Visceral Ischemia, Aortic Expansion, and Ehlers-Danlos Syndrome

This dangerous and complicated combination required a multifaceted solution.

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Controversy exists regarding the optimal treatment for patients with acute type B aortic dissection. Given the lack of well-controlled prospective trials, physicians often find themselves forced to make decisions in the absence of scientific data. However, the severity and acuity of the problem, coupled with the extensive morbidity and mortality associated with conventional repair for aortic dissection, has forced many clinicians to employ investigational approaches to this disease, resulting in several reports pertaining to the treatment of such problems with endovascular prostheses. Unfortunately, the definition of a successful procedure also remains the subject of considerable speculation.

Figure 1. Maximum image projection (MIP) reconstruction of the preoperative contrast CT scan. The distal dissection has a pronounced false lumen with marked true lumen compression. The celiac, superior mesenteric, and left renal arteries are supplied off the true lumen, and the resulting ischemia is readily apparent on this image with an absent left nephrogram (red arrow).

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Purists may argue that successful graft deployment, coupled with lack of aneurysm growth or conversion would constitute success; however, many seasoned clinicians have argued patient survival until hospital discharge should be the ultimate goal. Given the historical mortality approaching 80% in patients presenting with renal or visceral ischemia, the latter goal seems admirable. The case presented in this report illustrates the thought process accompanying a complex dissection associated with visceral and renal ischemia in a 28-year-old woman with Ehlers-Danlos syndrome.

PATIENT HISTORY

This patient was transferred to our institution 7 days after repair of a 6.5-cm infrarenal aneurysm. A polyester bifurcated graft had been anastomosed to the infrarenal aorta and both common iliac arteries. The patient was discharged shortly after her aneurysm repair but was readmitted with back and abdominal

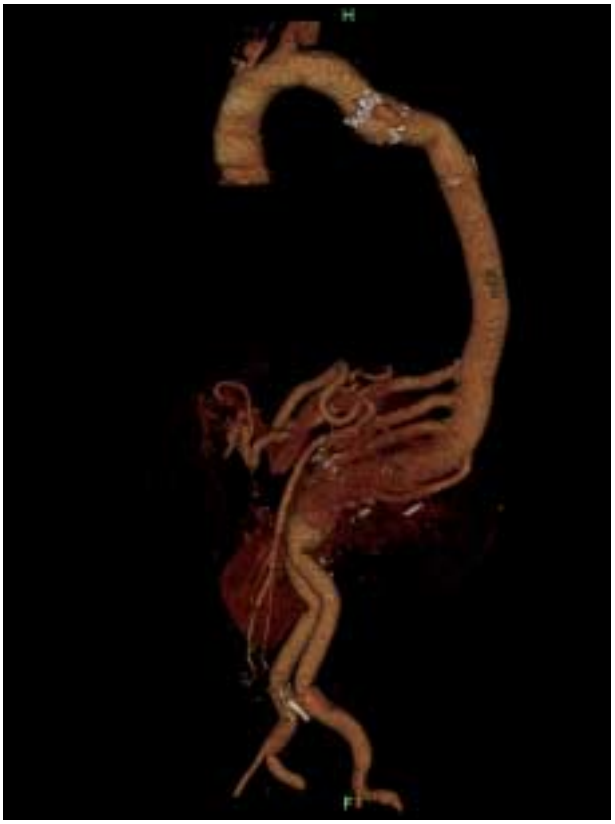

Figure 6. 3D reconstruction of the contrast CT scan performed after open thoracoabdominal repair. The celiac, superior mesenteric artery, and both renal arteries have been reimplanted and the proximal anastomosis of the thoracic graft has been supported with a 26-mm X 33-mm Excluder stent graft. There is excellent flow in all visceral segments.

The patient was initially treated in the intensive care unit for medical management of high blood pressure. Her mesenteric symptoms resolved, and her creatinine normalized. She was reimaged approximately 2 weeks after the initial intervention. CT scan demonstrated all visceral flow arising from the true lumen, improved left renal perfusion, but persistent false lumen flow with a markedly increased aortic diameter (from 34 mm before endovascular intervention to 42 mm after intervention) (Figure 5). The endovascular procedure had ameliorated her acute ischemic symptoms, but failed to entirely eliminate false lumen flow and consequently failed to protect the aorta from rapid dilatation.

The patient was returned to the operating room for an open operation to surgically replace her aorta from immediately distal to the left subclavian artery to the previously placed infrarenal graft. Separate branches to each of her visceral arteries were constructed. Hemostasis at the proximal anastomosis was imperfectly achieved, and an additional stent graft was placed under direct vision within the anastomosed segment. The patient then had an uneventful recovery without neurological sequelae. A final CT scan with contrast prior to the patient's discharge again demonstrated excellent flow in all visceral vessels with both kidneys well perfused (Figure 6).

DISCUSSION

Distal aortic dissections result in an annual mortality rate in excess of that reported for ruptured aneurysms. Death is a result of end-organ ischemia in up to 80% of all cases.[1-6] Aortic rupture is less common in the acute setting but still occurs in up to 20% of patients during their life span. The indications for intervention have included rupture, intractable pain or hypertension, end-organ ischemia, and degeneration of the aortic wall causing aneurysm formation. Although a great deal of progress has been made since the inception of the management of aortic dissection clinical sequelae in 1935, contemporary reports still harbor exceptionally high mortality rates in the subset of patients that experience ischemic complications (16%-25%).[1,4,5,7]

The development of an ischemic syndrome is the most frequently encountered morbidity and requires rapid treatment to prevent mortality. In our institution, it is also the most frequent indication for intervention in the setting of an acute dissection. Unfortunately, conventional open surgical options in these circumstances have been associated with mortality rates of up to 80%. Newer endovascular methods including percutaneous fenestration,[4,8] visceral vessel stenting from the true lumen, and placement of endovascular stent grafts over the major fenestration,[9-12] have been noted to markedly diminish the mortality rate for this condition, but lack long-term results and have generally been reported in series with small numbers of patients. However, an in-depth knowledge of medical therapies, open repair, and endovascular interventional options are prerequisites to optimal patient care. Knowing when to apply a particular treatment, being able to define the success of that treatment in the overall context of patient management, and knowing when to combine multiple techniques will help us to treat these patients better and reduce the associated morbidity and mortality.[6]

SUMMARY

This article illustrates complications occurring in a patient after an open infrarenal aneurysm repair, complications leading to marked mesenteric and renal ischemia, complications after endovascular manage-

ment of this problem, complications encountered during the open surgical conversion, and ultimately a successful outcome. One could easily argue that a failure was present at each stage of therapy, yet each therapeutic endeavor successfully achieved the intended objective. In other words, the infrarenal aneurysm was repaired yet, resulted in a thoracic dissection. The dissection was managed medically until an interventional procedure was performed. The interventional procedure was successful at treating the ischemia but did not prevent aortic growth. The surgery, assisted by endovascular techniques, treated the aortic growth resulting in a good outcome. This would not likely have been attained if performed in the setting of acute ischemia. ■

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