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Acute multi-visceral thrombosis and ischemia in a 3-year-old child

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ABSTRACT

Acute multi-visceral ischemia secondary to multi-vessel thrombosis is rare in the pediatric population and to our knowledge it has not been previously reported. We present a 3-year old boy with acute thrombosis of the celiac trunk, superior mesenteric artery, and left renal artery in addition to stenoses of his intracranial carotid arteries. The patient presented with extensive small and large bowel ischemia and perforations, gastric perforation, gallbladder necrosis, acute kidney injury and stroke. Here we describe our medical, surgical and percutaneous interventions for the successful treatment of this child.

1. Introduction

Visceral and renal artery stenoses are commonly associated with midaortic syndrome (MAS), however, they rarely result in acute thrombosis or ischemic necrosis of end organs. Even inflammatory vasculopathies are not commonly associated with acute thrombosis. Acute mesenteric ischemia (AMI) in the adult population has an incidence of 4.3–10/10,000 and is associated with a high mortality rate [1]. AMI in the pediatric population has been described in the literature, though due to its rarity, the precise incidence is unknown. To our knowledge, acute multi-visceral ischemia secondary to multiple large vessel arterial thrombosis has not been previously reported. Here we present the successful management of a 3-year-old previously healthy male who presented with celiac trunk, superior mesenteric artery (SMA), and left renal artery thrombosis with stenoses of intracranial carotid arteries.

2. Case report

2.1. Initial presentation and course at outside institution

A 3-year-old previously healthy male presented to a local emergency department with abdominal pain, bilious vomiting, and blood pressure of 200/100 mmHg. A few weeks prior, the patient had Streptococcal pharyngitis and vague abdominal pain with decreased oral intake. Further evaluation included an echocardiogram that revealed left ventricular hypertrophy and a CT angiogram that demonstrated an asymmetric small left kidney with left renal artery thrombosis, right renal artery stenosis, diffuse narrowing of the abdominal aorta, and occlusion of the origin of the celiac trunk and the SMA (Fig. 1a and b). Five days after presentation, the patient underwent surgical exploration which revealed a bowel perforation requiring resection of 10–15 cm of ileum, and creation of ileostomy and mucous fistula. He simultaneously developed altered mental status and was found to have posterior reversible encephalopathy syndrome (PRES), but cerebral angiogram did not demonstrate any abnormalities of the major cerebral vasculature. At that point, the working diagnosis was Takayasu's arteritis. He was initiated on steroids and infliximab, and his antihypertensive regime consisted of intravenous nitroprusside and nicardipine. On postoperative day (POD) 4, the patient was transferred to our institution.

2.2. Initial hospital course at our institution

Upon arrival to our institution, the patient was already intubated and sedated. His systolic blood pressure was maintained around 160–170 mmHg in order to maximize collateral perfusion of his abdominal viscera given thrombosis of the celiac artery and SMA. His abdomen was distended and both the ileostomy and mucous fistula were obviously necrotic. The patient was hemodynamically stable and we decided to first proceed with an attempt to re-establish flow through the mesenteric and renal vasculature via a percutaneous endovascular

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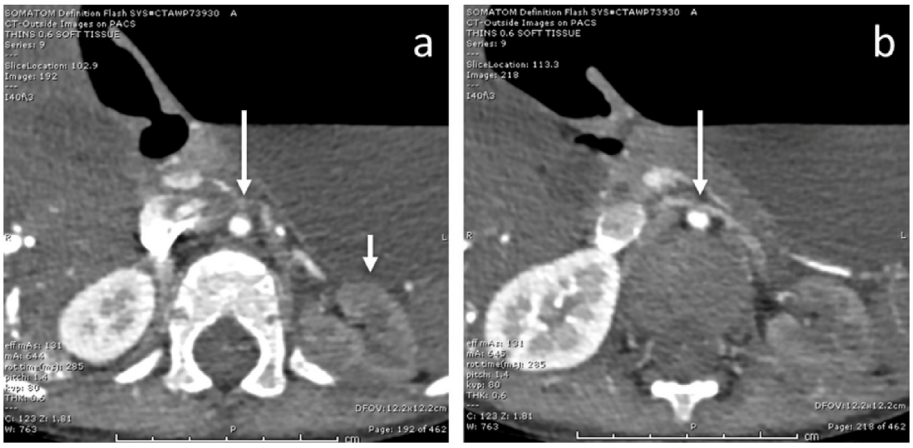

Fig. 1. Axial sections of CT angiogram of the abdomen demonstrating (a) occlusion of the origin of the celiac artery (long arrow) and poor perfusion to the left kidney (short arrow) and (b) occlusion of the origin of SMA (arrow).

approach. The patient underwent aortography and balloon dilation of his SMA, celiac artery, inferior mesenteric artery (IMA) and left renal artery, along with local tPA administration in the left renal artery, with successful establishment of patency of these arteries. On hospital day #2, he underwent abdominal exploration where he was found to have various stages of ischemia necrosis involving the entire small bowel for which he underwent several small bowel resections and suture reinforcement of other areas of questionable viability. In addition, cholecystectomy was performed for necrosis of the gallbladder and primary repair of a gastric perforation along the lesser curve was performed. Intraoperative Doppler examination demonstrated weak arterial signals in the small bowel mesentery and a weakly pulsatile SMA signal. Given these findings and in order to maximize the chance of survival of the remaining small intestine, we proceeded with revascularization of the SMA. The IMA was transected several centimeters distal to its takeoff from the aorta and an end-to-side anastomosis was performed between the proximal segment of the IMA and the SMA. The distal IMA was reimplanted onto the left common iliac artery.

Following the initial operation, the patient underwent 3 additional planned abdominal re-explorations resulting in: 1. Multiple small bowel resections; 2. Management of two duodenal perforations (second and fourth portions) with tube duodenostomy; 3. Creation of a new end-ileostomy for decompression of discontinuous small bowel; and 4. Right colectomy. Eventually, 20 days after the initial operation at our institution, the patient underwent fascial closure and closure of the subcutaneous tissue by secondary intention using wound vacuum-assisted closure device. During this period the patient was maintained on systemic anticoagulation using heparin infusion and aspirin *per rectum*.

Once paralysis was lifted following final closure of the abdomen, the patient was noted to lack extremity movements. MRA of his brain revealed new ischemic infarction in his right frontal lobe and increase of the steno-occlusive vasculopathy in the circle of Willis, most notably involving the bilateral supraclinoid internal carotid arteries. In addition, extensive microhemorrhages were present and anticoagulation was replaced with aspirin alone. Gradually, the patient had improvement in his neurologic function and after two months he was extubated.

On hospital day #87, a repeat CTA showed severe narrowing of the SMA just beyond its ostium, mild narrowing of the celiac artery origin (Fig. 2a) and occlusion of the IMA, and patent but beaded and irregular renal arteries (Fig. 2b). Consequently, the patient underwent aortic angiography with celiac and bilateral renal artery balloon angioplasty. The IMA-to-SMA bypass graft was noted to be patent.

The patient was eventually discharged from the hospital after three months on amlodipine and labetalol as well as home total parenteral nutrition (TPN). The patient's abdominal anatomy included a blind-ending 4th portion of the duodenum with two duodenostomy tubes to gravity drainage, a de-functionalized segment of small bowel ending in an ileostomy, and an intact left colon as a Hartman's pouch. With regards to his renal function, the patient developed acute kidney injury (AKI) during his hospitalization reaching a peak serum creatinine level of 2.3 mg/dl, which returned to a normal level at the time of his discharge from the hospital.

Nine weeks after discharge, the patient underwent a surveillance aortic angiography which revealed a stenotic celiac trunk and occluded SMA and IMA. Given these findings, he underwent balloon dilatation of the celiac trunk and placement of a 3.6 × 16 mm stent.

The patient was subsequently maintained on home TPN and allowed to take food by mouth for comfort. After 3.5 months following discharge, the patient returned for re-establishment of gastrointestinal continuity. Since the perfusion to the gastrointestinal tract was only through the stented celiac trunk, we decided to improve the blood flow to his intestines by performing a bypass prior to performing any bowel anastomosis. Therefore, the patient underwent an infrarenal aorta to common hepatic artery bypass with a composite right saphenous and right gonadal vein grafts (Fig. 3) as well as removal of one of the duodenostomy tubes and repair of the 2nd portion of the duodenum. One week later the patient underwent re-establishment of complete intestinal continuity by performing duodenojejunostomy and jejunocolostomy. At this point, the patient had 40 cm of small bowel remaining. The patient had an uneventful postoperative course and was discharged from the hospital 2 weeks later.

2.3. Outpatient follow-up

The patient was eventually transitioned to full enteral feeds over the subsequent several months. However, 6 months later he had several bouts of pancreatitis which responded to medical management including TPN. Imaging demonstrated near complete occlusion of the celiac stent and thrombosis of the aortic to hepatic artery bypass graft without any evidence of ischemia to the viscera. Of note, he had been maintained on daily aspirin following arterial bypass surgery. The patient is now 2.5 years from initial presentation and is tolerating oral intake and gastrostomy feeds well and is being actively weaned off TPN. He has had a complete neurological recovery and is attending kindergarten.

3. Discussion

Our patient presented with an extremely challenging set of problems arising from acute thrombosis resulting in ischemia of multiple organs. A multidisciplinary team approach was absolutely necessary to care for this patient in order to maximize the viability of the abdominal