Endovascular stent graft exclusion of a thoracic arteriovenous malformation in a patient with Cobb syndrome

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Cobb syndrome is a rare neurocutaneous disorder characterized by spinal vascular abnormalities in association with a vascular lesion of the skin at the same metamere. Patients present after the onset of symptoms such as paraplegia, back pain, and, less commonly, fatigue due to heart failure. Available treatment options to date have included neurosurgical resection and endovascular embolization. We present a patient with Cobb syndrome with a progressively symptomatic arteriovenous malformation involving the thoracic vertebrae and left hemithorax, with extensive blood supply from the T3 to T10 intercostal arteries, and demonstrate successful endovascular thoracic stent graft placement and coil embolization. (J Vasc Surg Cases 2016;2:7-9.)

Cobb syndrome, or cutaneomeningospinal angiomatosis, is a nonfamilial, neurocutaneous disorder characterized by spinal vascular abnormalities in association with a vascular lesion of the skin at the same metamere.[1] Patients are diagnosed at any age but most commonly in late childhood and typically after the onset of neurologic symptoms such as paraplegia, quadriplegia, back pain, and, less frequently, fatigue due to heart failure.[2-5] Available treatment options to date have been mainly limited to neurosurgical resection and endovascular embolization with coils or liquid embolic agents, with occasional use of irradiation or corticosteroid therapy.[2,3,6] We present a case of a progressively symptomatic high-output cardiac disease caused by a thoracic arteriovenous malformation (AVM) in a patient with Cobb syndrome and demonstrate successful endovascular thoracic stent graft placement and coil embolization. The patient consented to the publication of this case report.

CASE REPORT

History and examination. A 37-year-old man with a history of multiple subcutaneous skin nodules since birth initially presented to his primary care physician with complaints of increasing dyspnea on exertion, fatigue, and back discomfort. He had no other known medical problems. On examination, he had a bruit throughout the precordium and left hemithorax. He also had a palpable supraclavicular thrill, and examination of his skin revealed multiple subcutaneous nodules.

The patient underwent computed tomography (CT) and magnetic resonance imaging, which demonstrated a massive AVM involving the thoracic vertebrae and left hemithorax, with extensive blood supply from the T3 to T10 intercostal arteries and dilated azygous and hemiazygous venous structures (Fig 1). His cardiac output based on an echocardiogram was 10 L/min. A specimen from a soft tissue biopsy of his skin lesion was consistent with angiolipoma. At this stage the diagnosis of Cobb syndrome was made.

Intervention and postintervention course. The patient underwent placement of 26-mm × 10-cm and 28-mm × 10-cm TAG stent grafts (W. L. Gore and Associates, Flagstaff, Ariz) to cover the blood supply to his AVM from the third intercostal artery to immediately above the celiac artery. Additional coil embolization of the T10 intercostal artery using four 7-mm × 3-mm endovascular coils was chosen instead of extending stent graft coverage inferiorly because of the proximity of the celiac artery and completion angiography demonstrated reduction in arterial flow to the AVM (Fig 2).

Because of concern for the risk of thrombosis in the patient's dilated azygous and hemiazygous system with the sudden change in hemodynamics and decreased cardiac output, he remained anticoagulated on warfarin for 3 months postprocedure. At the 1-year follow-up, the patient reported complete relief of back pain and resolution of dyspnea on exertion. His postoperative echocardiogram confirmed a decrease in cardiac output to 5 L/min. The follow-up CT scan at that time showed increased attenuation of the aorta with decreased attenuation of the AVM and azygous vein, suggesting decreased collateral vascularity of his previous AVM; however, the overall size of the azygous and hemiazygous system remained grossly unchanged.

DISCUSSION

Cobb syndrome is a rare neurocutaneous disorder associated with a number of different spinal vascular

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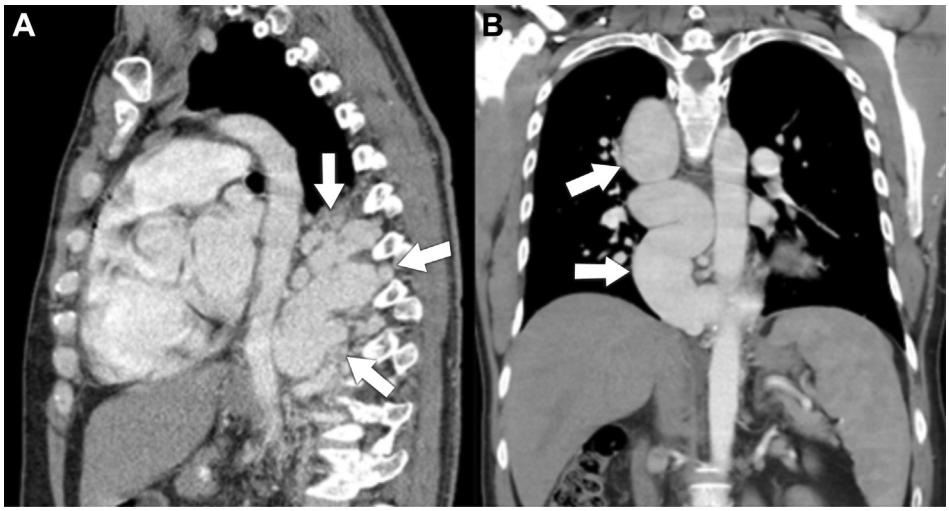

Fig 1. Computed tomography (CT) imaging of **(A)** arteriovenous malformation (AVM) with blood supply from T3 to T10 intercostal arteries (*arrows*) and **(B)** dilated azygous and hemiazygous venous structures (*arrows*).

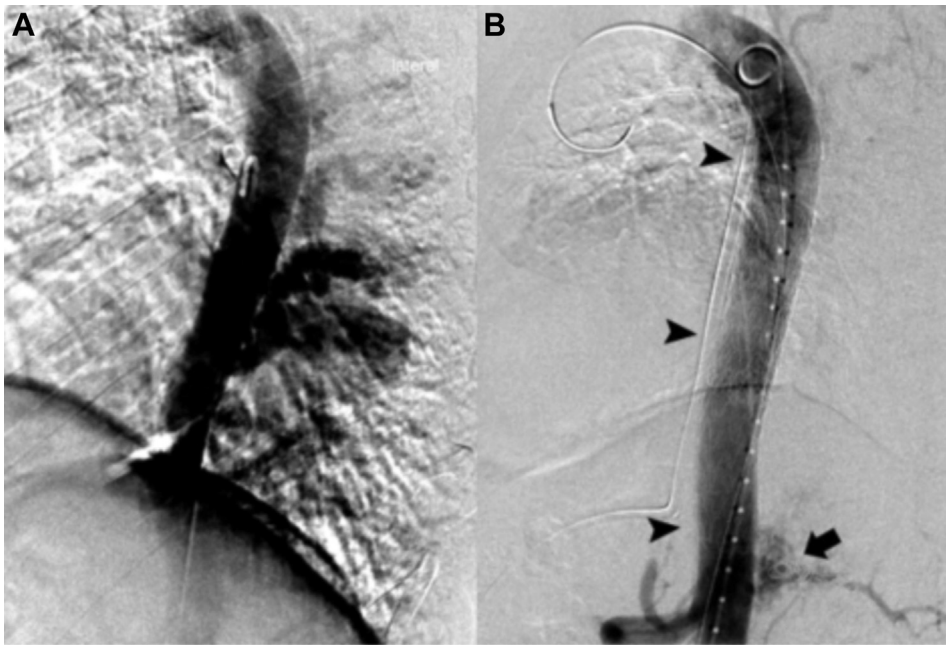

Fig 2. Preoperative angiogram shows **(A)** with multiple enlarged arteries supplying the arteriovenous malformation (AVM) and **(B)** exclusion of the AVM after graft placement (*arrowheads*) with coil embolization at T10 (*arrow*).

pathologies, most commonly AVMs and occasionally cavernous hemangiomas and angiomas.[7-10] This syndrome is thought to have an under-reported prevalence because the correct diagnosis is usually not recognized in patients until they are symptomatic, most often with effects of physical spinal cord compression or steal syndrome.[3-6,11] Increased awareness of the syndrome could contribute to recognizing that cutaneous vascular lesions may signify a spinal or paraspinal pathology leading to neurologic or cardiovascular symptoms.[7]

Treatment options have generally been restricted to surgical intervention, endovascular embolization with coils or liquid embolic agents, irradiation, and steroid therapy. The patient presented in this case report had progressively symptomatic high output cardiac disease secondary to steal syndrome from his thoracic AVM, and underwent successful endovascular thoracic stent graft placement and coil embolization. His subcutaneous skin nodules, determined to be angiolipomas on the biopsy specimen, were not resected because they were asymptomatic and benign,

and will be monitored for progression. The appropriate treatment option, however, needs to be tailored to fit the individual patient.

Our patient had an AVM that was being supplied by numerous intercostal arteries, which made possible covering most of the AVM with an aortic stent graft instead of multiple coils. Proximity of the inferior border of the AVM to the celiac artery precluded attempting complete coverage with the aortic stent graft, and coils were used at the T10 intercostal artery.

The reported incidence of spinal cord ischemia after thoracic endovascular aortic repair is 4% to 7%, and risk factors include the extent of disease and prior distal aortic repair.[12,13] The anterior spinal artery, which supplies the spinal cord along with the posterior spinal artery, receives collateral supply from the intercostal and lumbar segmental arteries. We believed in this patient, there was a low probability of the spinal cord perfusion relying on the intercostal arteries because the AVM was part of a low resistance system. Because of this, coupled with the risk of placement of a lumbar drain due to its proximity to the spinal cord, we elected to not place a drain after discussion of the risk of paraplegia after the procedure with the patient.

Reduction in the blood supply to the AVM with a 50% decrease in cardiac output led to improvement in the patient's symptoms, and repeat CT scan 1 year later revealed decreased collateral vascularity of his previous AVM. He has had follow-up to 18 months postoperatively and remains with significant relief of symptoms. Predicting his long-term outcome is challenging due to the limited follow-up in this population.

A recent retrospective literature review of 28 patients with Cobb syndrome managed with embolization demonstrated a statistically significant angiographic progression of disease compared with patients with spinal cord AVM alone.[14] Although brain AVMs require complete obliteration of the nidus rather than inflow occlusion to decrease the risk of recurrence and hemorrhage, management of spinal AVMs necessitates consideration of the generally poor prognosis of these patients and the high risk of treatment-related complications. Partial treatment of spinal AVMs can result in stabilization of disease and improvement of symptoms, even without total angiographic cure.[15-17] We preferred an initial endovascular repair in this patient because it is less invasive than surgical resection, with decreased morbidity and mortality, but it is possible that inflow occlusion may result in additional inflow being recruited in the future. Coverage of the arterial inflow may also limit access to the nidus for further endovascular intervention, but open surgical resection remains an option for management of possible future recurrence.

CONCLUSIONS

Cobb syndrome is a rare and unusual disorder that must be considered when patients present with vascular cutaneous malformations and neurologic deficits. Optimal treatment must be tailored to the individual anatomy and pathophysiology and may require a combination of endovascular techniques. To our knowledge, this is the first reported thoracic endovascular stent graft repair of the descending aorta for a patient with Cobb syndrome. In selected patients, endovascular stent graft placement may present a feasible option compared with endovascular coiling alone and does not carry the morbidity of a surgical resection.

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