Aortic quadfurcation with persistent left sciatic artery: an extremely rare anatomic variant in a 3-year-old boy

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ABSTRACT

Reports of aortoiliac variant anatomy are rare, especially in the pediatric population. A 3-year-old male patient with hypertension and left foot polydactyly with syndactyly was referred to our interventional radiology service for evaluation of a possible renovascular cause of the hypertension. Angiograms revealed an extremely rare anatomic variant consisting of the absence of the common iliac arteries bilaterally, resulting from quadfurcation of the abdominal aorta into the bilateral internal and external iliac arteries. Additionally, a persistent left sciatic artery was identified. (J Vasc Surg Cases and Innovative Techniques 2021;7:262-5.)

Keywords: Angiogram; Anatomic variant; Aortoiliac; Pediatrics; Quadfurcation

Aortoiliac anatomic variations are rare, with few cases described in reported studies.[1] Most reported cases have been in adults and were discovered incidentally through the increasingly widespread use of diagnostic imaging performed to determine the cause of abdominal or lower extremity pain or cadaveric dissection.[2-14] The identification and knowledge of these anatomic variants is important when planning both open surgical and endovascular approaches. We present the case of a pediatric patient with extremely rare variant anatomy in which the distal aorta had divided into the bilateral internal and external iliac arteries without discernable common iliac arteries (aortic quadfurcation). The parents of the patient provided written informed consent for the report of their child's case details and images.

CASE REPORT

A 3-year-old boy with poorly controlled hypertension despite two medications, proteinuria, and left foot polydactyly with syndactyly had been referred to the interventional radiology service for evaluation of a renovascular cause of the hypertension after an initial workup. The initial evaluation had included renal ultrasonography, abdominal magnetic resonance angiography (MRA), and renal biopsy. The renal ultrasound examination showed enlarged echogenic kidneys bilaterally, and abdominal MRA demonstrated no vascular cause for the hypertension (Fig 1). The appearance of the distal aorta was believed to be normal, and the presence of a left persistent sciatic artery was identified (Fig 2). Open surgical wedge renal biopsy revealed glomeruli with proliferative changes and rare crescents but no evidence of concomitant immune complex-mediated glomerulonephritis. Mild chronic tubulointerstitial disease was noted. Because the presence of a treatable vascular cause of hypertension would have made a significant difference in the treatment of the patient and because the clinical concern for such a lesion remained high, we decided to perform catheter angiography.

With the patient under general anesthesia, abdominal aortography, followed by selective bilateral renal arteriography, was performed via right common femoral arterial access using a 4F system. Aortic pressure measurements were performed from the aortic arch through the infrarenal aorta, which demonstrated no significant pressure gradient. Two renal arteries were present bilaterally, which appeared normal. Distally, in both kidneys, the arteries were abnormally tortuous, with areas of occlusion and aneurysm formation (Fig 3, *A*). Additionally, instead of the typical aortic bifurcation, the aorta had quadfurcated into the bilateral external and internal iliac arteries at the L4–L5 disc space, with the absence of bilateral common iliac arteries. The left internal iliac artery also continued as a persistent sciatic artery (Fig 3, *B*) and was the dominant blood supply to the left lower extremity. The left femoral artery gradually disappeared just above the knee joint, with the left persistent sciatic artery continuing as the left popliteal artery.

DISCUSSION

Anatomic variations of the aortoiliac system are rare and occur with less frequency than those of the thoracic aorta. Although the exact incidence is unknown, a previous study of 8000 angiograms found only six cases of aortoiliac and iliofemoral variations.[1] Previously described anomalies included the absence of the internal iliac arteries bilaterally, aplasia of the external iliac arteries bilaterally, aplasia of the common iliac artery unilaterally, and unilateral absence of the external iliac artery.[6] In addition, quadfurcation of the abdominal aorta has been described in two postmortem adult cadaveric studies and in a 22-year-old woman who had undergone surgery

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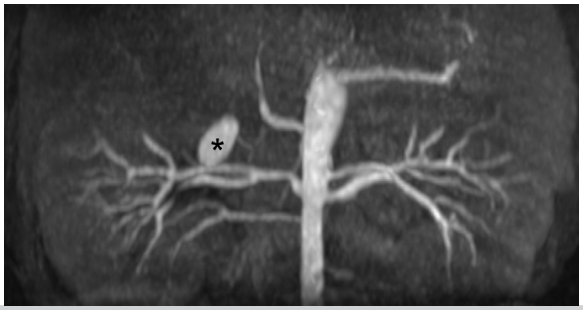

Fig 1. Reconstructed coronal maximum intensity projection inversion recovery sequence of magnetic resonance image showing renal arteries without major stenosis. Note the presence of the gallbladder, not a renal artery aneurysm, on this reconstructed image (*black asterisk*).

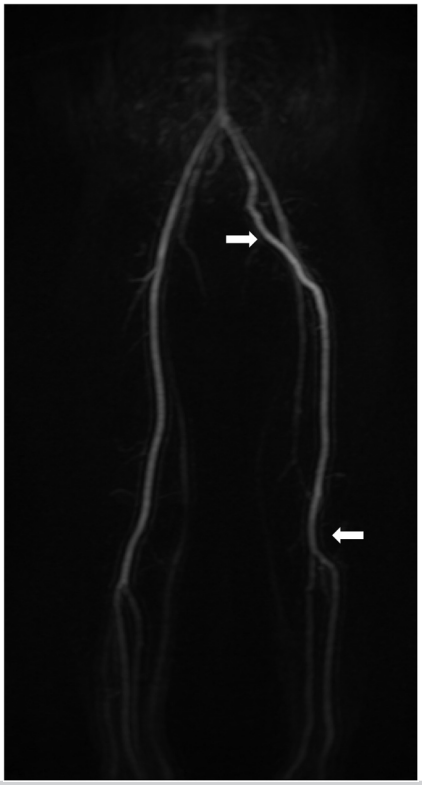

Fig 2. Subtracted coronal time-resolved image of contrast kinetics image showing persistent left sciatic artery as the dominant artery supplying the left lower extremity (*white arrows*). The presence of aortic quadfurcation would have been extremely challenging to identify on the image.

for placenta accreta.[3,5,15] Our pediatric patient exhibited two distinct vascular anomalies: the absence of both common iliac arteries and a persistent sciatic artery. Tamisier et al[13] grouped the congenital anomalies of the external iliac artery into three categories: (1) anomalous origin or course of the artery; (2) hypoplasia or atresia compensated for by a persistent sciatic artery, such as occurred in our patient; and (3) isolated hypoplasia or atresia, potentially leading to ischemia.

The occurrence of aortoiliac variant anatomy stems from the embryologic development of the aorta. Formation of the aorta begins during the third week of gestation when cells from the endocardial mesenchyme migrate along the neural groove to form two dorsal aortae.[16] During the fourth week of gestation, the umbilical arteries form an initial connection with the dorsal aortae.[17] At approximately the 6-mm embryo stage (fifth week of gestation), the umbilical arteries break their initial connection with the dorsal aorta and form anastomoses with the fifth pair of lumbar intersegmental artery branches.[17] The dorsal root of the umbilical arteries then give rise to the primitive sciatic artery, which becomes the initial dominant supply for the lower limb buds, eventually forming the adult popliteal and peroneal vessels. This connection with the lumbar intersegmental artery branch will later become the internal iliac arteries.[18] Along the umbilical arteries and proximal to the sciatic arteries, the external iliac arteries begin to form. By the 12-mm embryo stage, the external iliac arteries will have developed into the common and superficial femoral arteries, and, by the 18-mm embryo stage, flow to the lower extremities will occur predominantly through the femoral artery via the external iliac artery. In typical embryologic development, the sciatic artery will begin to regress and flow to the popliteal artery through this vessel will be discontinued by the 22-mm embryo stage.[18,19] The portion of the fifth lumbar intersegmental artery branches proximal to the internal and external iliac arteries forms the common iliac arteries.[20]

In the present patient, the left sciatic artery failed to regress and remained as the dominant blood supply to the lower extremity. Concomitantly, hypoplastic development of the left external iliac artery had occurred. The absence of both common iliac arteries could have resulted from malformation of the fifth pair of intersegmental artery branches, causing both internal and external iliac arteries to develop separately on the dorsal aorta, as described by Kara et al.[9] Alternatively, formation of the external iliac artery along the umbilical artery in a higher than normal position proximal to the sciatic artery or directly from the fifth lumbar intersegmental artery could account for this absence.

Identification of a treatable anatomic cause is an important aspect of the evaluation and treatment of children and adults with hypertension. Intervention can potentially prevent the long-term consequences from exposure to chronically elevated blood pressure, such as stroke and left ventricular hypertrophy. In the present patient, the findings of peripheral renal arterial tortuosity with occlusion and aneurysm formation were most consistent with developmental dysplasia. These lesions are not amenable to percutaneous angioplasty or other forms of minimally invasive or open surgical intervention. The lack of a correctable vascular cause of hypertension and the progression of kidney disease resulted in our

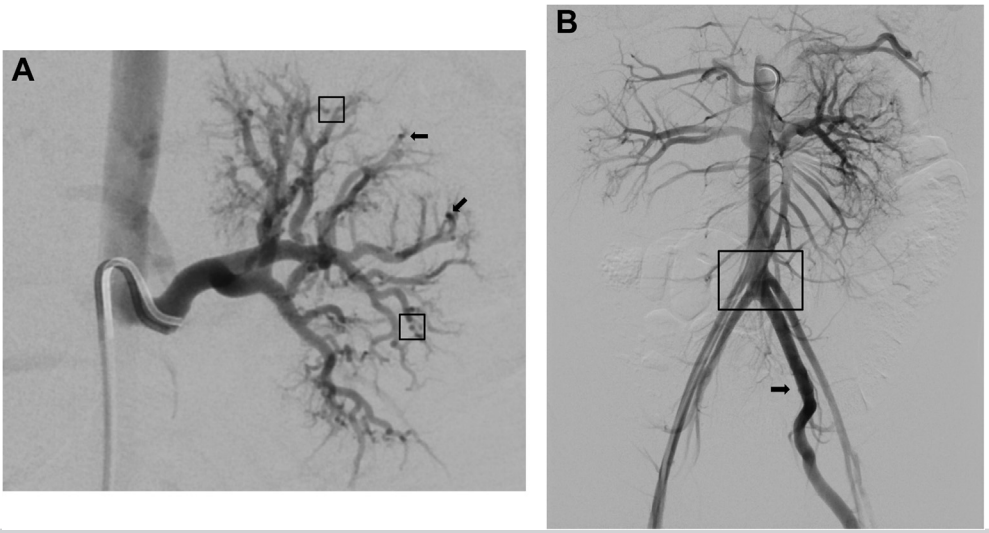

Fig 3. A, Frontal digital subtraction angiogram of one left renal artery demonstrating areas of occlusion (*black rectangles*) and areas of aneurysm formation (*black arrows*). **B,** Frontal digital subtraction abdominal aortogram image demonstrating aortic quadfurcation (*black rectangle*) and left persistent sciatic artery (*black arrow*).

patient being listed for renal transplantation. Knowledge of the pelvic arterial anatomy will be useful for the transplant surgeon to select the best possible place to create an arterial anastomosis for the transplanted kidney. In the present patient, the surgeon might prefer to use the right external iliac artery because it is larger than the left. This could, in turn, influence which side would be catheterized if the patient were to require arterial evaluation of the transplanted kidney.

The aortic quadfurcation was not identified on the initial MRA of the abdomen. In hindsight, one might have been able to visualize the abnormality. However, such a possibility would have been difficult to comment on prospectively given the uncommon nature of the finding and the inferior spatial resolution of MRA compared with that of conventional angiography. As imaging techniques continue to evolve, the frequency of the identification of arterial anomalies is likely to increase.

CONCLUSION

To the best of our knowledge, we have presented the first case of the absence of both common iliac arteries with a persistent unilateral sciatic artery in a pediatric patient. The identification of such anomalies is paramount in the setting of potential renal transplantation or other interventions.

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