Successful treatment of disseminated intravascular coagulation associated with aortic dissection

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ABSTRACT

Disseminated intravascular coagulation (DIC) is an infrequent aortic dissection complication, and its optimal treatment remains controversial. A 55-year-old woman developed DIC associated with Stanford type B aortic dissection, which improved by administration of low-molecular-weight heparin combined with tranexamic acid, but the dissecting aneurysm of the descending aorta was dilated. After thoracic endovascular aortic repair for occlusion of entry tears detected by transesophageal echocardiography, DIC improved without anticoagulant therapy. Three months after treatment, the patient is doing well without complications. Endovascular repair is effective for DIC due to aortic dissection that requires anticoagulant therapy. (J Vasc Surg Cases and Innovative Techniques 2018;4:268-71.)

Keywords: Aortic dissection; Disseminated intravascular coagulation; Endovascular therapy

Chronic disseminated intravascular coagulation (DIC) is a well-known complication of aortic aneurysm, but DIC due to aortic dissection is infrequent. Several treatments of DIC from aortic dissection have been reported.[1-7] Although surgical intervention is sometimes necessary for dilation of the dissecting aortic aneurysm, optimal medical therapy is recommended because of the high risk of surgical repair for bleeding tendency. In addition, endovascular repair for Stanford type B aortic dissection is currently performed frequently and safely.[8-10]

We describe a case of refractory DIC associated with aortic dissection that improved with thoracic endovascular aortic repair (TEVAR). We obtained the patient's consent to the submission and publication of this case report, and she signed a standard publication consent form of our institution.

CASE REPORT

A 55-year-old woman was admitted with sudden back pain. Enhanced computed tomography (CT) revealed acute type B aortic dissection with partial thrombosis of a false lumen, extending from the distal aortic arch to the left common iliac artery. All visceral vessels were patent, the celiac and superior mesenteric arteries originated from the true lumen, and both renal arteries originated from both true and false lumens. There was no clear entry at the descending aorta, and one tear was identified at the celiac artery level (Fig 1, *A*). Moreover, she was diagnosed with Child-Pugh class B liver cirrhosis because of several portosystemic shunts. With uncomplicated type B aortic dissection, she received optimal medical treatment and was discharged 1 month later.

She presented 2 months after discharge with right gluteal ecchymoma and multiple purpura of the extremities. Laboratory examination showed the following values: hemoglobin, 7.0 g/dL; platelets, 12.3 $\times$ 10^3/μL; fibrin degradation product (FDP), 40.7 μg/mL; fibrinogen, 243 mg/dL; plasmin-α_2-plasmin inhibitor complex, 5.1 μg/mL (normal range, <0.8 μg/mL); and thrombin-antithrombin complex, 25.3 ng/dL (normal range, <3.0 ng/dL). Thus, she was admitted for DIC. Antifibrinolytic therapy was started (tranexamic acid 1 g/d intravenously). However, the ecchymoma and purpuras spread; hence, low-molecular-weight heparin (LMWH; heparin calcium, 5000 units subcutaneously, twice daily) was added. Subsequently, the ecchymoma, purpura, and coagulopathy resolved. She was discharged on day 113 after onset of the aortic dissection.

A 6-month follow-up CT scan revealed recanalization of the thrombosed false lumen at the distal aortic arch and dilation of the descending aorta (from 32 to 44 mm), without obvious entries (Fig 1, *B*). Thus, TEVAR was performed as aortic rupture prophylaxis, despite the high surgical risk with DIC and liver cirrhosis. Recanalization and extension of the false lumen could have been caused by LMWH; however, LMWH continuation was considered necessary because of the possibility of DIC exacerbation by its discontinuation. We believed that TEVAR could not only prevent rupture of the dissecting aortic aneurysm but also improve DIC for thrombosis of the false lumen. Cerebrospinal fluid drainage (CSFD) to prevent spinal cord ischemia was not performed because of potential complications, such as epidural hematoma and subarachnoid hemorrhage, that occur with DIC and anticoagulation. Instead, steroid was administered (methylprednisolone, 2 g intravenously) before stent graft deployment, and perioperative hypertensive management was performed.

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Author conflict of interest: none.

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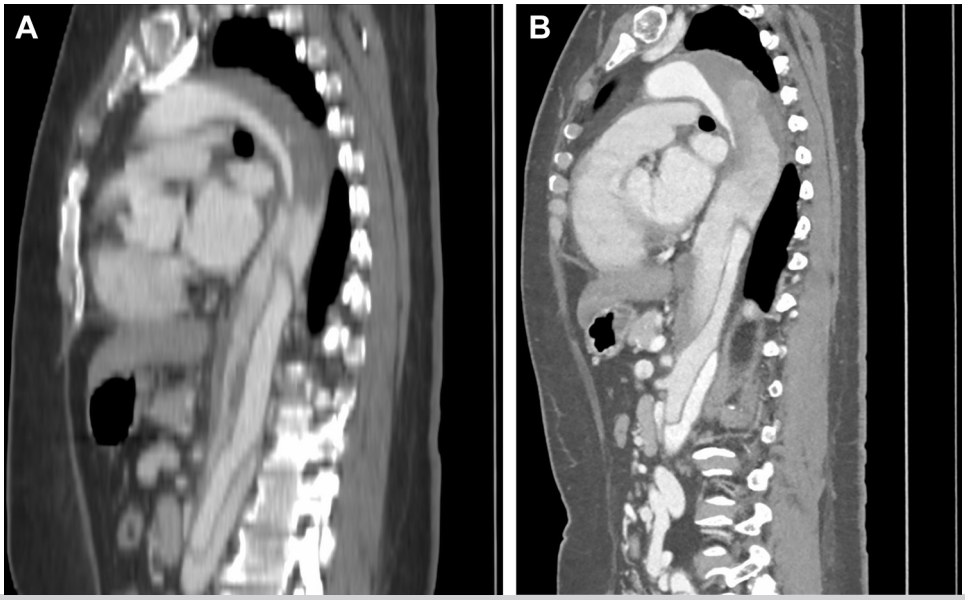

Fig 1. Enhanced preoperative computed tomography (CT) of type B acute aortic dissection. **A,** Enhanced CT scan at onset shows a patent false lumen with partial thrombosis from the descending aorta to the thoracoabdominal aorta. **B,** Recanalization of the thrombosed false lumen and dilation of the descending aorta 6 months after onset.

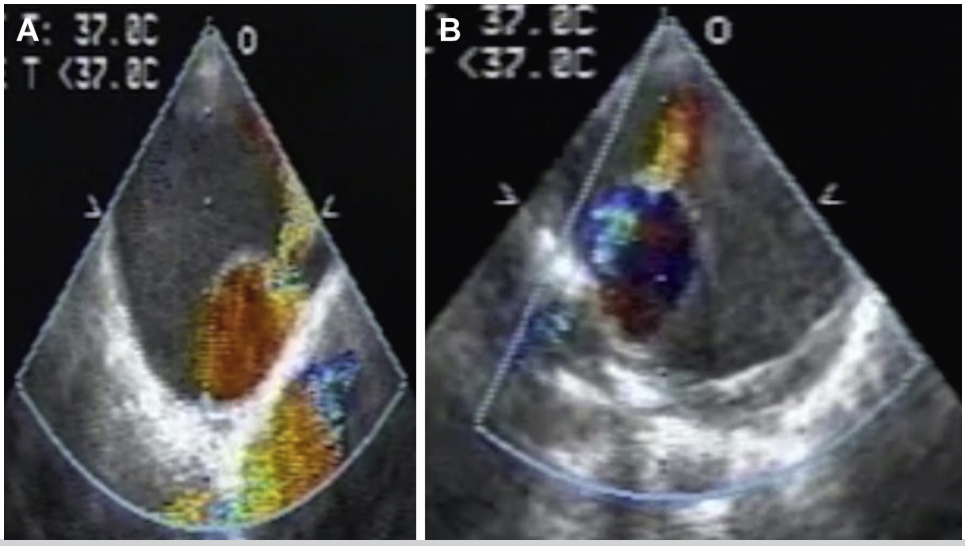

Fig 2. Presurgical transesophageal echocardiography. Both images show stagnant vascular flow of false lumen. **A,** Vascular flow from true lumen to false lumen at T8 spinal level. **B,** Vascular flow from true lumen to false lumen at celiac artery level.

Presurgical transesophageal echocardiography revealed several entry tears at the T6, T8, and T9 spinal levels and the celiac artery level. Color Doppler imaging showed vascular flow, which was turbulent, from the true lumen to the false lumen in the systolic phase at both tear sites (Fig 2). TEVAR was performed to stop the turbulent flow and to reduce flow to the false lumen by closing the entry tears. The Gore cTAG thoracic stent graft (W. L. Gore & Associates, Flagstaff, Ariz) was introduced over a Super Stiff guidewire (Cook Medical, Bloomington, Ind) through a right femoral arteriotomy. Two cTAG thoracic stent grafts were deployed to cover the entry tears from the distal left subclavian artery to the descending aorta at the T10 level (cover length, 180 mm). Angiography confirmed no endoleak or retrograde type A aortic dissection; the procedure proceeded without complications. No hemorrhagic event occurred without anticoagulant therapy after surgery. The patient was discharged on postoperative day 9 without anticoagulant therapy. Enhanced CT on postoperative

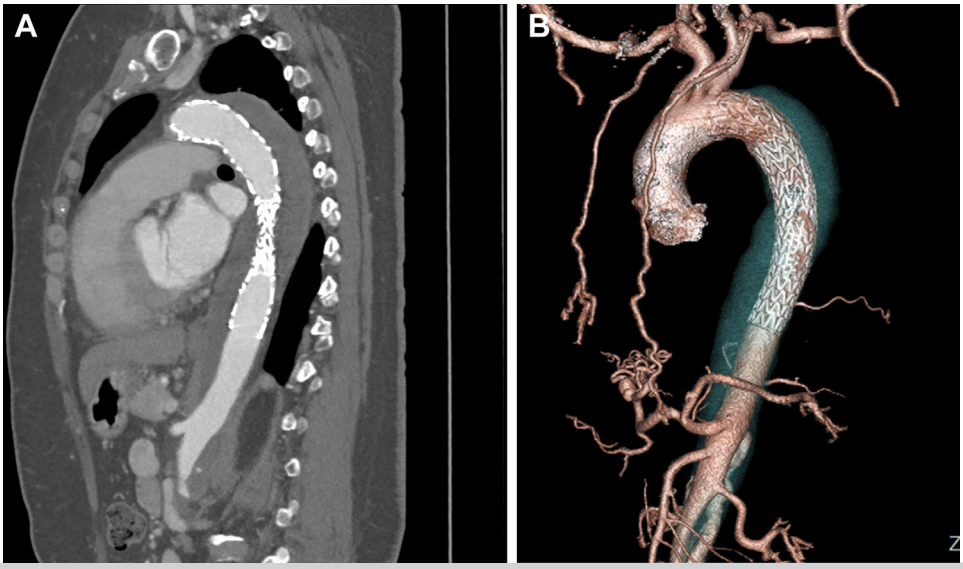

Fig 3. Postoperative computed tomography (CT) angiography at 3-month follow-up. **A,** Complete thrombosis is shown from the distal aortic arch to the descending aorta with no dilation of the dissecting aortic aneurysm. The patent false lumen is seen from the abdominal aorta to the proximal common iliac artery. **B,** Three-dimensional CT angiogram.

day 7 showed complete thrombosis of the false lumen from the distal aortic arch to the descending aorta. However, FDP and D-dimer levels were elevated 2 weeks after discharge. Tranexamic acid (1500 mg/d) was added; subsequently, FDP and D-dimer levels improved. The most recent laboratory data were as follows: FDP, 13 μg/mL; D-dimer, 5.9 μg/mL; fibrinogen, 393 mg/dL; plasmin-α_2-plasmin inhibitor complex, 0.5 μg/mL; and thrombin-antithrombin complex, 9.1 ng/dL. CT angiography at 3-month follow-up demonstrated a completely thrombosed false lumen from the distal aortic arch to the descending aorta at the celiac artery level. The descending aorta diameter was reduced (from 44 to 40 mm; Fig 3).

DISCUSSION

Thrombocytopenia and bleeding tendency are seen in approximately 0.5% to 6% of large aortic aneurysms,[1] but the frequency of comorbid DIC due to aortic dissection is unknown. Various treatments have been reported to be effective for chronic DIC associated with vascular disease.[1-7] In particular, direct oral anticoagulants and LMWH are effective and convenient.[1,4-7] Because of the high risk of surgical repair for bleeding tendency, conservative treatment is the first choice for aorta-related DIC. However, several studies have reported that endovascular repair is more effective than open surgery.[11,12] Yet, it was previously reported that endoleak from endovascular repair deteriorated DIC[13,14]; therefore, endovascular repair for aorta-related DIC is controversial.

In general, DIC caused by aortic dissection is often classified as enhanced fibrinolytic DIC.[2] The pathogenesis of DIC related to aortic dissection remains unclear; however, it has been reported that aortic dissection with partial thrombosed type, turbulent flow in the false lumen, and extensive false lumen can cause DIC.[11] In such cases, anticoagulant therapy should be combined with antifibrinolytic therapy. However, aorta-related DIC with anticoagulant therapy sometimes causes aortic extension and rupture.[4] Endovascular treatment can be more effective and safer than anticoagulant therapy because aortic rupture is prevented and stagnation and turbulent flow in the false lumen can be completely resolved. The safety and efficacy of TEVAR for both uncomplicated and complicated aortic dissection has been confirmed[8-10]; therefore, TEVAR can be an optimal and safe treatment of DIC associated with aortic dissection.

CSFD is generally performed because of the high risk of paraplegia, which can occur with extensive coverage of long segments of the thoracic aorta (stent graft coverage >300 mm) and compromise of important intercostal collaterals (T8-L1 spinal level).[15] However, hemorrhagic complications of CSFD can occur with anticoagulation, causing neurologic sequelae. With coagulopathy, alternative paraplegia prophylaxis (eg, perioperative hypertensive management and steroid infusion before stent graft deployment) should be selected.

In our case, the location of entry tears and turbulent flow in the false lumen could be detected by transesophageal echocardiography, and TEVAR was performed successfully without paraplegia or other complications, without CSFD. TEVAR improved the turbulent flow and reduced blood flow in the false lumen, resulting in improvement of DIC; anticoagulant therapy was

unnecessary. Endovascular therapy for refractory aorta-related DIC is a good option for aortic rupture prophylaxis and avoidance of anticoagulant therapy.

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