Journal of Pediatric Surgery Case Reports 38 (2018) 27–30

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

Idiopathic abdominal aortic dissection causing aortoiliac occlusive disease in a teenage boy

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ARTICLE INFO

Keywords:
Pediatric
Abdominal aortic dissection
Peripheral vascular disease

ABSTRACT

Abdominal aortic dissection (AAD) is very rare in the pediatric population in the absence of trauma, connective tissue disease, or infection. Our patient presented with an AAD causing aortoiliac occlusive disease (AIOD) and left popliteal arterial disease which is not described in the pediatric population. Presentation of the AIOD was classic in our pediatric patient though a distracting history of strained calf muscle during a sporting event delayed diagnosis by several months. Once the diagnosis was established, the interesting problem of managing this disease was met with possible risks of other vascular disease or even other organ disease. Further imaging and a genetic evaluation did not elucidate an etiology for our patient, only a suspicion. Post-surgical intervention was met with symptom resolution but warranted careful surveillance and follow-up in clinic by both genetics and vascular surgery.

1. Introduction

Idiopathic abdominal aortic dissection (AAD) of the pediatric patient has not been previously described in the literature. Aortoiliac occlusive disease (AIOD) in the setting of AAD makes this case of greater interest. Aortic disease is typically seen in the thoracic aorta, in cases of congenital heart disease, connective tissue disorders, or significant trauma, and with high mortality [1–4]. Our institution reports a pediatric case of AIOD secondary to AAD with left popliteal arterial disease of no apparent etiology.

2. Case report

A teenage boy of no significant neonatal/medical history underwent orthopedic evaluation after sustaining right calf pain attributed to a strained muscle during a sporting event in 2016. The clinic visit resulted in a diagnosis of gastrocnemius muscle strain and a referral to outpatient physical therapy (PT). The physical therapist identified pain with walking, running, and sharp pains at night that would awaken the patient. The patient underwent biweekly sessions with the physical therapist and adhered to a home exercise regimen. After 4 months of consistent PT, the physical therapist noted little improvement in the patient's pain with exercise and encouraged the family to see the patient's pediatrician. The pediatrician ordered an arterial duplex ultrasound of the bilateral lower extremities. The results of this study were remarkable for:

1. The mid left superficial femoral artery (SFA) becoming diminutive with no identified flow but a large number of collateral vessels that reconstituted the left SFA distally.
2. The mid right SFA being more diminutive in its caliber than the distal portion of the artery.

The patient was referred to an outside vascular surgeon with work up of CT angiogram chest, abdomen, and pelvis that identified an infrarenal 6.3 mm focus of dissection extending into the bilateral common iliac arteries, right greater than left (Fig. 1). The left SFA was diminutive proximally and reconstituted by collaterals distally. The left popliteal artery was occluded though the left anterior tibial artery was patent (Fig. 2). The patient was then referred to our vascular surgery department due to the complexity of the case. The plan was made to undergo genetic testing to rule out possible collagen vascular disorders. The genetics team began their evaluation and did not establish familial traits in a family pedigree that identified a connective tissue disorder. The plan was to undergo Thoracic Aortic Aneurysm and Dissection (TAAD) panel which would be followed up postoperatively.

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https://doi.org/10.1016/j.epsc.2018.07.031
Received 26 July 2018; Accepted 31 July 2018
Available online 06 August 2018

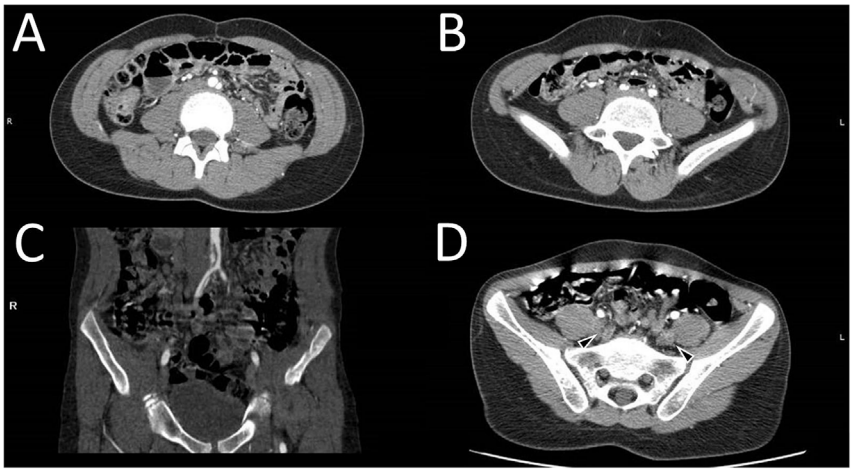

Fig. 1. Preoperative CT of the Abdomen and Pelvis Demonstrating Aortic Dissection Extending into the Bilateral Iliac Arteries. A-D is computed tomography with intravenous contrast of the abdomen and pelvis: A) axial section of the abdominal dissection just before the bifurcation B) axial and C) coronal section of abdominal aortic dissection extending into the bilateral common iliac arteries D) absence of internal iliac arteries (black arrows).

Fig. 2. Preoperative CT scan 3D Reconstruction of Runoff Vessels Demonstrating Left Popliteal Occlusion with Well-Developed Collaterals. Computed tomography of a 3D reconstruction of bilateral lower extremities. Left popliteal artery is absent, but collateral vessels are present (white arrows).

The patient underwent an open aortoiliac bypass with 12 × 6 cm bifurcated Dacron graft. Briefly, a midline excision was made. Bilateral distal external iliac arteries were exposed. The aorta was exposed just inferior to the renal arteries to the level of the inferior mesenteric artery. The aorta was palpated and noted to be firm and inflamed. The patient was heparinized and proximal and distal control was achieved. The 12 × 6 cm bifurcated Dacron graft was anastomosed end to side via a longitudinal arteriotomy in the aorta and bilateral external iliac arteries. Some give in the graft was allowed to accommodate for growth. Distal pulses were biphasic on handheld doppler. The patient was extubated and transferred to the pediatric intensive care unit (PICU) for continuous hemodynamic monitoring and hourly neurovascular checks. The patient was also started on aspirin and completed a 24hr dose of prophylactic cefazolin.

The entirety of the patient's hospital stay was uneventful. The patient was transferred out of the PICU on POD#2 tolerating a regular diet, without requiring any invasive vascular access or catheters, and began ambulating with physical therapy. By POD#4, the patient had return of distal palpable pulses, and by POD#5, he was deemed safe for discharge home with outpatient physical therapy, aspirin 81 mg daily, and follow-up with vascular surgery and genetics.

Mid-month, the patient was seen in vascular clinic and was progressing with PT, had improved leg pain, and continued to have strong femoral pulses (3+, not present preoperatively) palpable pedal pulses (1+). Post-operative imaging was scheduled for 3 months after genetics follow-up. The genetics team was suspicious of Ehlers Danlos Type IV even though the TAAD panel was negative. They planned for chest/abdomen/pelvis MRI every 6 months and recommended that all family members undergo MRI or CT of chest/abdomen/pelvis as well. Genetics clinic follow up was scheduled for 12 months. Several weeks after the genetics visit, the post-operative imaging, a CT angiogram of the abdomen/pelvis, was completed noting a widely patent anastomosis