Persistent disseminated intravascular coagulation despite correction of endoleaks after thoracoabdominal endovascular aneurysm repair

Ann H. Kim, MD,[a] Alexander H. King, MS,[a] Alvin H. Schmaier, MD,[b] *and* Jae S. Cho, MD,[a] *Cleveland, Ohio*

ABSTRACT

Disseminated intravascular coagulation (DIC) is a rare complication of endovascular aortic repair, commonly associated with type I or type III endoleaks. DIC is also known as consumption coagulopathy because excessive thrombin formation and secondary fibrinolysis leads to consumption of coagulation factors with hyperfibrinolysis and activation of platelets, which can lead to excessive bleeding. We present the case of an 80-year-old woman who had undergone thoracic endovascular aortic repair for a type B aortic dissection that was complicated by a series of recurrent endoleak-induced DICs requiring multiple thoracic endovascular aortic repair extensions to cover the entire thoracoabdominal aorta. The DIC persisted despite the resolution of the endoleaks. (J Vasc Surg Cases Innov Tech 2021;7:730-3.)

Keywords: Disseminated intravascular coagulation; Endoleak; Thoracic endovascular aortic repair

CASE REPORT

A 77-year-old woman had initially presented to an outside emergency room because of acute onset of shortness of breath, tachycardia, and a 2-month history of worsening chronic back pain. Computed tomography (CT) angiography (CTA) revealed a type B aortic dissection with a 5-cm distal thoracic aneurysm and hemorrhagic pleural effusion. Her medical history included hypertension, hyperlipidemia, and diabetes mellitus. Her medications included metoprolol succinate (100 mg) and atorvastatin (10 mg) daily. She had a remote smoking history and denied alcohol and illicit drug use. She was transferred to our hospital and underwent thoracic endovascular aortic repair (TEVAR) on May 25, 2016 using 36 × 200-mm and 36 × 167-mm Valiant stent grafts (Medtronic, Dublin, Ireland) with a 5-cm overlap covering the aorta from zone II to the T10 vertebral body. Her fibrinogen levels were 697 mg/dL and 558 mg/dL before and after the surgery and her platelet counts were 244 $\times 10^9$/L and 230 $\times 10^9$/L, respectively.

However, 11 months later, she returned with aneurysmal degeneration of the perivisceral aorta. Renovisceral debranching (a 14 × 7-mm expanded polytetrafluoroethylene [ePTFE] graft from the left common iliac artery to the left renal artery and superior mesenteric artery and a 12 × 6-mm ePTFE graft from the right common iliac artery to the right renal artery and common hepatic artery) was performed. Two weeks later, on April 27, 2017, she underwent left common carotid to left subclavian artery bypass grafting with a 6-mm ePTFE graft and a distal TEVAR extension using 36 × 32 × 150-mm and 40 × 40 × 200-mm Valiant stent grafts with extensive overlap.

A routine surveillance CTA 1 year later in April 2018 demonstrated a type Ib endoleak. She underwent another distal TEVAR extension with 36 × 36 × 200-mm Valiant stent grafts down to the distal abdominal aorta. However, 8 months later, follow-up CTA showed both a type Ia endoleak and a type III endoleak (Fig 1). An ascending aorto–innominate and left carotid bypass grafting with proximal extension TEVAR was planned. However, during a cardiac evaluation for the planned operation, she was found to be anemic with hemoglobin of 6.5 g/dL and thrombocytopenia (74 × 10^9/L). A colonoscopy and upper gastrointestinal endoscopy were both negative for active bleeding. The hematology workup attributed the anemia to iron deficiency, and iron supplements were started.

During the next several months, she developed recurring hematoma around her buttocks and thighs. She also had persistent thrombocytopenia. Her prothrombin time and international normalized ratio were persistently abnormal, although the activated partial thromboplastin time (aPTT), liver function test results, and albumin were normal. After ruling out other etiologies, the hematology consultation concluded that she had disseminated intravascular coagulation (DIC; International Society on Thrombosis and Hemostasis score, 5) secondary to a persistent endoleak on the basis of thrombocytopenia (platelet count, 72 × 10^9/L [normal, 200-450 $\times 10^9$/L]), hypofibrinogenemia (131 mg/dL [normal, 200-375 mg/dL]), and markedly elevated D-dimer levels (20,552 ng/mL [normal, <550 ng/mL]).[1]

From the Division of Vascular Surgery and Endovascular Therapy, Department of Surgery,[a] and Division of Hematology/Oncology, Department of Medicine,[b] Case Western Reserve University School of Medicine, University Hospitals, Cleveland Medical Center.

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Correspondence: Jae S. Cho, MD, Professor, Division of Vascular Surgery and Endovascular Therapy, Case Western Reserve University School of Medicine, University Hospitals, Cleveland Medical Center, 11100 Euclid Ave, LKS 7060, Cleveland, OH 44106-7060 (e-mail: jsc163@case.edu).

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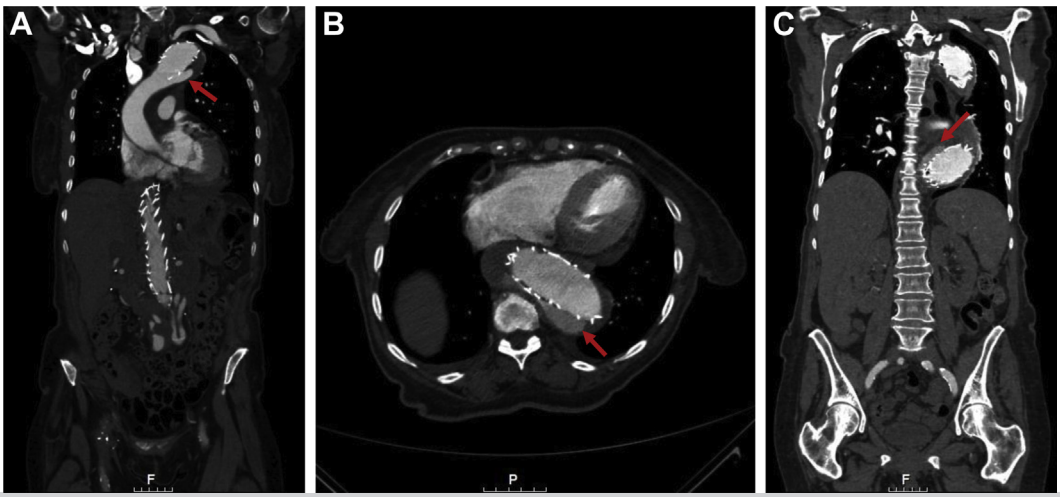

Fig 1. Computed tomography (CT) angiogram 8 months after her second thoracic endovascular aortic repair (TEVAR) extension demonstrating a type Ia endoleak at the arch **(A)** and a type III endoleak at the level of the diaphragm **(B** and **C)**, as demonstrated by *red arrows*.

After the cardiac evaluation was completed, she underwent ascending aorta to innominate and left carotid artery bypass grafting (Fig 2) and concomitant proximal TEVAR extension with a 37 × 37 × 220-mm Navion stent graft (Medtronic) in April 2019. The completion angiogram showed no evidence of an endoleak. By postoperative day (POD) 7, she had had normalization of fibrinogen (323 mg/dL) and aPTT (29 seconds), stabilization of the platelet count (121-153 $\times 10^9$/L). However, her D-dimer levels remained elevated (17,932 ng/dL). She was discharged on POD 15.

Three months later, she presented with a 3-day history of hematuria. Her D-dimer level had increased to 43,000 ng/mL and fibrinogen had decreased to 164 mg/dL. The platelet count was 87 × 10^9/L, and the aPTT was 30 seconds, indicating DIC exacerbation. CTA demonstrated a large type III endoleak (Fig 3). Her hemoglobin was 8.7 g/dL. The existing stent grafts were relined from the proximal descending to the distal abdominal segments with two 37 × 37 × 220-mm Navion stent grafts (Medtronic) on August 21, 2019, with complete resolution of the type III endoleak. Her D-dimer level had again decreased to 18,012 ng/dL, with normalization of fibrinogen, platelet count, and aPTT. She was discharged to a rehabilitation facility on POD 8 and to home 1 week later.

Nine months later, on May 20, 2020, she was noted to have a new type III endoleak with expansion of the aneurysm sac on CT (Fig 3). She underwent another session of TEVAR in July 2020 with coverage of the leaking segment using a 40-mm × 200-mm Relay stent graft (Terumo Aortic, Inchinnan, UK).

During follow-up, resolution of the previous endoleak on CTA at 3, 6, and 9 months after surgery was noted (Fig 3). No new endoleaks were visualized at any follow-up point. Her laboratory studies showed a persistently elevated D-dimer level (49,124 ng/dL), a normal fibrinogen level (255 mg/dL), and a normal platelet count (160 × 10^9/L). Chronic DIC with compensation was diagnosed. The normalization of the fibrinogen and platelets indicated that the patient's liver and bone marrow, respectively, were able to compensate for the increased consumption from the DIC. However, the basic disease process has not been ameliorated because the D-dimer level has remained persistently elevated.

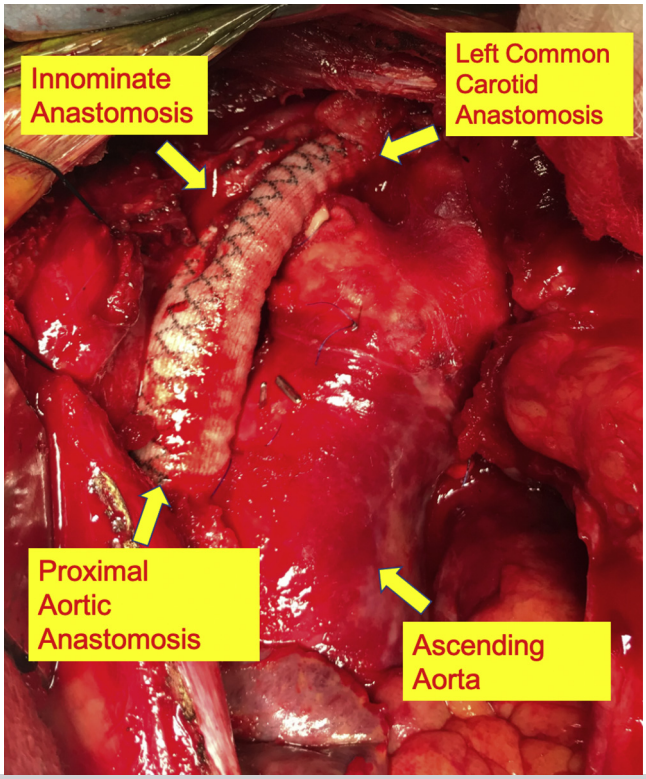

Fig 2. Our patient underwent an ascending aorta to innominate and left common carotid artery bypass, followed by a proximal thoracic endovascular aortic repair (TEVAR) extension for persistent type Ia and type III endoleaks.

The patient provided written informed consent for the report of her case details and imaging studies.

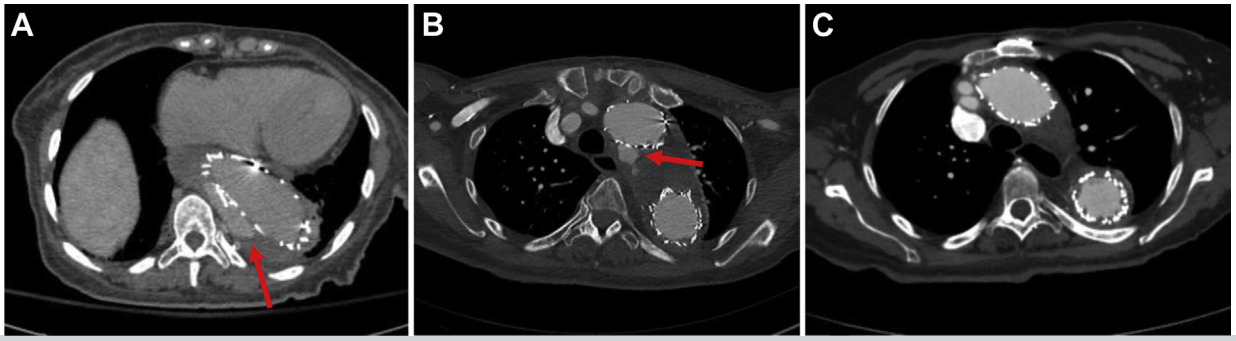

Fig 3. A, Computed tomography (CT) angiogram from August 2019 demonstrating a large type III endoleak (*arrow*) in the setting of worsening disseminated intravascular coagulation (DIC), which resolved with relining the stent grafts with Navion endografts (Medtronic, Dublin, Ireland). **B,** A new, more proximal, type III endoleak (*arrow*) had developed 9 months later with expansion of the aneurysm sac. It was treated with a Relay stent graft (Terumo Aortic, Inchinnan, UK). **C,** The most recent CT angiogram from February 2021 showing resolution of the endoleak.

DISCUSSION

We have illustrated a rare case of DIC in the setting of aortic endoleaks. In our patient's case, she continued to have compensated DIC after successful endovascular treatments, with elevated D-dimer levels but a normalized platelet count and fibrinogen level. DIC is caused by excessive and persistent thrombin and plasmin formation, resulting in the consumption of coagulation factors and platelets and hyperfibrinolysis. The shear stress of blood flowing through an area of high turbulence that occurs with type Ia and III high-flow endoleaks with formation of vascular channels within the thrombus of the false lumen or in the aneurysm sac leads to intravascular hemolysis. Expression of acidic phospholipids on the lysed red blood cell surface potentiates blood coagulation activation and secondary fibrinolysis. Additionally, the constant exposure of blood to the denuded endothelium and vascular smooth muscle tissue factor leads to activation of the coagulation cascade, with consumption of clotting factors and secondary fibrinolysis. Common laboratory indicators include thrombocytopenia, a prolonged prothrombin time, aPTT, elevated D-dimer levels, and hypofibrinogenemia.[2,3] The development of DIC secondary to an endoleak after endovascular aortic aneurysm repair has been previously described.[3-10] Thrombocytopenia was reported for all the previously described cases because it is a feature of DIC. It likely develops from local thrombin formation involved in clot formation, which also activates platelets.[4,11] However, the possibility that thrombocytopenia occurs from a separate disease state that subjects patients to a heightened risk of DIC once an endoleak has developed cannot be excluded.[4]

Although no specific guidelines exist for the medical treatment of endoleak-induced DIC, unfractionated heparin or low-molecular-weight heparin (at a reduced dose, ~25% of standard dose) has generally been used for symptom control.[2] Its use should be short term and should be considered a temporary bridge until a more definitive treatment can be provided. In patients with endoleak, previous case reports have shown that treating the endoleak will ameliorate the DIC.[4-8,12,13]

The recurrent and persistent nature of DIC in the present patient is unique. The chronic DIC in our patient was likely due to the presence of persistent type I and type III endoleaks, with high flow vascular channels leading to fragmented red blood cells and coagulation activation. Persistent DIC, even after the correction of endoleaks, is difficult to explain. It might result from an undetected endoleak or thrombus in the excluded, thrombosed false lumen undergoing hemolysis and being adsorbed into the systemic circulation, resulting in the DIC parameters. The patient has continued to be followed up closely to allow for the institution of surgical and medical correction as needed in accordance with the patient's performance status.

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