Severe Obstructive Calcification of the Descending Aorta: A Case Report of “Coral Reef Aorta”

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An 82-year-old man suffering from lower back pain and dyspnea presented to our institute in a state of shock. Computed tomography showed subtotal occlusion of the descending aorta with massive atherosclerotic calcification. As the proximal portion of the superior mesenteric artery was obstructed, emergency bypass from the right axillary artery to the bilateral external iliac arteries was performed, but the patient died 2 days later. Autopsy revealed that reddish-brown and verrucous masses obstructed the descending aorta, and high-grade thickening of the intima and extensive deposits of calcium in the lumina and medial layer were detected in the descending aorta histologically.

Keywords: coral reef aorta, calcification, aorta

Introduction

Obstructive atherosclerotic disease of large vessels generally occurs in the infrarenal aorta. Localized obstruction of the suprarenal aorta is rare, and obstruction of the descending aorta might be rarer. In 1984, Qvarfordt et al.1) reported nine patients with heavily calcified suprarenal obstructive atherosclerotic lesions and named this entity “coral reef aorta.” Heavily calcified plaques grow into the lumen and can cause significant stenosis and malperfusion of the visceral arteries or lower limbs. We report a patient who required emergent operation because of hemodynamically significant symptomatic stenosis of the descending aorta, which caused ischemia of the intestine and lower limbs.

Case Report

An 82-year-old man presented to the emergency department with complaints of acute lower back pain and dyspnea. He had a history of hypertension and peripheral arterial disease but no family history of aortic disease. His renal function was in the normal range, and he did not have diabetes mellitus. He had undergone percutaneous transluminal angioplasty for stenosis of the bilateral iliac arteries three times in the last 9 years. After angioplasty,
intermittent claudication continued to appear on his left foot and his ankle brachial index remained at 0.6–0.8 on the right side and 0.5–0.6 on the left side as the stents did not expand sufficiently against severe calcification of the common iliac arteries. Computed tomography (CT) after angioplasty 10 months earlier (Fig. 1) showed severe calcification of the descending aorta and bilateral common iliac arteries. He was in shock, with tachypnea and impaired consciousness. Physical examination revealed cyanosis in the lower half of the body, board-like rigidity of the abdomen, and a pulseless bilateral femoral artery. Arterial blood gas showed severe metabolic acidosis, with a pH of 7.113, BE of $-17.3$ mmol/L, and anion gap of 21.8. A blood test revealed a serum calcium level of 9.1 mg/dL and a serum phosphate level of 5.9 mg/dL.

Emergent thoracoabdominal CT scan showed subtotal occlusion of the descending aorta with massive atherosclerotic calcification (Figs. 2A–2C). Hyperdense gritty materials had accumulated at the aortic bifurcation (Figs. 2A, 2B, and 2D), which had not been observed 10 months earlier. The celiac artery was not detected. The superior mesenteric artery (SMA) was obstructed approximately 10 cm from its orifice by the hyperdense mass, and the peripheral branches of the SMA showed no contrast.

To improve the decreased blood flow to the lower half of the body due to the obstruction of the descending aorta, we performed emergent bypass surgery from the right axillary artery to the bilateral external iliac arteries with an 8 × 8-mm expanded polytetrafluoroethylene graft. The pulsation of the bilateral femoral arteries was highly palpable after bypass surgery, and endoluminal re-vascularization of the SMA was attempted. However, aortography showed no blood flow in the peripheral branches of the SMA. After prompt laparotomy, resection of the necrotic small intestine was performed. Despite the intensive care, including continuous arterial injection of an anticoagulant, he died on the second postoperative day.

Autopsy revealed a highly calcified aorta with no elasticity. Rock-hard, reddish-brown, and verrucous masses obstructed the lumen of the descending aorta (Fig. 2E). Deposits of atheromatous material and a rugged intima surface were found, particularly around the ostium of
the visceral arteries. Calcified plaques and ulcers of the intima were found everywhere (Figs. 3A and 3B). A verrucous calcified mass also accumulated and obstructed the bifurcation of the aorta. The celiac artery could not be detected from either inside or outside the aorta. Histologically, high-grade thickening of the intima and extensive deposits of calcium in the lumina and medial layer were detected in the descending aorta (Figs. 3C and 3D), and similar lesions were found in the entire aorta, including the brachiocephalic (Fig. 3E) and internal carotid artery (Fig. 3F). Intraluminal calcium deposits nearly obstructed the SMA (Fig. 3G), and small deposits were found in the left renal artery (Fig. 3H). These observations suggested that coral reef calcification occurred not only in the aorta.
but also in its branches.

Discussion

Qvarfordt et al. described nine patients with heavily calcified suprarenal aortic disease and named this rare entity “coral reef aorta,” which is characterized by severe atherosclerosis and calcification of the abdominal aorta with stenosis. Heavily calcified plaques grow into the lumen more extensively than those observed in routine atherosclerosis, and the lesions tend to be located on the posterior surface of the aorta. All nine patients reported in the above study were women, but the majority of later studies have not found any significant difference in gender distribution. The estimated frequency of this disease ranges between 0.6% and 1.8%, and the age distribution is as wide as 3–79 years. From our investigations, a difference in prevalence according to ethnicities has not been reported.

The common symptoms of coral reef aorta are renovascular hypertension, intermittent claudication, and visceral ischemia. Although the pathogenesis of coral reef aorta remains uncertain, Grotmeyer et al., and Schlieper et al. suggest low serum levels of calcification inhibitors such as fetuin-A and uncarboxylated matrix gla protein (ucMGP) are related to its development. In this particular patient, fetuin-A and ucMGP levels were not measured because of the emergency situation, but serum ions known to contribute to vascular calcification were normal.

Many reports have argued the necessity of surgery because coral reef aorta is considered potentially life-threatening if untreated and its in-hospital mortality rate is reportedly as high as 11.6%–13.3%.,. Conventional surgeries such as thromboendarterectomy, replacement of the thoracoabdominal aorta, or bypass surgery to the iliac artery are performed in accordance with the localization of the lesion. The operative mortality was reported to be as high as 8.7%–11.6%, and the rate of postoperative complications requiring corrective surgery was reported to be 13.9%–15.9%. The placement of stent grafts and laparoscopic aortic reef removal were recently reported. In treatments using stent grafts, however, the surgeon should be aware of the possibility that detachment of the atherosclerotic debris causes distal embolization, and stent graft fracture occurs because of severe calcification. In addition, paraplegia may occur when a stent graft is placed at the thoracoabdominal transition. Because the application of these surgeries is limited by anatomical aspects, case-by-case evaluation is mandatory when treating coral reef aorta.

Conclusion

We reported a rare case of severe obstructive calcification in the descending aorta, called “coral reef aorta.” Coral reef aorta is potentially life-threatening and the in-hospital mortality rate is still high.

Disclosure Statement

All authors have no conflict of interest.

Author Contributions

Writing: TI
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

References

1) Qvarfordt PG, Reilly LM, Sedwitz MM, et al. “Coral reef” atherosclerosis of the suprarenal aorta: unique clinical entity. J Vasc Surg 1984; 1: 903-9.
2) Grotmeyer D, Pourhassan S, Rehbein H, et al. The coral reef aorta—a single centre experience in 70 patients. Int J Angiol 2007; 16: 98-105.
3) Schlieper G, Grotmeyer D, Aretz A, et al. Analysis of calcifications in patients with coral reef aorta. Ann Vasc Surg 2010; 24: 408-14.
4) Minnee RC, Idu MM, Balm R. Coral reef aorta: case reports and review of the literature. Eur J Vasc Endovasc Surg 2005; 29: 537.
5) Belczak SQ, Sincos IR, Aun R, et al. Coral reef aorta, emergency surgical: case report and literature review. Einstein (Sao Paulo) 2014; 12: 237-41.
6) Sagban AT, Grotmeyer D, Rehbein H, et al. Occlusive aortic disease as coral reef aorta—experience in 80 cases. Zentralbl Chir 2010; 135: 438-44.
7) Holfeld J, Gottardi R, Zimpfer D, et al. Treatment of symptomatic coral reef aorta by endovascular stent-graft placement. Ann Thorac Surg 2008; 85: 1817-9.
8) Di Centa I, Coggia M, Javerliat I, et al. Total laparoscopic suprarenal aortic coral reef removal. J Vasc Surg 2006; 44: 194-7.