Surg Today (2004) 34:354–356
DOI 10.1007/s00595-003-2694-0

SURGERY TODAY

Case Reports

Shaggy Aorta Syndrome After Acute Arterial Macroembolism: Report of a Case

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Abstract
We report the case of a patient who underwent treatment for a macroembolism in the right lower leg, which led to shaggy aorta syndrome. Anticoagulant therapy for the macroembolism and intra-aortic catheterization exacerbated the patient's renal function and triggered another massive microembolization of the visceral arteries, with a fatal outcome. To minimize the incremental complications inherent to this syndrome, awareness and prompt diagnosis with enhanced computed tomography or intravenous digital subtraction aortography are essential. Axillo-bifemoral bypass with bilateral external iliac artery ligations, performed with optimal timing, could save patients with shaggy aorta syndrome.

Key words Shaggy aorta syndrome · Arterial embolism · Intravenous digital subtraction angiography · Enhanced computed tomography

Introduction

Shaggy aorta syndrome is a rare disorder causing peripheral, renal, and visceral ischemia, caused by multiple cholesterol emboli from the aorta. The term "shaggy" was derived from the visual characteristics on angiograms and computed tomography (CT) films of the aorta, which show a spiculated and irregularly shaped surface of the aortic wall. We describe a very unusual case of a macroembolism followed by shaggy aorta syndrome, and discuss the diagnosis and treatment of this complicated disease.

Reprint requests to: N. Hayashida
Received: May 29, 2002 / Accepted: July 8, 2003

Case Report

A 59-year-old man was admitted to our hospital after the sudden onset of severe pain in his right lower leg. The initial diagnosis was acute arterial obstruction, and an emergency thrombectomy was performed. The hematologic laboratory data and blood chemical values are summarized in Table 1. General heparinization was given as 50 mg intravenously before the thrombectomy. White emboli containing red thrombi were successfully removed, following which peripheral arterial pulsation was restored, and the right lower extremity pain subsided. A continuous intravenous heparin infusion of 240 mg/day was subsequently initiated, and the dose was decreased to 100 mg/day by 24 h after thrombectomy. However, 12 h after the thrombectomy, the patient started complaining of severe abdominal pain, and there was a small amount of mucus and blood in his stool.

Aortography was immediately performed to locate a possible cause of mesenteric ischemia. Irregularities and ulcerative lesions of the abdominal aorta were seen, but there was no evidence of stenosis in the superior mesenteric artery (Fig. 1). The inferior mesenteric artery and the left internal iliac artery were not depicted. Enhanced computed tomography (CT) of the aorta also showed irregularly shaped intraluminal thrombi in the descending and abdominal aorta (Fig. 2). Shaggy aorta syndrome was strongly suspected and an exploratory laparotomy was done 1 day after the thrombectomy because of the persistent severe abdominal pain. The sigmoid colon was slightly pale, but good pulsation was confirmed in the sigmoid and marginal arteries. Therefore, colon surgery was not deemed necessary at the time. Several hours after the operation, anuria and livedo reticularis in the lower trunk and bilateral lower extremities were noted, but his hemodynamics were still stable, with a regular heart rate of 80 beats/min, and a systolic blood pressure of 110 mmHg. Continuous

Table 1. Hematologic, biochemical, and blood gas values

Variables	On admission	Before 1st lap.	Before 2nd lap.
Hematocrit (%)	45.2	45.9	39.8
White cell count (per mm^3)	17700	25900	14850
Platelet count (per mm^3)	240000	245000	145000
C-Reactive protein (mg/dl)	0.4	11.0	38.7
Creatinine kinase (U/l)	239	40630	92790
Prothrombin time (INR)	1.07	1.16	1.49
Activated partial thromboplastin time (s)	N/E	39.9	76.8
pH	N/E	7.452	7.400
Base excess (mmol/l)	N/E	−0.3	−5.8

N/E, not examined; lap, laparotomy

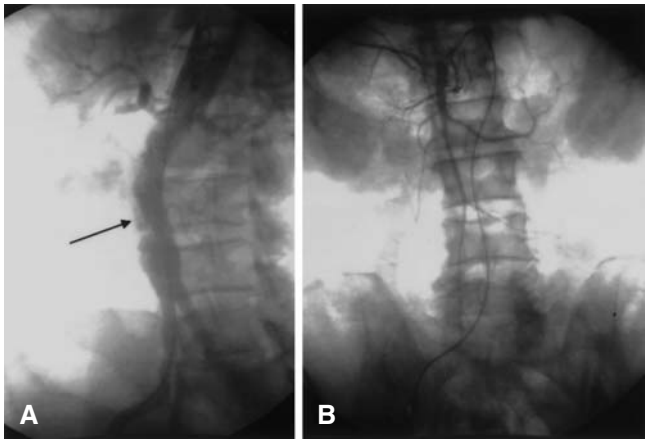

Fig. 1. A Aortogram showed severe irregularity of the infrarenal abdominal aortic wall, including ulceration (*arrow*). **B** Superior mesenteric arteriogram did not show any stenosis

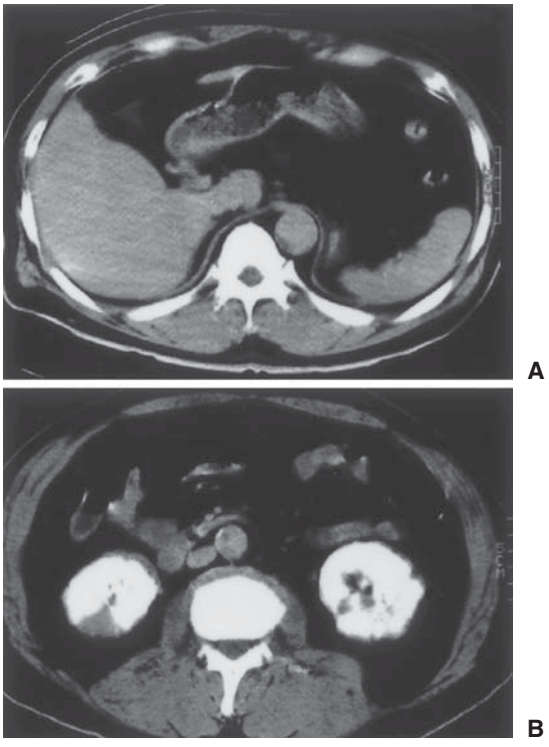

Fig. 2. Enhanced computed tomography demonstrated irregularly shaped intraluminal thrombi in the suprarenal (**A**) and infrarenal (**B**) abdominal aorta

hemodialysis filtration was started and 3 days after the thrombectomy, a turbid malodorous discharge began draining out of an intra-abdominal tube. Immediate re-exploration of the abdomen revealed massive inoperable intestinal necrosis. The patient died the next day.

Autopsy findings revealed that the superior mesenteric artery, the mesenteric branches, and the marginal arteries were patent, but the origin of the inferior mesenteric artery was not. The resected cecum showed transmural necroses. Histologic investigation of the abdominal aorta disclosed the presence of cholesterol crystals in the fragile atheromatous thrombi (Fig. 3).

Discussion

Shaggy aorta syndrome is a rare and poorly understood disorder. Hollier et al.[1] and Kazmier[2] defined this syndrome as spontaneous atheromatous visceral embolization from diffuse aortic atherosclerotic disease. This case was very unusual because atheromatous visceral embolization occurred while treating the macroembolism. The patient's symptoms were progressive, severe, and ultimately incorrigible. Thus, we stress the importance of confirming the diagnosis early so that appropriate therapy can be initiated without delay.

Not all patients who have the characteristic shaggy-like findings on CT and angiogram have microembolization. Shaggy aorta syndrome can only be suspected clinically, even with the help of imaging

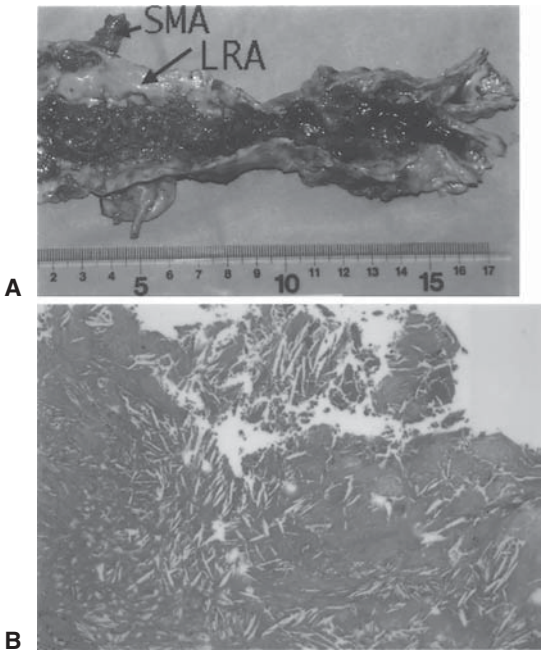

Fig. 3. A Photography of the postmortem abdominal aortic specimen, showing diffuse atheromatous thrombi with ulceration on the flow surface. *SMA*, superior mesenteric artery; *LRA*, orifice of the left renal artery. **B** Histologic specimen of mural thrombus on the flow surface, demonstrating needle-like cholesterol crystals (H&E, ×40)

diagnostic modalities such as CT and angiogram. The diagnosis is only confirmed by the presence of cholesterol crystals in the arterioles and small arteries of the muscle, skin, and kidneys. Unfortunately, we did not take a biopsy, but performed arteriography in the hope of determining the cause of the acute abdominal symptoms after the thrombectomy. The possibility of this syndrome did not occur to us at the time; however, in retrospect, it may have been more appropriate to perform an enhanced CT examination before angiography. A diagnostic approach involving any intra-arterial catheterization may always have a high potential of inducing severe embolization, even if deliberately manipulated.[3] Intravenous digital subtraction aortography could be an alternative option.

The discontinuation of anticoagulants in the presence of shaggy aorta syndrome has been advised.[4,5] In our hospital, heparin is routinely given to prevent secondary thrombosis after thrombectomy, but this might have aggravated the symptoms following thrombectomy in this patient. Therefore, action to prevent the worst possible sequela must be taken if this syndrome is suspected.

Complete removal of the diseased aorta is usually impossible, according to the literature.[1,2] Hollier et al. described the efficacy of an axillo-bifemoral bypass with bilateral external iliac artery ligations, emphasizing the acceleration of fibrin deposition on the aortic wall due to the reduction in blood flow and flow velocity.[1] When we reconsider the therapeutic options for our patient, an axillo-bifemoral bypass with bilateral iliac ligations might have saved him.

In summary, an enhanced CT scan should be done before intra-arterial catheterization if shaggy aorta syndrome is suspected. The point of suspicion may arise at any time, but particularly when a patient complains of abdominal pain after treatment for a macroembolism of the lower limb, as in our patient. Intravenous digital subtraction aortography may be a helpful diagnostic alternative.

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