

Multiple idiopathic aneurysms in a 10-year-old child

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CASE SUMMARY

A 10-year-old African-American child with chronic kidney disease, anemia, hypertension, history of stroke, and multiple aneurysms seen on prior imaging, presented with respiratory distress, swelling and cough. Review of systems revealed recent oliguria.

An extensive diagnostic workup for the cause of the multiple aneurysms was performed. An infectious or immune-mediated etiology was originally suspected, though testing for HIV, tuberculosis, and fungal infections was negative. P-ANCA, C-ANCA, MPO Ab, Pr3 Ab, and ANA were negative. Complement levels, ferritin, and Von Willebrand antigen were within normal limits.

After the aforementioned workup, a genetic cause was felt to be more likely. There was no significant family history on the mother's side, and the father's family history was largely unknown. A urine spot mucopolysaccharidosis test was negative. Genetics testing of the following genes was performed, though no pathogenic mutations or clinically

significant variants were detected: ACTA2, CBS, COL3A1, FBN1, FBN2, MYH11, SLC2A10, SMAD3, TGFB1 and TGFB2. The patient was found to be heterozygous for the c.3651+5_+11del7insG variant of the MYH11 gene, though was of unknown significance.

IMAGING FINDINGS

Ultrasonographic evaluation of the kidneys was performed based on the patient's presenting symptoms and history of multiple aneurysms. Imaging revealed a 4.4 × 2.5 cm partially thrombosed left renal artery aneurysm, as well as an aneurysm of the right renal artery (Figures 1 and 2).

After reviewing the risks and benefits, CT of the abdomen and pelvis with intravenous contrast was performed, which demonstrated aneurysmal dilatation involving the infrarenal abdominal aorta. Large partially thrombosed aneurysms with mural calcifications also affected the bilateral main renal arteries, which caused mass effect on the bilateral kidneys. Addi-

tional findings included partial visualization of a large hematoma in the right pararenal space, as well as intraperitoneal free fluid, a large pericardial effusion, and bilateral pleural effusions (Figures 3 and 4); three-dimensional reconstructed images were also obtained (Figure 5).

A Tc-99m DMSA nuclear medicine scan revealed a large defect in the superior pole and hilar region of the right kidney. The left kidney demonstrated a large defect in the medial portion of the midpole (Figure 6). The findings are consistent with CT imaging of mass effect from the renal artery aneurysms.

Angiography was performed, with CO₂ as a safer alternative to iodinated contrast material, via a 4-French S&S catheter. The renal arterial and infrarenal abdominal aortic aneurysms were again identified (Figure 7). The patient was also found to have a right arteriovenous shunt (Figure 8).

DIAGNOSIS

Multiple idiopathic aneurysms

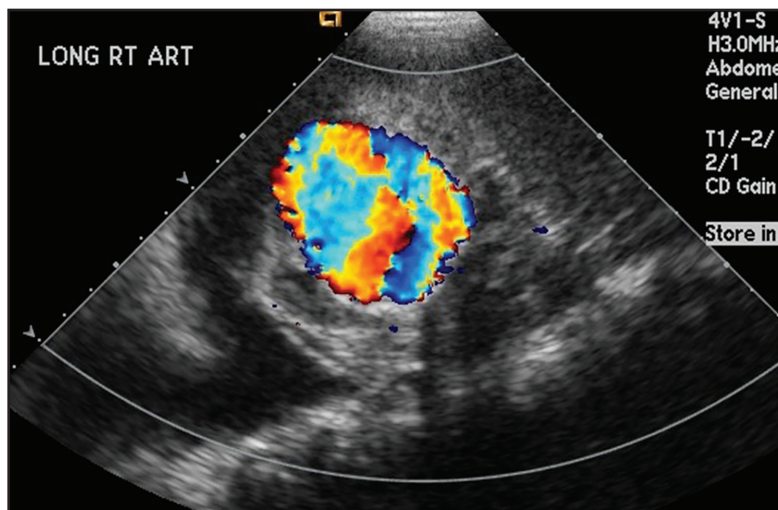
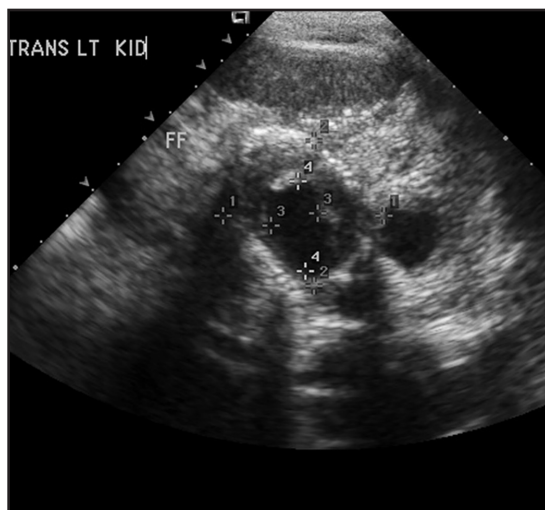


FIGURE 1. Ultrasonography demonstrating a 4.4×2.5 cm partially thrombosed left renal artery aneurysm.

FIGURE 2. The right renal artery aneurysm seen on Doppler imaging.



FIGURE 3. Coronal CT abdomen and pelvis with IV contrast demonstrates an aneurysm of the infrarenal abdominal aorta (black arrow), and large partially thrombosed aneurysms of the bilateral renal arteries with mural calcifications (white arrows). Additional findings include a right lower quadrant hematoma (broken black arrow), as well as intraperitoneal free fluid, pleural effusions, and a pericardial effusion (broken white arrows).



FIGURE 4. Axial CT abdomen and pelvis with IV contrast displays the partially thrombosed bilateral renal artery aneurysms with mass effect on the bilateral kidneys (white arrows).

DISCUSSION

Multiple arterial aneurysms in the pediatric population are distinctly rare phenomena. They are likely secondary to an underlying source, such as trauma, infection, vasculitides, vascu-

lar dysplasias, and non-inflammatory medial degeneration, among other etiologies.¹ Genetic syndromes and collagen vascular diseases are another potential source of multiple aneurysms in children, and include entities such as

Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, thoracic aortic aneurysm and dissection (TAAD), as well as Turner syndrome, neurofibromatosis, and autosomal dominant polycystic kidney disease

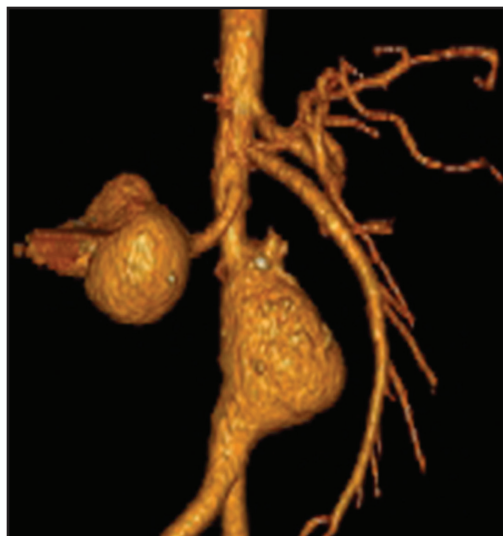


FIGURE 5. An obliquely oriented three-dimensional reconstruction shows the infrarenal abdominal aortic aneurysm. The right renal artery aneurysm is partially visualized.

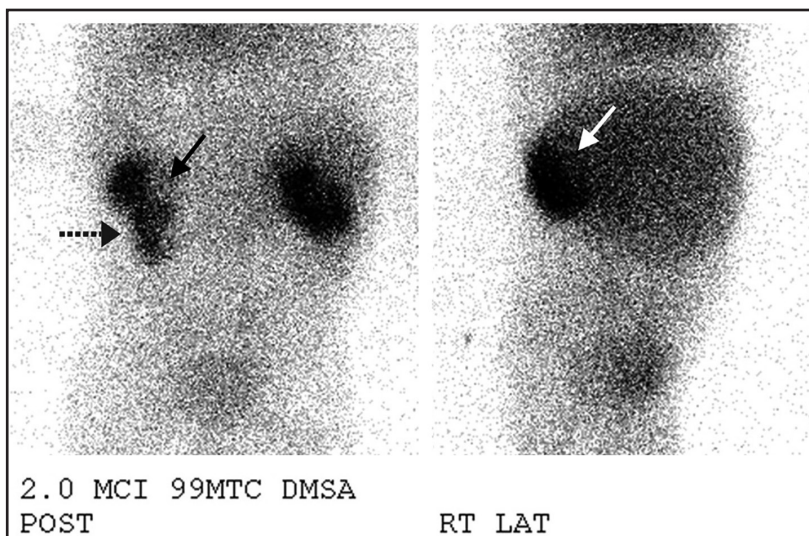


FIGURE 6. Tc-99m DMSA scan reveals large renal defects from aneurysmal mass effect in the medial portion of the left kidney (black arrow), and the superior pole and hilar region of the right kidney (white arrow). A small cortical defect in the inferior pole of the left kidney is also noted (dashed arrow).

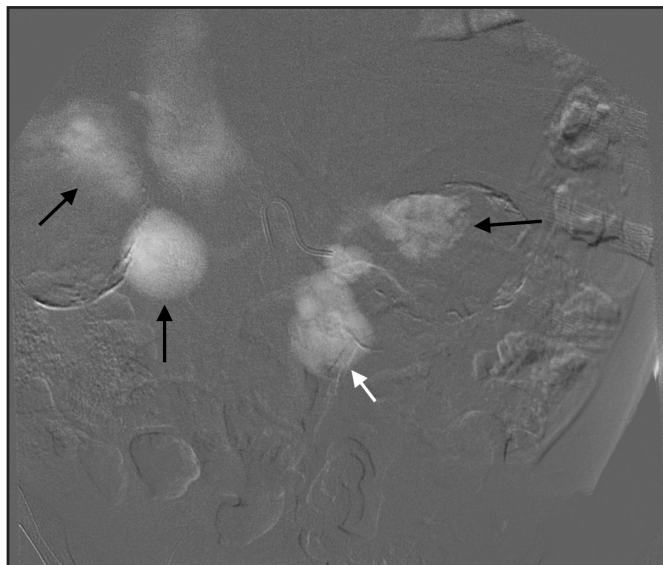


FIGURE 7. Angiogram with CO₂ contrast highlights the bilateral renal aneurysms (black arrows) and infrarenal abdominal aorta aneurysm (white arrow).

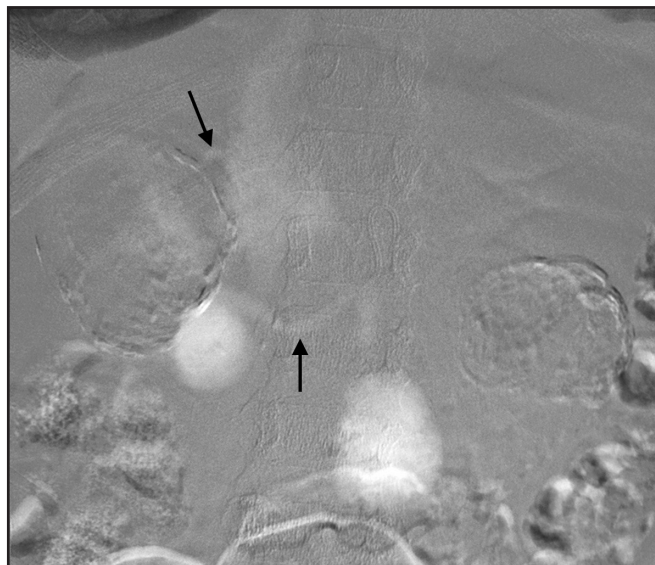


FIGURE 8. CO₂ angiogram again demonstrates the right renal and infrarenal abdominal aorta aneurysms, as well as a right renal AV shunt (black arrows).

(ADPKD).² The least common of all types is idiopathic, for which all other etiologies are first excluded by clinical and laboratory evaluation.

While the aforementioned syndromes may be diagnosed clinically and corroborated with genetic testing, the predilections for specific vascular

territories may also help differentiate among them, though significant overlap precludes definitive radiologic delineation. As an example, Turner syndrome and TAAD preferentially affect the thoracic aorta, while ADPKD may involve the thoracic, cervicocephalic, or intracranial

vasculature.² The renal arteries and abdominal aorta may be affected with neurofibromatosis, while aneurysms of both the thoracic and abdominal aortas are associated with Marfan syndrome and Ehlers-Danlos syndrome.² As such, in this case the differential diagnosis was able to be somewhat revised

once patient imaging was obtained. However, the subsequent outcome of the genetic analysis excluded the aneurysmal syndromes as potential causes, resulting in an idiopathic etiology for these multiple aneurysms.

Idiopathic arterial aneurysms are reported infrequently in the literature since Miguel first described a case in 1834.¹ Since then, although the small number of cases precludes definitive associations, some important information has been obtained. They most commonly involve the aortoiliac system, renal arteries, and upper extremities. There is a slight preponderance towards boys, with a mean age of presentation less than 10 years. Bilateral involvement is common, as is the simultaneous occurrence of both abdominal and renal aneurysms. Additionally, long-term follow up is important, as interval development of additional aneurysms has been reported in a handful of cases.^{1,3}

An important note in this case is that the patient's autopsy reported medial elastin disruption of the aneurysmal walls, as well as regional fragmentation of elastin fibers in the skin, suggesting an underlying elastic fiber disorder. Review of prior case reports demonstrate similar findings including decreased elastin fibers and fragmentation of those fibers.³⁻⁵ Such findings in cases with continued new sites of aneurysm formation suggest an ongoing elastolytic process as a possible cause of this disease.³

Clinically, idiopathic aneurysms and aneurysmal syndromes can present variably, though certain features

are more common. Large and medium vessel aneurysms involving the renal arteries often present with renal impairment, and large aneurysms can cause a mass effect leading to hepatic venous congestion; both of these effects can further lead to generalized edema. Complaints of patients with aneurysms may include any of the symptoms of abdominal fullness, pain in the abdomen or lower back, or as a pulsatile tumor, but are otherwise rarely symptomatic unless non-vascular organ systems are compromised.^{3,4}

Treatment for these aneurysms consists of open surgical or endovascular techniques, the latter of which include graft placement and coil embolization.^{3,4} Use of traditional contrast may be contraindicated during angiography depending on any preexisting kidney damage. However CO₂ contrast angiography has begun to play a broader role in these situations, in which the aneurysms are seen as contrast retaining dilatations of the affected vessels. Of note, the aforementioned interventions treat the existing aneurysms though are not considered curative. If surgical intervention is not feasible, studies in animal models and in vitro studies suggest that β -blockers, and antibiotics such as doxycycline or azithromycin, may help limit growth in aneurysms.^{5,6} However, to date there is no evidence that these therapies definitively prevent expansion.

CONCLUSION

Multiple aneurysms in the pediatric population are unusual phenomena. An idiopathic etiology is very rare, with

only a few cases reported in the literature. Our case focused on a 10-year-old boy with hypertension, chronic kidney disease, and recent oliguria, who on multiple imaging modalities demonstrated aneurysms of the bilateral renal arteries and infrarenal abdominal aorta.

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Prepared by Dr. Patel while a Radiology Resident, Mr. Arndt while a Medical Student, Dr. Nguyen while an Assistant Professor, and Dr. Ezell while an Assistant Professor in the Department of Radiology at Tulane University School of Medicine, New Orleans, LA.