Management of celiac trunk and superior mesenteric artery synchronous aneurysms as an extremely rare manifestation of Wegener granulomatosis

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ABSTRACT

Large-vessel aneurysm is an extremely rare complication of Wegener granulomatosis. We report a case of Wegener granulomatosis in a 49-year-old woman with large synchronous aneurysms of the celiac trunk (54 mm) and superior mesenteric artery (42 mm) who presented with abdominal pain. Because of the large diameter of the aneurysms and their proximity to each other, a combination of endovascular and hybrid repair was used for management. After surgical debranching and endovascular repair, the patient was discharged in good general condition. We concluded that abdominal pain in Wegener granulomatosis can be a rare manifestation of a large visceral aneurysm. (J Vasc Surg Cases and Innovative Techniques 2019;5:525-8.)

Keywords: Wegener granulomatosis; Aneurysm; Celiac artery; Superior mesenteric artery

Wegener granulomatosis is an autoimmune, systemic inflammatory disorder that occurs as a necrotizing vasculitis and involves small and medium-sized vessels of the upper airways and kidneys.[1-3] Arterial aneurysm, one of the rare presentations of this disease, can be caused by chronic inflammation.[4,5] However, aneurysm in Wegener granulomatosis patients usually affects the small and medium-sized vessels, and involvement of the large vessels is extremely rare.[6-8]

We report a rare case of Wegener granulomatosis in a patient who presented with large aneurysms in the proximal ostia of the celiac trunk and the superior mesenteric artery (SMA). The patient consented to publication of the report of her case.

CASE REPORT

A 49-year-old woman was referred to the emergency department with a chief complaint of abdominal pain for 40 days. The patient's pain progressively became worse and was located at the epigastric region with radiation to the back. The pain worsened on lying down but would not improve with any analgesic medicine. The patient had nausea without vomiting and a low-grade fever, significant weight loss, and noncardiac chest pain for 2 months before admission to our emergency department.

The past medical history was significant for Wegener granulomatosis that had been diagnosed 2 years before this admission.

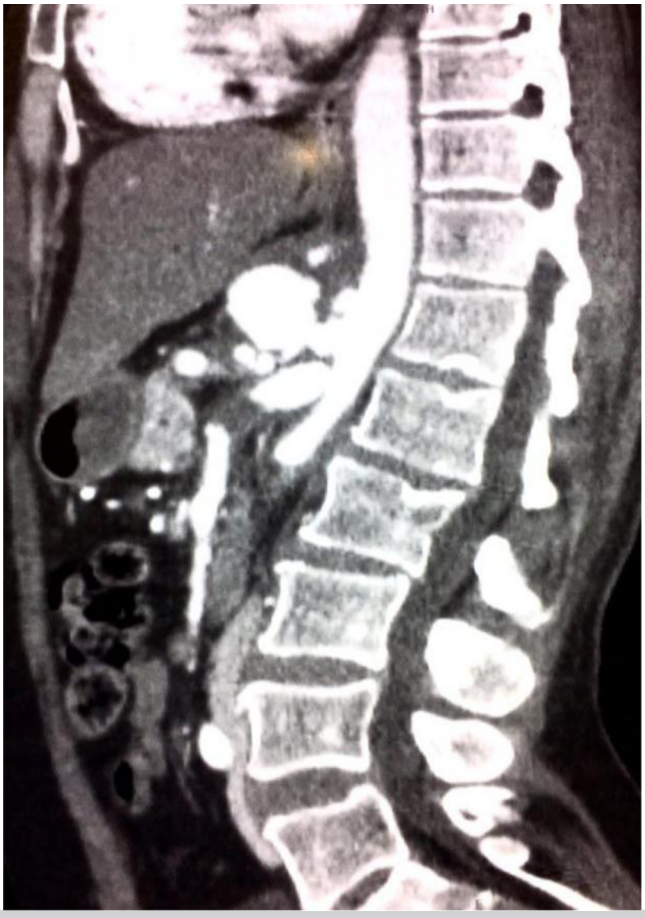

Fig 1. Sagittal section of spiral computed tomography (CT) scan. Aneurysms are seen on this view.

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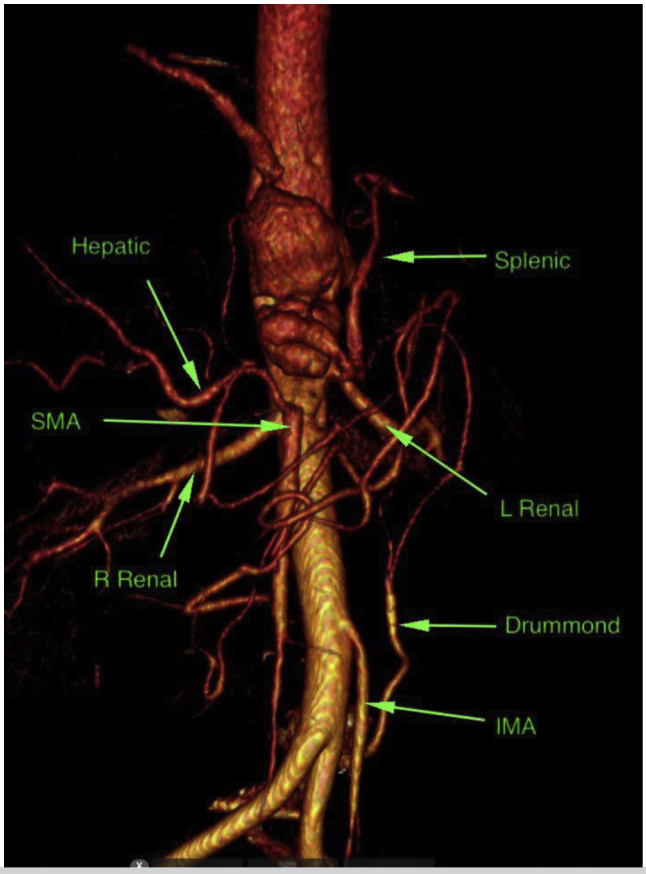

Fig 2. Three-dimensional reconstruction of the patient's preoperative computed tomography (CT) angiography image. Superior mesenteric artery (*SMA*) and celiac artery aneurysms are seen. *IMA,* Inferior mesenteric artery.

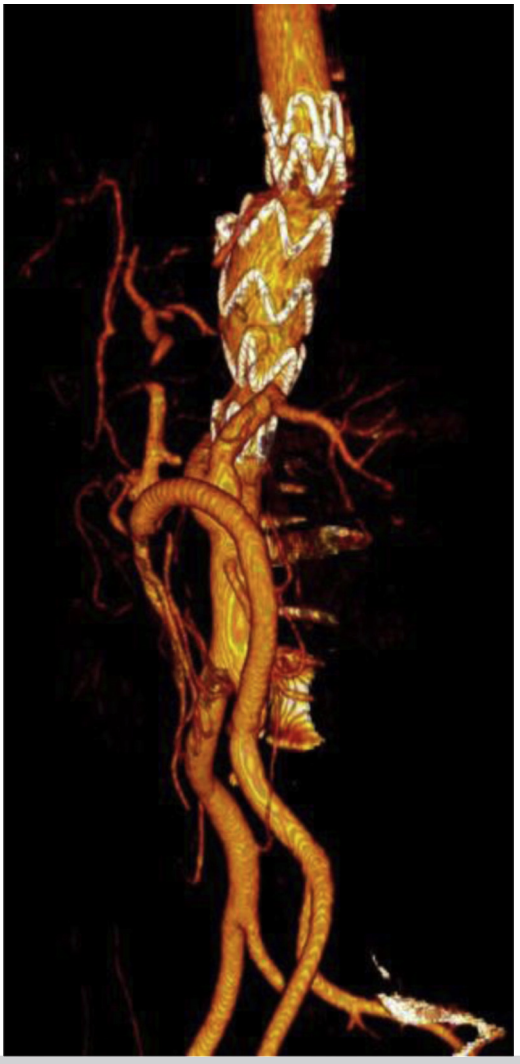

Fig 3. Computed tomography (CT) angiography image after surgery.

The diagnosis of Wegener granulomatosis was made by clinical presentation (such as sinusitis, hemoptysis, and hematuria) and laboratory data (such as presence of cytoplasmic antineutrophil cytoplasmic antibody) in addition to microscopic evaluation of a kidney biopsy specimen. She also had active tuberculosis since the previous year that was diagnosed by bronchoscopy and bronchoalveolar lavage. The tuberculosis has been controlled by drug therapy.

The patient's temperature was 37.3°C; respiratory rate, 18 breaths/min; and blood pressure, 130/80 mm Hg. On physical examination, the abdomen had generalized tenderness, but rebound tenderness and guarding were absent. Furthermore, pulmonary and cardiovascular examination findings were normal. On laboratory examination, the patient had an erythrocyte sedimentation rate of 85 mm/h and C-reactive protein level of 2 mg/dL.

During clinical examination of the patient, spiral computed tomography (CT) of the chest and abdomen with intravenous administration of contrast material was requested. On spiral CT, a 54-mm saccular pseudoaneurysm was identified at the level of the celiac trunk, and another 42-mm saccular pseudoaneurysm was seen at the level of the SMA (Fig 1). Moreover, the origins of the celiac trunk and SMA were mostly occluded, apart from the trunk of the splenic artery. Most of the flow into the SMA and celiac trunk vessels was through pancreatic, adrenal, and inferior mesenteric artery collaterals. The SMA was cut off after the aneurysm because of thrombosis, and immediately after the runoff of the SMA, the common hepatic artery was separated from it.

The patient was taken to a hybrid suite for visceral and renal debranching with endovascular exclusion of the origins of the aneurysmal vessels (Fig 2). Surgical bypass grafting of the renal arteries with ligation of the origin of both arteries was done, followed by bypass grafting of the SMA. The anastomosis of the renal bypass was end to side, and the proximal ends of the renal arteries were ligated to prevent turbulent flow and endoleak after insertion of the stent. In bypass grafting of the renal arteries, the inflow source was the infrarenal aorta; furthermore, in bypass grafting of the SMA, the inflow source was the left common iliac. Selective angiography was conducted within the bypass grafts to ensure patency of the SMA and the renal arteries and effective ligation of the renal arteries.

After bypass grafting, thoracic endovascular aortic repair was done with placement of a thoracic tubular stent graft (Zenith

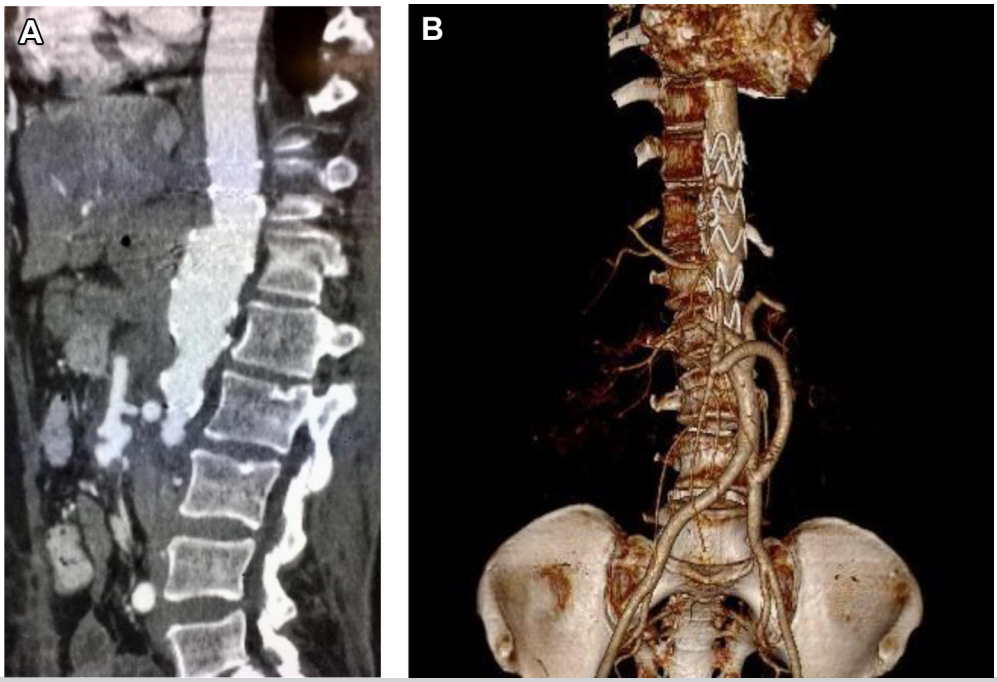

Fig 4. A, Computed tomography (CT) sagittal view 6 months after the operation. **B,** Three-dimensional reconstruction of the CT angiography image.

Alpha Thoracic device, ZTA-P-28-109; Cook Medical, Bloomington, Ind). In addition, five coils (8-mm and 10-mm diameter; MReye Embolization Coils; Cook Medical) were inserted into the celiac trunk by a catheter to prevent retrograde filling of the pseudoaneurysm after thoracic endovascular aortic repair.

After the operation, the patient's abdominal pain subsided, and after 48 hours, CT angiography confirmed the technical success of the procedure (Fig 3). The patient was discharged after 7 days of hospitalization in good general condition. The treatment was continued medically by methylprednisolone and cyclophosphamide. After 6 months, CT angiography was done for follow-up study (Fig 4). There were not any signs of large-vessel aneurysm, and the condition of the patient was good after 6 months.

DISCUSSION

Wegener granulomatosis, one of the antineutrophil cytoplasmic antibody-associated vasculitides, is characterized by necrotizing inflammation of the small and medium-sized vessels.[9,10] This disease usually affects the small vessels of the upper airway (in 90% of cases) and the kidneys (in 75% of cases), but it can also involve skin, joints, and the cardiovascular and nervous systems.[3,6] Large-vessel aneurysms are mostly seen in giant cell and Takayasu arteritis, and they are rare in Wegener granulomatosis.[11]

There have been some reports of large-vessel aneurysms, such as abdominal aorta, renal artery, hepatic artery, subclavian artery, and left gastric artery, in Wegener granulomatosis patients.[12-14] However, to the best of our knowledge, this is the first case that involves the celiac trunk and SMA simultaneously, which is an extremely rare complication of Wegener granulomatosis.

The primary presentations of the large visceral aneurysms of Wegener granulomatosis are abdominal pain and intraperitoneal hemorrhage, which could be fatal if the aneurysm ruptures.[4,14] The basis for treatment of large-vessel aneurysms in Wegener granulomatosis is medical treatment, such as corticosteroids and cyclophosphamide, but this cannot prevent rupture of the aneurysm.[5,15] Our patient was treated medically by corticosteroids before presentation, but because of abdominal pain and large visceral aneurysms, surgical approaches were preferred for management.

We used an endovascular approach followed by a hybrid procedure for management of the aneurysms. An exclusively endovascular procedure was not feasible because of the proximity of the two aneurysms to the visceral arteries; although fenestrated endovascular aneurysm repair could have been done for the patient, it was not possible because of obstruction of the SMA and the long time for the grafts to be made. In addition, because of the long thoracoabdominal incision, long time for aortic clamping, and risk of ischemia for intestines and kidneys, a purely open surgical approach was not suitable for the patient.

Medical treatment was used to continue the treatment. The patient had used methylprednisolone before the operation; however, considering the severity of the disease that caused the aneurysms, cyclophosphamide was added to methylprednisolone for the medical treatment. The combination of methylprednisolone and cyclophosphamide is useful in >90% of severe cases of Wegener granulomatosis.[5]

CONCLUSIONS

Large-vessel aneurysms should be considered a rare manifestation of Wegener granulomatosis. In addition, rupture of a large aneurysm is considered a surgical emergency because it could be fatal. Abdominal pain may be helpful for detecting large-vessel aneurysm. Endovascular surgery followed by a hybrid repair can be a feasible approach in large and multiple aneurysms with proximity to the visceral arteries.

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