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Open Surgical Treatment of a Severe Case of Obstructive Calcifying Aortic Disease

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The obstructive calcifying aortic disease refers to severe calcifications of the descending aorta that obstruct or slow blood flow. Here, we report the case of a 65-year-old woman with recent onset of a very tight intermittent claudication and concomitant severe and uncontrolled hypertension, treated with a bypass graft between the proximal descending thoracic aorta and the supra-visceral abdominal aorta.

Severe stenosis or total occlusion of the descending aorta usually involving the origin of the visceral and renal arteries is a rare pathologic entity usually called middle aortic syndrome.[1] The commonest etiologies are congenital disorders or inflammatory disease, and adolescents or young people are more commonly affected. Herein, we describe the case of an unusual localization limited to the descending aorta and not involving the visceral vessels without any congenital or inflammatory explanation.

CASE REPORT

A 65-year-old woman was admitted to our department because of worsening intermittent claudication of the lower limbs and uncontrolled severe hypertension despite 3 medications (β blockers, diuretic, α1-adrenergic blocker). Her past medical history also included type 2 diabetes mellitus treated with oral hypoglycemic agents and dyslipidemia treated with statins; no history of smoke was present. Over the past months, she experienced worsening of general health, with episodes of intense asthenia, headache, dizziness and disorientation, and dyspnea after mild exertion. Physical examination revealed bilateral hyposphygmic femoral pulses and the absence of peripheral pulses. Ankle-brachial index (ABI) was 0.3 on both sides.

Echo-color Doppler of the proximal abdominal aorta and iliac arteries showed poststenotic flow in the celiac trunk, superior mesenteric artery, both renal arteries, and the iliac and femoropopliteal axes bilaterally. The supra-aortic vessels appeared normal. Echocardiography demonstrated mild left ventricular hypertrophy, ectasia of the ascending aorta, and calcific stenosis of the descending aorta downstream from the isthmus. A computed tomography angiography (CTA) showed a normal ascending aorta and aortic arch, a severely calcified descending aorta with a pseudocoarctation in the proximal portion (18-mm diameter) and tight stenosis in the mid-distal segment, with reduction of the vessel lumen to a few millimeters just above the celiac trunk (Fig. 1). The splanchnic arteries, the infrarenal aorta, and the renal and the iliac arteries, although severely hypoperfused, were all morphologically normal and free from calcifications.

Preoperative blood chemistry showed no significant anomalies. Spirometry demonstrated mild-to-moderate restrictive lung function. Coronary angiography revealed no abnormalities of the coronary arteries, and aortography showed an 85−mm Hg pressure gradient at the lower extremity of the restricted aorta. Total body positron emission tomography was performed to rule out focal inflammatory processes in the aorta, but it did not show any abnormal tracer distribution. Antibody testing for TPHA, VDRL, c-ANA, p-ANA, and anti ds-DNA was negative.

An endovascular approach was ruled out because of the presence of full thickness and uninterrupted

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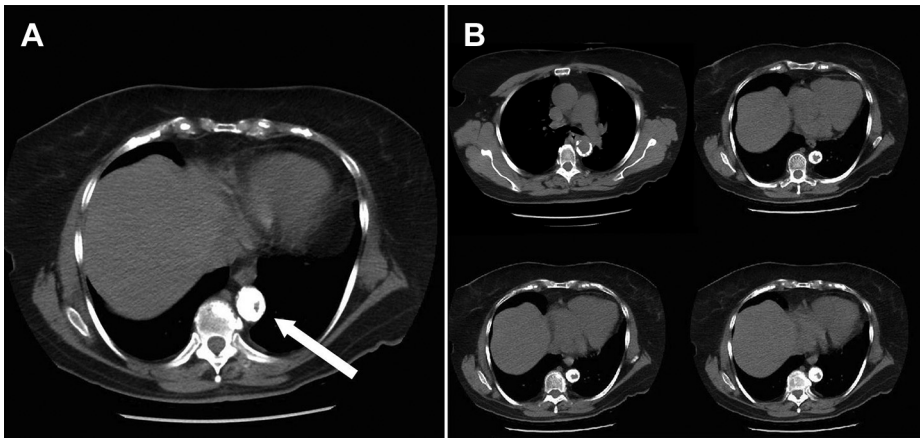
Fig. 1. (A) Preoperative CTA scan without contrast showing the tightest stenotic point of the thoracic aorta (*arrow*). **(B)** Full-thickness calcifications of the vessel wall at different levels.

calcifications along the entire descending aorta. Because of the patient's severe symptoms, surgery was then planned.

Through a thoracophrenolombotomy at the sixth intercostal space, the aorta was isolated from the left subclavian artery down to the renal arteries. After tangential clamping of the descending aorta immediately distally to the left subclavian artery, an end-to-side anastomosis was performed between the aorta and a 20-mm Dacron graft with a 3/0 polypropylene (Fig. 2A). Then, an aortotomy was made proximally to the celiac trunk, after tangential clamping of the vessel and an end-to-side beveled anastomosis was performed with a 3/0 polypropylene involving the ostia of the celiac trunk and the superior mesenteric artery (Fig. 2B).

Histopathology and microbiology showed nothing but calcifications of the stenotic abdominal aortic wall with absence of bacterial or viral pathogens. The postoperative course was uneventful. Distal pulses could be appreciated; the ABI was 1 and intermittent claudication resolved. Three months later, blood pressure was brought under control with a single therapy (α1 adrenergic blocker) and headaches remitted. A CTA was performed 1, 6, and 12 months after surgery and demonstrated a patent graft without vascular anastomosis defects (Fig. 2C).

DISCUSSION

Aortic coarctation refers to an abnormal narrowing of the aortic lumen, with hemodynamically significant alteration of downstream blood flow. The middle aortic coarctation is a rare syndrome characterized by severe calcifications of the descending aorta, mostly affecting adolescents or young adults.[1] It may be caused by congenital or inflammatory disorders, whose commonest disease is the Takayasu

arteritis.[2] A similar nosologic entity is the ''coral reef aorta,'' which however involves the portion between the diaphragm and the suprarenal part of this vessel (visceral aorta).[3] Mild stenosis may be completely asymptomatic; with the progression of the disease, symptoms may arise, such as dyspnea, lipothymic episodes, fatigue, headache, upper limb hypertension, and lower limb hypoperfusion with claudication.[1]

Diagnostic suspicion of a steno-occlusion of the thoracic aorta may be prompted by echo-color Doppler findings of poststenotic blood flow abnormalities in the splanchnic and renal arteries and the downstream vessels bilaterally, as seen in this patient. As the disorder can localize to any segment of the aorta, second-level imaging will better reveal the relationship between the steno-occlusion and the emergence of supra-aortic vessels and help to evaluate the extension of lumen narrowing, which actually involves the infrarenal aorta in <2% of cases.[4] On the contrary, the visceral abdominal aorta is more often involved, and the renal and the splanchnic arteries (87% and 62% of cases, respectively).[5]

In the search for possible causes of extensive stenosis of the descending thoracic aorta, clues from the patient's medical history and clinical presentation and angiographic and histopathologic findings will help in differential diagnosis between chronic inflammatory processes (arteritis, connective tissue disease),[6] atherosclerosis, or infection. In our patient, laboratory and histopathologic examinations were negative. Symptoms arose in adult age; thus, a congenital etiology is unlikely. Therefore, this might not be a case of middle aortic syndrome. The only explanation we can postulate is a
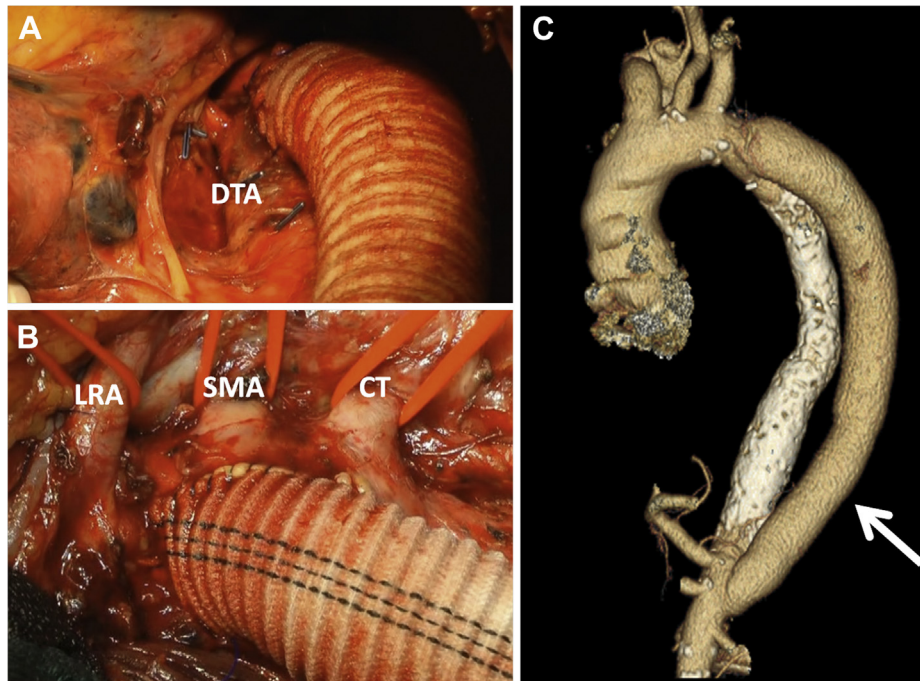
Fig. 2. (A) Proximal anastomosis at the origin of the descending thoracic aorta (DTA); **(B)** Distal anastomosis at the celiac trunk (CT) and the superior mesenteric artery (SMA) and above the left renal artery (LRA); **(C)** Postoperative 3D CTA scan at 12 months showing the patency of the graft (*arrow*).

progressively worsening calcification process of unknown origin of the descending thoracic aorta, which led to a left ventricular hypertrophy and uncontrolled hypertension, despite a multidrug antihypertensive therapy. Many anatomic and morphologic patterns have been described for this syndrome.[2] Although the calcific component was suggestive of atherosclerotic degeneration, the descending thoracic aorta was the only vessel involved, whereas the splanchnic arteries, the infrarenal aorta, the iliac arteries, the supra-aortic trunks, and the coronary arteries all appeared normal. We searched the literature to better identify this clinical case. In 1982, Guilmet et al.[7] reported the first patient with severe aortic narrowing limited to the descending thoracic aorta not involving the visceral vessels, because of hard calcifications and they called this condition ''obstructive calcifying aortic disease.'' He treated it with resection and Dacron repair. Three years later, Patra et al.[8] published 2 cases, one of them involving only the descending thoracic aorta and the other one the thoracoabdominal aorta; in the first case, again, the patient underwent open repair. Recently, a similar case involving the descending aorta but not the visceral vessels treated with surgical bypass has been published by Price et al.,[9] but some substantial differences with our case must be pointed out. First,

we performed a thoracophrenolombotomy, with circumferential section of the diaphragm, instead of 2 different incisions (thoracotomy plus lombotomy) and the creation of a tunnel through it. Second, our patient had a late onset of symptoms, with no history of juvenile hypertension; this could rule out a congenital etiology. Third, we performed the distal anastomosis at a supraceliac level instead of at the aortic bifurcation. This allowed an antegrade blood flow to all visceral vessels, which were not involved by calcifications.

As a surgical alternative, we might have considered a less-invasive extra anatomic approach such as an axillobifemoral bypass, but we considered the patient fit enough to undergo a more radical, hemodynamically effective, and long-lasting treatment without significant surgical risks.

Endovascular techniques were not suitable for this case as calcifications involved almost all the descending thoracic aorta. Children and high-risk patients with focal stenoses might, instead, be eligible candidates for at least an attempt of balloon angioplasty and stenting.[10]

The obstructive calcifying aortic disease is a polyedric condition which needs prompt intervention to avoid severe complications. Open surgery remains the gold standard; if calcifications do not involve the visceral vessels, an off-pump bypass

with tangential clampings is a relatively safe and hemodynamically effective procedure.

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