cm abdominal aortic aneurysm that seemed to be confined to the infrarenal aorta and not to involve the iliac arteries. ECG, chest X-ray and routine blood investigations were normal. Aortography was not performed.

Operative findings. At laparotomy, the liver, spleen, kidneys and gut were grossly normal. Although there were numerous hard, markedly enlarged para-aortic nodes, dissection of the aneurysm was easy as these nodes were not adherent to the aortic wall. The aneurysm commenced 6 cm below the renal arteries, increased in size gradually and was largest 1 cm proximal to the aortic bifurcation. There was no clear "neck" to the aneurysm. On opening the aorta, the lumen was approximately twice normal size, while the thick, fleshy wall had a smooth, glistening inner lining. No atherosclerotic plaques were seen, but a thin layer of firmly adherent thrombus was present in some areas. The greyish, fleshy aneurysm wall could not hold sutures securely enough to occlude the lumbar arteries, all of which were patent. Most of the aortic wall had to be dissected from within to get at the point where the lumbar arteries leave the aorta in order to securely transfix and ligate them. The inferior mesenteric artery was similarly ligated. Because of the gradual taper and absence of a neck proximally, and also because the affected area was too friable to hold sutures, the aorta was completely transected well above the aneurysm so that the proximal anastomosis could be constructed using the full thickness of grossly normal aorta. Distally, the aneurysm extended to the bifurcation with normal iliac arteries, and it appeared that a 16 mm tube dacron graft would fit easily in that position.

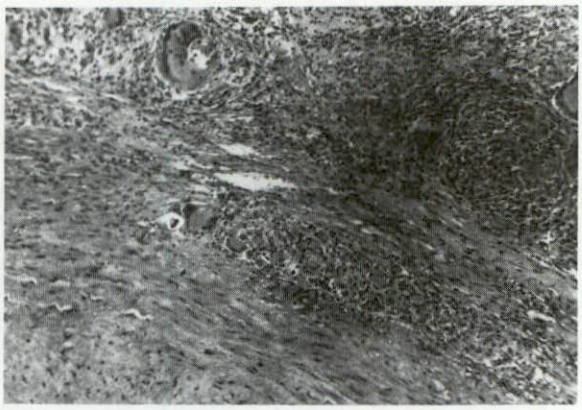

Fig 1 Section of aortic wall showing sarcoid granulomas in the adventitia and media. (H&E × 60)

However, in constructing the distal anastomosis, the posterior wall at the bifurcation would not hold sutures securely, and a 16 × 8 mm short segment of bifurcation graft was used for anastomosis into the common iliacs 1 cm from the aorta. The sac was closed over the graft while 2 nodes and a piece of aortic wall were sent for pathology. The patient recovered well, and repeat chest X-ray 6 weeks postoperatively showed no evidence of pulmonary or hilar node involvement.

6 months later, however, he presented with a 1 cm cutaneous lesion on the forehead, which on biopsy showed classical sarcoid granulomas. Chest X-ray at this time was still normal.

Pathology. The aortic wall showed numerous non-caseating epitheliod granulomas with giant cells in the adventitia which, at some areas, infiltrated the media. The intima showed hyaline degeneration but no evidence of atherosclerotic changes (Fig 1). The 2 nodes showed extensive hyalinisation and clusters of non-caseating epithelioid granuloma. Special stains for mycobacterium tuberculosis and fungus were negative. The histological features were classical of sarcoidosis in both the aorta and lymph nodes.

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Discussion. Sarcoidosis of the aorta is extremely rare, and other conditions with which it may be confused include other granulomatous diseases and giant cell aortitis. In our case, classical sarcoid granulomas involving both the aorta and para-aortic nodes were seen. In giant cell aortitis the nodes do not show granulomas. Further, the histologic confirmation of a cutaneous sarcoid lesion, 6 months later, establishes this as a case of sarcoid aortitis.

Although sarcoidosis involving small vessels of the lung[2], eye[3], brain[4], and mesentery[5] have been described, "sarcoid aortitis" has only been noted in 4 cases, 3 of which involved the thoracic aorta[1,6,8]. The description of an infrarenal sarcoid abdominal aortic aneurysm by Maeda *et al*[1] differs in several respects from our case.

Their patient had extensive atherosclerosis with a thickened calcified aorta, thrombus in the lumen, and aneurysmal dilatation of the entire infrarenal aorta – gross findings very similar to an atherosclerotic aneurysm, though histologically, sarcoid granulomas were found in the media and intima. In our case, the 6 cm of uninvolved infrarenal aorta was quite normal in size and consistency, with a normal intimal surface and no atherosclerotic plaques. The aneurysm wall itself, though thick, was quite soft and fleshy, with no calcification and very little thrombus in the lumen. If, during surgery for an aortic aneurysm, these features are found, along with enlarged para-aortic nodes, the diagnosis of sarcoidosis of the aorta should be considered.

The operative surgery for this condition may differ from that of an atherosclerotic aneurysm in the following respects:

(1) Because the fleshy wall does not hold sutures well, it may be necessary to dissect the lumbar orifices from within the aneurysm to transfix them securely.
(2) There is no neck proximally and it is better to transect the aortic wall completely, well above the grossly diseased area, to ensure that sutures will hold securely on the aortic wall.
(3) Although the aneurysm stops at the bifurcation, the posterior wall may be too friable to hold sutures. A very short bifurcation graft may be safer than a tube graft, even if the common iliac arteries appear grossly healthy.
(4) Since there is very little thrombus in the lumen and the intima appears grossly normal, there is little risk of distal embolism during dissection.

The natural history of this condition is not known, and although our patient has done well for 9 months, we do not know if the disease process will eventually extend proximally or distally to involve the suture lines.

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Heterotopic pancreas: a rare cause of obstructive jaundice

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A case of obstructive jaundice due to pancreatic heterotopia is presented. The pre-operative and per-operative diagnosis was either cholangiocarcinoma or carcinoma of the head of the pancreas, for which a cholecystojejunostomy was performed, followed by a pancreaticoduodenectomy at a later date. This is the third reported case to survive surgical treatment and it differs markedly from the previous two.

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Heterotopic pancreas (also known as ectopic pancreas or aberrant pancreas) is a condition in which pancreatic tissue is abnormally situated, the most common sites being stomach, duodenum and jejunum. The condition is most frequently found at post mortem or laparotomy as an incidental finding[1,2]; its clinical significance has not been established[3].

Obstructive jaundice due to heterotopic pancreatic tissue around the ampulla of Vater is rare, and to our knowledge only 5 cases have been recorded[4].

Case history. A 58-year-old man presented with an 8-week history of painless jaundice, generalised itching, pale stools and dark urine, and weight loss of 9.5 kg. On examination, he