

Multisaccular Aneurysm Developing in Association with Abdominal Aortic Coarctation: Report of a Surgical Case

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This paper reports a rare case of a 65-year-old woman diagnosed with a multisaccular, abdominal aortic aneurysm (AAA), 35 mm in diameter, which was revealed developing just distal to an abdominal aortic coarctation (AAC), with a 20 mmHg pressure gradient. The patient underwent corrective surgery for both lesions, with success. Intraoperatively, the aneurysm wall was found to be so thin and transparent that the inner blood turbulence could be seen, and it appeared highly susceptible to rupture. When a saccular, thin-walled AAA develops in association with AAC, early surgical intervention is mandatory regardless of the size of the aneurysm. (Ann Thorac Cardiovasc Surg 2003; 9: 326–9)

Key words: abdominal aorta, aneurysm, coarctation

Introduction

It is extremely rare for an abdominal aortic aneurysm (AAA) to develop in association with abdominal aortic coarctation (AAC) although a thoracic aortic aneurysm (TAA) sometimes does with a thoracic aortic coarctation (TAC)^{1,2)} since AAC by itself is a relatively rare vascular lesion accounting for 0.5-2.0% of all coarctations of the thoracic and abdominal aortae.³⁻⁵⁾ Recently, however, we successfully operated on a patient with a multisaccular AAA developing just distal to AAC.

Case Report

A 65-year-old woman, 152.0 cm tall and weighing 43.6 kg, was referred to us for an AAA which had been incidentally found five years previously. The patient had received regular medical follow-up with standard antihypertensive drugs since the aneurysm size had remained small. Upon physical examination, the systemic blood pressure

measured by the upper right arm was 140/62 mmHg, and the upper extremity blood pressure was always more than 20 mmHg higher than that in the lower extremities. There was a pulsating, hen's egg-sized mass in the periumbilical region, and there were thrills and bruits (Levine degree 4/6) along the upper abdominal aorta. Abdominal computed tomography (CT) revealed an AAA, around which no mantle sign⁶⁾ was seen. Abdominal three dimensional (3D) CT clearly demonstrated the multisaccular configuration of the AAA in conjunction with the abdominal aorta and adjacent visceral arteries (Fig. 1). There appeared to be coarctation of the infrarenal aorta. Conventional aortography revealed that the right renal artery branched off significantly distal to the left one, and the infrarenal aorta tapered and was as small as the common iliac artery, presenting coarctation (Fig. 2). Just distal to the coarctation site, a multisaccular aneurysm was clearly shown by aortography. Based on these images, the wall of the aneurysm was thought to be very thin, without mural thrombi and atheromatous plaques. The more distal abdominal aorta showed aneurysmal dilation in two other sites. On the other hand, neither stenotic nor aneurysmal lesions were observed with the visceral arteries. Therefore the conclusive diagnosis was a multisaccular AAA developing in association with AAC. The patient underwent corrective surgery for both lesions.

The operative findings and reconstructive procedure are illustrated in Fig. 3. The external diameters of the su-

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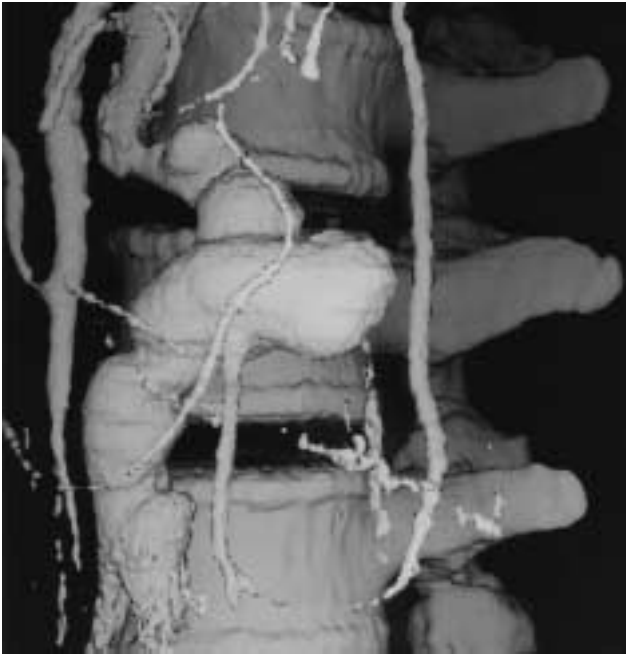


Fig. 1. Three dimensional computed tomography in the left anterior oblique view. A multisaccular aneurysm is clearly seen developing just distal to the aortic narrowing, that is, coarctation. The largest part of the aneurysm is located at the root of the inferior mesenteric artery which communicates with a thick meandering artery.

prerenal, interrenal, and infrarenal aortae measured 18 mm, 15 mm, and 10 mm, respectively. There was an unusually thick meandering artery originating from a splenic artery and terminating in the root of the inferior mesenteric artery (IMA). A multisaccular aneurysm was observed developing just distal to the coarctation site. The maximum diameter of the aneurysm was 35 mm at the IMA root, where the aneurysm wall was so thin and transparent that inner blood turbulence could be seen. Dissection of the peritoneum from the walls of AAC and AAA was quite easy since there was neither abnormal adhesion nor thickening of the peritoneum. Using the direct needle-puncture method, the pressure gradient across the AAC was determined to be 22 mmHg. After systemic heparinization and isolation of the abdominal aorta, with the most proximal cross-clamp placed between the right and left renal arteries, the aorta was transected 7 mm distal to the right renal artery. The left anterior aspect of the proximal stump of the aorta was cranially incised as high as possible (Fig. 3, inset) not only to make it easy to cannulate the right renal artery with an 8-French catheter for renal protection but also to enlarge the stenotic site. Via the catheter, the right kidney was perfused with 500 ml



Fig. 2. Aortogram in the anteroposterior projection. The infrarenal abdominal aorta tapers, presenting coarctation which is masked by the engorged superior mesenteric artery, and distal to the coarctation a multisaccular aneurysm is observed. Note the unusually long distance between the right and left renal arteries. An artery observed just distal to the right renal artery is an engorged lumbar artery as one of collaterals to the lower half of the body.

of 4°C lactated Ringer's solution using a pressure cuff set around 80 mmHg. A prosthetic vascular graft (12 mm in diameter), with a tongue-shaped sleeve, was anastomosed in an end-to-end way using a Dacron felt strip (Fig. 3, inset). The right renal circulation was restored by moving the most proximal aortic clamp on to the graft. After doubly ligating the IMA root and cross-clamping both iliac arteries, the aneurysm and distal abdominal aorta were longitudinally incised in a segmental manner, which was useful to control a significant influx of blood from lumbar arteries. There was neither mural thrombus nor atheromatous plaque in the multisaccular aneurysm. The distal aorta was transected just proximal to the iliac bifurcation, where the graft end was directly anastomosed.

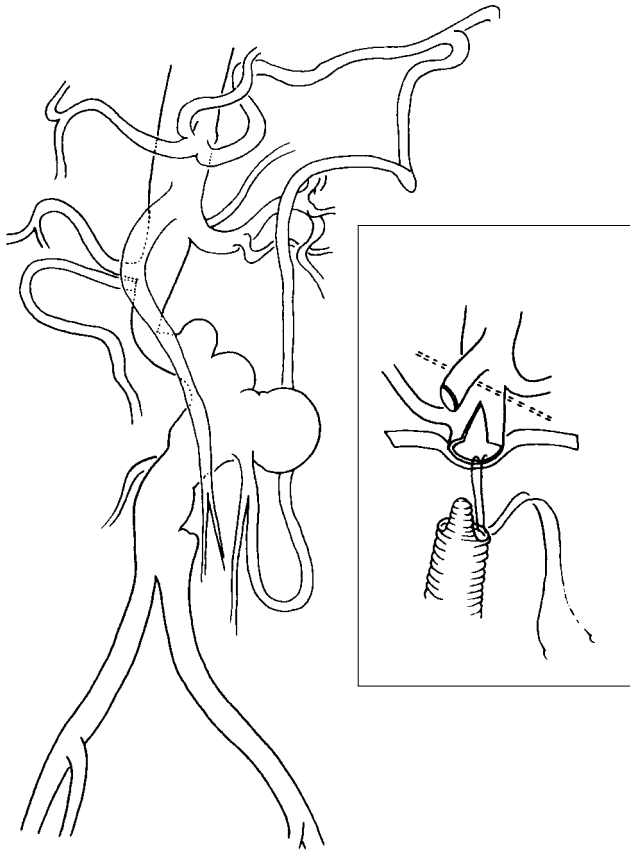


Fig. 3. Diagrams of the intraoperative vascular anatomy and the reconstructive procedure. The vascular anatomy is illustrated in accordance with that shown in Fig. 2. The inset shows the aortic reconstructive procedure, and the broken double lines indicate the site of the most proximal cross-clamp.

Postoperatively, the pressure difference between the upper and lower extremities eventually disappeared.

Macroscopic examination of the surgical specimen from the coarctation site revealed that it was not the thickening of the aortic wall but the decrease of the external diameter that caused the luminal stenosis of the aorta. Microscopically, hyaline and/or basophilic degeneration with fibrous proliferation in the aortic wall was observed, and muscle fibers were disrupted in some parts and completely disappeared in other parts; the magnitude of these changes varied from site to site. There were no histological findings which indicated the presence of some inflammatory tissue responses. It was noteworthy that some parts of the multisaccular aneurysm wall consisted of only a very thin layer of collagen fibers, and that the elastic lamina and smooth muscle fibers had completely disappeared. There were, however, no histopathological findings suggesting the etiology of coarctation and/or

multisaccular form of the aneurysm.

Discussion

AAC is a rare vascular lesion compared with TAC. According to a review of 119 cases of AAC, the mean and median ages of the patients were 21.8 ± 16.0 (1 SD) and 17 years, respectively.⁴⁾ Therefore the present patient was among the oldest. The most common location of the aortic narrowing was interrenal (52%), and the remaining cases were confined to the suprarenal, infrarenal and diffuse entire abdominal aorta in 11%, 25% and 12% of the 119 patients, respectively.⁴⁾ Thus, infrarenal narrowing as in our case is relatively uncommon.

Whether AAC is congenital or acquired is controversial, multiple renal arteries, which are considered to be a significant finding supporting the congenital origin,⁴⁾ were not observed in our patient. However, an unusually long distance between the two renal arteries in the present case was considered to be suggestive of a congenital disorder. The decreased external diameter of the aorta which caused stenosis was also attributable to a developmental disorder, favoring a congenital origin.⁵⁾ Furthermore, the patient's medical history, clinical signs, and operative and histological findings did not indicate the presence of any inflammatory process. Therefore we thought that the AAC in this patient was of a congenital origin.

Vascular disorders other than multiple renal arteries, such as stenotic renal arteries, splenic arterial occlusive lesions, and visceral artery aneurysms, have been noted in association with AAC.³⁻⁵⁾ In the present case, however, no such vascular lesions were observed except for a large meandering artery, which was considered to have been a major collateral to the lower extremities via IMA. The major vascular disorder associated in this patient was a multisaccular AAA just distal to the AAC.

The incidence of the association of aortic aneurysms with whole aortic coarctations was reported to be 17%, and among 106 cases of aortic coarctation with aneurysms, 103 were TAC and the remaining three were atypical coarctations; eventually those three were also considered as atypical TAC rather than AAC.¹⁾ Thus aortic aneurysms developing adjacent to AAC have rarely been documented. Accordingly, we discussed the aortic aneurysm in the present case by referring to those developing in association with TAC.

A great majority (83%) of previously reported such aneurysms located adjacent to the stenotic site, and the most common site (51%) was distal to the stenosis as in

the present case.¹⁾ The aneurysms were of the saccular, dissecting and fusiform type in 77.0%, 18.8% and 4.1%, respectively, of 122 aneurysms in 106 patients.¹⁾ It was also reported that the walls of the saccular aneurysms were often so thin that the inner blood turbulence could be seen^{2,7)} as in this case. Although saccular aneurysms are the most common type reported, the multisaccular type is rare and interesting.^{2,7)}

The following etiologies of aortic aneurysms associated with TAC have been proposed:^{1,7)} predisposition to aortic wall weakness, acquired aortic wall weakness after an inflammation, persistent hypertension, and jet-stream blood turbulence formed by stenosis and/or backflow from collaterals into the aorta. In our case, complex factors may have contributed to the development of the saccular aneurysm. Inflammation, however, could be excluded since there were no clinical, operative or histological findings that suggested an inflammatory origin. It was noteworthy that the largest saccular part of the AAA was observed at the root of the IMA, where the aorta must have received an extraordinary amount of blood influx from the large meandering artery. That is, in addition to the poststenotic blood turbulence, the jet stream of the backflow into the aorta from the large meandering artery via the IMA might have played one of the important roles in the development of the aneurysm in this case.

One of the causes of death in patients with aortic coarctation is rupture of the associated aneurysm, which accounted for 11% of the deaths and occurred at a mean age of 25 years in one study.⁸⁾ Survival of more than 60 years as with the present patient appears to be very rare among those having both aortic coarctations and aneurysms. In the series of 106 such patients only one

survived such a long term.⁴⁾ Therefore we think that the present case is extremely rare.

In conclusion, when a saccular, thin-walled AAA develops in association with AAC, early surgical intervention is mandatory regardless of the size of the aneurysm.

References

1. Skandalakis JE, Edwards BF, Gray SW, Davis BM, Hopkins WA. Collective review. Coarctation of aorta with aneurysm. *Intern Abst Surg* 1960; **3**: 307–26.
2. Inoue T. Aortic aneurysms associated with aortic coarctation. *Heart* 1995; **27**: 1089–92. (in Japanese)
3. DeBakey MF, Garrett E, Howell JF, Morris GC Jr. Coarctation of the abdominal aorta with renal artery stenosis: surgical consideration. *Ann Surg* 1967; **165**: 830–43.
4. Graham LM, Zelenock GB, Erlandson EE, Coran AG, Lindenauer SM, Stanley JC. Abdominal aortic coarctation and segmental hypoplasia. *Surgery* 1979; **86**: 519–29.
5. Dogan R, Demircin M, Hamaloglu E, Balkanci F, Gungen Y, Bozer AY. Coarctation of the abdominal aorta with left renal artery and splenic artery aneurysms. *J Cardiovasc Surg (Torino)* 1996; **37**: 457–61.
6. Ikeda K, Abe T, Itou M, Tamiya Y, Tanaka T, Kazui T. Successful surgical treatment of primary aorto-duodenal fistula associated with inflammatory abdominal aortic aneurysm: a case report. *Ann Thorac Cardiovasc Surg* 1999; **5**: 194–7.
7. Mizuno A, Kurosawa H, Koyanagi K, Aoki I. Successful treatment of cluster-like aneurysm associated with coarctation of the aorta. *Jpn J Cardiovasc Surg* 2001; **30**: 55–7. (in Japanese)
8. Campbell M. Natural history of coarctation of the aorta. *Br Heart J* 1970; **32**: 633–40.