Cryptogenic acute lower extremities and multiorgan ischemia in an 8-year-old girl

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ABSTRACT

Pediatric acute limb ischemia is rare but can have devastating effects on children. The most common causes of acute limb ischemia in the pediatric age group are iatrogenic injury during cardiac catheterization and traumatic vessel injury. Embolic events have been described less often. We present the case of an 8-year-old girl with cryptogenic extensive bilateral lower extremity ischemia and embolization to multiple visceral organs. Our findings have highlighted the importance of interdisciplinary workup, timely intervention, and the advantage of intraoperative imaging for revascularization. (J Vasc Surg Cases Innov Tech 2022;8:565-8.)

Keywords: Acute limb ischemia; Pediatric; Surgical embolectomy; Thromboembolism

Acute limb ischemia (ALI) is an emergent condition that requires immediate vascular management.[1] Although substantial data are available regarding the management and outcomes of ALI in the adult population, ALI in pediatric patients is very rare and requires additional considerations, including differences in the pharmacologic responses to anticoagulation therapy and the long-term sequelae of thrombosis.[2] Although ALI in children will most often result from iatrogenic or traumatic causes, in a subset of pediatric patients with ALI, the cause of ALI will never be established. These patients will often be treated with anticoagulation therapy.[2] The so-called cryptogenic emboli have seldom been described in the literature overall, even in adults.[3] In adults with cryptogenic emboli, recurrence rates have approached 25%, and lifelong anticoagulation therapy will often be warranted.[3] We present the case of an otherwise healthy 8-year-old girl with profound bilateral lower extremity ALI and concomitant visceral embolization treated with emergent revascularization. The patient's parents provided written informed consent for the report of her case details and imaging studies.

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CASE REPORT

An 8-year-old girl with no prior medical or surgical history had been transferred to the pediatric intensive care unit at our institution for vascular surgery and pediatric surgery evaluations from an outside hospital emergency department after presenting with acute-onset bilateral lower extremity pain and difficulty walking for 1 day. She had also reported abdominal discomfort 1 day before her presentation. The physical examination was notable for pale and cool feet bilaterally, with mild mottling below the knees and decreased motor and sensory neurologic function. No pedal Doppler signals were detected on the physical examination. No abdominal tenderness or peritoneal signs were present. The laboratory test results were significant for elevated creatinine kinase to 14,305 U/L, white blood cell count of 12,000/μL, and normal creatinine and lactate levels. Her C-reactive protein was 4.71 mg/dL. A full preliminary rheumatology workup was conducted, with unrevealing findings. A SARS-CoV-2 (severe acute respiratory syndrome coronavirus 2) test was negative. The initial imaging studies at the outside hospital had included a venous duplex ultrasound with no acute findings, and an arterial duplex ultrasound, which showed thrombosis of the bilateral superficial femoral arteries. The parents reported no family history of hypercoagulable disorders.

Computed tomography angiography of chest, abdomen, pelvis, and bilateral lower extremities showed bilateral renal infarcts, a small superior mesenteric artery embolus with distal reconstitution, without any signs of bowel ischemia, and extensive bilateral lower extremity emboli (Fig 1). The left external iliac artery (EIA) and common femoral artery (CFA) were occluded, with distal reconstitution and single vessel runoff. The right EIA was patent; however, the distal EIA and CFA were occluded, with reconstitution of the superficial femoral artery and profunda femoral artery, with no filling of the distal runoff (Fig 1).

The patient had been transferred with intravenous heparin. Because the patient did not have ongoing abdominal pain and no evidence of bowel compromise was found by computed tomography, we decided to treat the visceral emboli with

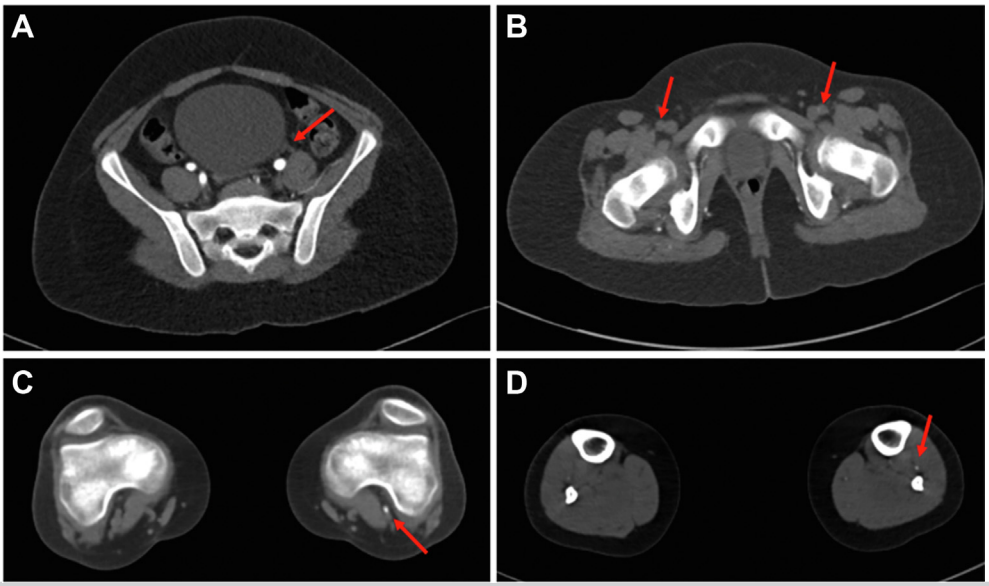

Fig 1. Computed tomography angiograms demonstrating complete occlusion of the left external iliac artery (EIA; **A**; *arrow*), bilateral occlusion of the common femoral arteries (CFAs; **B**; *arrows*). **C,** The popliteal artery in the right leg was completely occluded and minimally patent in the left leg. **D,** In the right leg, no filling was present in the tibioperoneal trunk, anterior tibial artery, posterior tibial artery, or peroneal artery, with no runoff branches. In the left leg, the anterior tibial artery was minimally patent; however, the tibioperoneal trunk and all other branches were occluded.

anticoagulation therapy and proceed with emergent bilateral lower extremity revascularization in a hybrid operating room. The bilateral CFAs were exposed, and bilateral iliofemoral and femoropopliteal embolectomies were performed using no. 2 and 3 Fogarty catheters. After extraction of a large volume of thrombus, 2 mg of tissue plasminogen activator was instilled in each leg distally to treat any residual thrombus. Next, bilateral four-compartment fasciotomies were performed, and the muscles appeared viable. A completion angiogram was performed, which showed excellent clearance of all thrombus and patent bilateral two-vessel runoff to the feet (Fig 2).

She had an uncomplicated postoperative course and underwent closure of the left and right fasciotomy wounds on postoperative days 3 and 7, respectively. She was evaluated by the hematology, rheumatology, and cardiology services. Transesophageal echocardiography did not show evidence of intracardiac thrombus or endocarditis but demonstrated a very small patent foramen ovale. It was thought that this patent foramen ovale was too small to have contributed to her extensive emboli, and venous source was found on the imaging studies. Pathologic examination of the emboli was nonrevealing. The findings from additional imaging studies, including brain magnetic imaging and upper extremity duplex ultrasound, were negative. Further rheumatologic workup was only notable for a heterozygous plasminogen activator inhibitor 1 mutation, which is associated with venous thrombosis. However, no venous source was ever found. She was discharged on postoperative day 13 with a 6-month course of 30 mg enoxaparin twice daily. The hypercoagulable evaluation findings were negative. At the 1-month follow-up in the vascular surgery clinic, the patient had 2+ palpable pedal pulses bilaterally and was rehabilitating well. At 6 months of follow-up, she demonstrated bilateral lower extremity patent vasculature, except for a short 50% asymptomatic focal stenosis of the proximal right superficial femoral artery. The superior mesenteric artery and bilateral renal arteries were patent, with resolution of previously seen bilateral renal infarcts and mild cortical scarring. At 8 months of follow-up, she was ambulating well without pain, and ultrasound showed patent vessels with stable stenosis of the right superficial femoral artery. At 1 year, she continued without activity limitations, with anticoagulation therapy continued indefinitely as recommended by the hematology team.

DISCUSSION

Our case report highlights a very rare vascular emergency in a pediatric patient with emboli to multiple organs and the importance of imaging studies before and after surgical intervention to ensure optimal revascularization. In the adult population, the mortality and limb loss associated with ALI can be as high as 40% and 50%, respectively, despite surgical treatment.[4] Although the reported rates of mortality and limb loss have been <10% in the pediatric population, early recognition is crucial.[5] Additionally, given the rarity of ALI in pediatric patients, the ALI might not be diagnosed in a timely manner, resulting in delayed treatment. In a limited case series of children aged <13 years, Dalsing et al[6] suggested that children with obvious signs of ischemia should receive emergent surgical intervention. Emergent intervention is especially important when the cause is

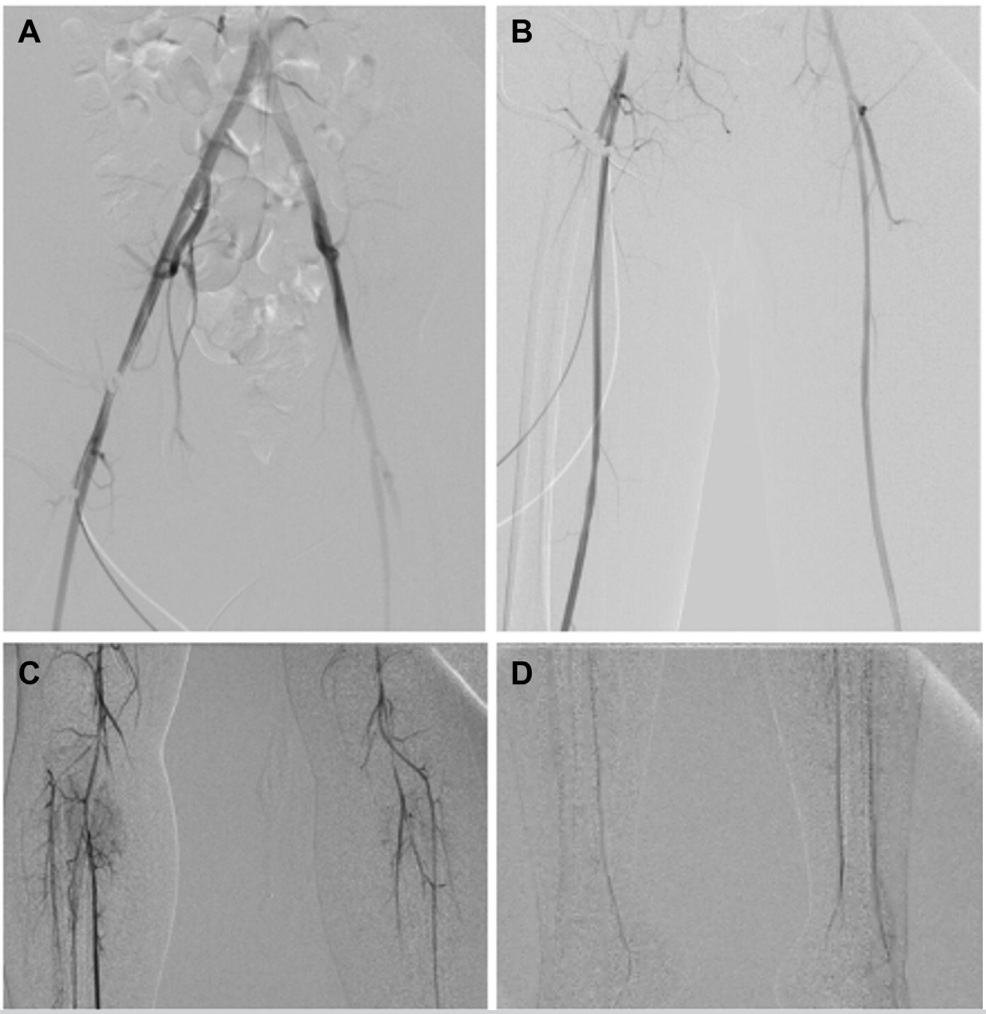

Fig 2. Arteriograms after embolectomy demonstrating restoration of flow bilaterally, with reconstitution in the iliac arteries, bilateral common femoral arteries (CFAs), profunda femoral arteries, superficial femoral arteries **(A,B)**, bilateral popliteal arteries, tibioperoneal trunks, and branches **(C)**. **D,** The anterior tibial arteries and posterior tibial arteries were also restored with satisfactory runoff.

unknown (eg, not attributable to an iatrogenic injury).[6] Multiple studies have demonstrated low public awareness among the adult population regarding ALI despite the high rates of peripheral arterial disease,[7] which often leads to delayed presentation. Awareness of ALI is, thus, likely even lower for the pediatric population, for whom atherosclerosis-associated ALI is almost unheard of.[8] For the present patient, the quick recognition and multidisciplinary expertise led to timely revascularization, preventing limb loss.[9,10] The treatment of pediatric ALI requires consideration of additional factors, including the technical challenges resulting from the smaller vessel size and the risks of anticoagulation therapy.[2,5]

Given the absence of atherosclerosis or evidence of arterial injury, the etiology of ALI for our patient remains unknown. The pattern of distribution of thrombus in the lower extremities and in the visceral vessels suggests an embolic process. In adults, thrombosis and embolism account for most ALI cases (24% and 46%, respectively); however, embolism leading to ALI has rarely been observed among pediatric patients.[11] Most pediatric ALI cases will result from iatrogenic injury during cardiac catheterization or trauma.[2,5,6,11] In pediatric patients, ALI due to embolism has been previously attributed to cardiac sources, such as atrial myxoma.[12] In the absence of any obvious trauma or cardiac risk factors, it can be more difficult to diagnose ALI in this population. Transesophageal echocardiography in our patient did not show evidence of intracardiac thrombus or endocarditis; hence, the nidus for embolism in our patient remains unknown and cryptogenic.

At present, no definitive clinical guidelines are available for the management of ALI in pediatric patients. The American College of Chest Physicians evidence-based clinical practice guidelines suggest therapeutic anticoagulation using intravenous unfractionated heparin or low-molecular-weight heparin for neonates or children presenting with acute femoral artery thrombosis,

although the level of evidence is weak and focused on occlusions resulting from iatrogenic arterial injury (grade 1C).[11] However, the present patient had extensive multilevel thrombosis and could have been potentially harmed by a trial of anticoagulation treatment only.

In absence of trials specific to the pediatric population, treatment guidelines have been extrapolated from adult studies, which might not be appropriate because pediatric patients will respond to ischemia differently than do adults.[2,5,11,13] Additional factors should be considered with pediatric cases, including the risk of radiation exposure.[14] A completion angiogram was warranted for our patient to ensure inline flow to the foot and the absence of significant residual thrombus that could have led to recurrence.

CONCLUSIONS

Pediatric ALI is uncommon and, if not managed promptly, can be limb- and/or life-threatening. We have described a rare case of pediatric cryptogenic ALI with multiorgan embolization with an unclear source, for which prompt surgical management with advanced imaging guidance led to successful treatment.

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