Acute-on-chronic type B aortic dissection presenting as cauda equina syndrome

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Abstract

A 60-year-old woman transferred with suspected cauda equina syndrome lacked lower extremity pulses on arrival. She rapidly developed visceral malperfusion due to underlying type B aortic dissection, necessitating aortic fenestration with thrombectomy. Despite misdiagnosis and delayed treatment, she could ambulate 1 year postoperatively. Aortic dissection remains integral to the differential diagnosis of patients presenting with acute paraparesis.

Keywords

Aortic dissection; vascular surgical procedures; delayed diagnosis.

Introduction

We present the case of a patient with an acute-on-chronic type B aortic dissection who initially presented with suspected cauda equina syndrome and developed visceral malperfusion. The patient underwent aortic fenestration with visceral and lower extremity thrombectomy and had near normal neurologic function 1 year postoperatively. This article discusses the diagnostic challenge and subsequent surgical management of complicated acute dissection of the descending aorta.

Case report

A 60-year old woman without significant past medical or family history awoke at midnight with sudden bilateral lower extremity numbness and paralysis. At a local hospital, she had absent rectal tone and non-contrast computed tomography (CT) revealed bulging L3–S1 spinal discs. She was transferred to our institution with suspected cauda equina syndrome. On her arrival, 7 h after symptom onset, she was hypertensive (157/96 mmHg), mildly tachycardic (90–100 beats/min), and had an increased white blood cell count (12.7 \times 10^3/ml). Urine output had been normal.
Otherwise, vital signs and laboratory values were unremarkable. Her symptoms did not require pain medication.

During neurosurgical work-up, she was found to have absent femoral and pedal pulses bilaterally. CT angiography of the chest, abdomen, and pelvis demonstrated a type B aortic dissection beginning in the mid-descending thoracic aorta and extending to the right common femoral artery and the left superficial femoral artery. The false lumen was thrombosed (Fig. 1). The left kidney was non-enhancing. The celiac trunk, superior mesenteric artery (SMA), and two right renal arteries were perfused from the true lumen, albeit through small channels of thrombus. The patient was anticoagulated with intravenous heparin and emergent lower extremity revascularization with extra-anatomic bypass was planned. In the pre-operative area, she developed abdominal pain, nausea, hematochezia, and coffee grounds emesis. Due to concern for acute visceral malperfusion, the operative strategy was revised to aortic fenestration with visceral and lower extremity thrombectomy.

The patient was positioned in a right lateral decubitus position. A left thoracoabdominal incision was made. The descending thoracic aorta was dissected out and controlled with clamps at the diaphragm and aortic bifurcation. The celiac trunk, SMA, and left renal artery were controlled with vessel loops. A longitudinal aortotomy was performed from just above the celiac artery to below the left renal artery. Acute- and chronic-appearing thrombi were extracted, and a chronic-appearing, fibrotic septum was identified. Balloon thrombectomies of the celiac trunk, SMA, and right renal artery were performed and unorganized thrombi were returned with good back-bleeding (Fig. 2). Balloon thrombectomy of the left renal artery was attempted, but no back-bleeding was obtained. Final pathology analysis of the specimens revealed bland, laminated, unorganized thrombus. Normal Doppler signals were restored in the celiac, superior mesenteric, and right renal arteries. Although the bowel was initially dusky, it appeared fully viable at the time of abdominal closure. Femoral arterial pulses were palpable bilaterally. Prophylactic 4-compartment calf fasciotomies were performed bilaterally.

The patient was admitted to the surgical intensive care unit (ICU) for postoperative management. Several hours postoperatively, her lactate level was increased (4.4 mmol/l), but decreased to
2.7 mmol/l within 12 h. Adequate urine output was maintained. Her nitroglycerine drip was weaned. She was extubated on postoperative day 2. Postoperatively, magnetic resonance imaging of the spine confirmed spinal cord infarction at T11–T12 and T12–L1. The patient was discharged to a rehabilitation facility on long-term anticoagulation with warfarin. One year postoperatively, she wears a brace for a foot drop, but ambulates without crutches.

Discussion

Impact of late diagnosis on morbidity and mortality

Acute type B aortic dissection (ABAD) is a rare disease, with an incidence of 5–7 cases per 1,000,000 person-years[1,2]. However, ABAD is a severe disease process that carries an in-hospital mortality rate of 11% overall and up to 71% in the highest risk patients, necessitating emergent diagnosis and management[3–5]. Unlike type A dissection, in which mortality arises from rupture and other cardiac complications, the morbidity of type B dissection lies in end-organ compromise of the viscera and extremities[6].

Female patients with dissection fare worse than their male counterparts. Significantly older age at presentation partly contributes to the gender discrepancy, as 56.4% of females versus 36.3% of males with ABAD present at age 70 years or older[7]. However, female patients with ABAD are also less likely to present with common symptoms of chest or back pain, causing them to present an average of 5 h later in the clinical course than men[2,8].

Atypical ABAD

The increased mortality associated with delayed diagnosis requires that clinicians be knowledgeable of the classic and atypical signs of dissection. Aortic dissection typically presents with the acute onset of severe, sharp chest or back pain. Our patient was female, had no chest or back pain, and had lower extremity paralysis without femoral pulses. This atypical presentation contributed to delayed diagnosis of ABAD. In a case series of 175 patients with ABAD, abrupt onset of pain (83.8%), most often in the back (63.8%) or chest (62.9%), was the most common sign[2]. This implies, however, that many patients presented without the classic signs of ABAD. Other less commonly recognized but critical signs of ABAD include systolic blood pressure greater than 150 (70.1% of patients), abdominal pain (42.7%), aortic insufficiency murmur (12.0%), pulse deficits (9.2%), and syncope (4.1%)[2]. In the case of our patient, earlier recognition of pulse deficits and hypertension would have prompted earlier treatment and possibly improved the outcome.

Emergent operative management of aortic dissection

ABAD has minimal risk of rupture and, among stable patients, medical treatment is preferred over surgery[9]. The exception to this rule occurs in the case of complicated type B dissection,
which is defined by the presence of renal, visceral, or limb malperfusion; refractory hypertension; or severe pain. In our patient, the development of bloody emesis and hematochezia heralded mesenteric ischemia, which necessitated a revision of the operative plan from axillary-femoral bypass to open aortic fenestration with visceral and lower extremity thrombectomy. 

Approximately 80% of cases of complicated ABAD are characterized by dynamic obstruction, in which flow is reduced by either prolapse of the dissection flap into a branch vessel lumen or compression of the true lumen. The treatment of choice is to equalize flow states between the true and false lumens via aortic fenestration, in which the dissection flap is extensively resected to restore true-to-false lumen communication. The remaining cases of static obstruction are characterized by sustained absent visceral arterial flow and warrant an extended septectomy. The goal of treatment is to restore true-to-false lumen communication.

The primary option for intervention with complicated ABAD remains aortic fenestration. However, improvements in the percutaneous approach have created a viable alternative to open surgery. Thoracic endovascular aortic repair has become a first-line intervention for rapid and minimally invasive treatment of complicated ABAD. Early results from non-randomized retrospective studies with endovascular therapy for acute aortic dissection have been promising, although robust long-term outcome data have not yet been documented. In the present case, given evidence of visceral ischemia and suspected thrombosis of the visceral branches, we felt open aortic fenestration with visceral thrombectomy was most appropriate. This approach also allowed visual assessment of bowel viability. In our case, a cerebrospinal fluid drain was not used due to the emergent nature of the operation.

After surgical intervention, ICU care remains an integral part of the recovery process for patients with ABAD to control hypertension and to assess for postoperative complications including visceral organ malperfusion. Postoperative hypertension afflicts 4–35% of patients, with the higher estimates attributed to patients requiring clamping of the aorta. One case report documents true lumen stenosis after surgical repair of a type A aortic dissection due to uncontrolled hypertension postoperatively. In the present case report, the patient was placed on a nitroglycerin drip to preclude such a hypertensive crisis. She was also monitored for decreased urine output, which remained normal.

**Teaching point**

Given the significant morbidity and mortality associated with ABAD, there is a need for increased awareness of its myriad presentations. Despite appropriate operative and postoperative ICU management, earlier diagnosis may have minimized the patient’s residual disabilities.

**Conflict of interest**

None.

**References**


Acute-on-chronic type B aortic dissection and cauda equina syndrome