Arterial aneurysms rarely occur in children. When present, an underlying pathologic process is usually discovered, such as infection, trauma, collagen vascular disease, arteritides, congenital arterial malformations, and others. Even less common are multiple pediatric aneurysms without a recognizable etiology (idiopathic aneurysms).1-12 We describe a 6-year-old boy who presented with symptomatic multiple arterial aneurysms. Along with the diagnostic workup, surgical treatment, and pathologic findings, a review of the published literature is presented.

CASE REPORT

A 6-year-old African American boy presented to the emergency department with a 2-day history of worsening right flank pain. He also had nausea, vomiting, and diarrhea; there was no history of dysuria or hematuria. His perinatal history was unremarkable (full term, normal vaginal delivery), and immunizations were up to date. His medical and surgical history was only significant for asthma treated with β agonist inhaler as needed.

On physical examination, he was afebrile and mildly hypertensive. The rest of the examination was unremarkable except for mild right flank tenderness on deep palpation. There were no stigmata of connective tissue diseases with arterial involvement such as hyperelastic skin, hypermobile joints, nor marfanoid habitus. Routine blood tests revealed leukocytosis to 16,000/mm³ with no left shift. Urinalysis was positive for microscopic hematuria and ketones. Plain abdominal radiographs were normal, but an abdominal ultrasound scan demonstrated a right renal artery aneurysm and an infrarenal abdominal aortic aneurysm (AAA). A subsequent computed tomography (CT) scan was performed of his abdomen and pelvis, followed by aortography (Fig 1). These revealed multiple areas of right kidney infarction, a 3-cm right renal artery aneurysm (RAA), with fistula formation to the right renal vein with early filling of the inferior vena cava, and a 3-cm AAA.

The initial workup included a search for infectious and immune-mediated etiologies. Blood cultures for aerobic, anaerobic, and acid-fast bacteria were negative. Fungal cultures were also negative. The erythrocyte sedimentation rate was 52 on admission and later dropped to 11. Results were negative for serum antineutrophil antibodies, perinuclear and cytoplasmic antineutrophil cytoplasmic autoantibodies, rapid plasma regain, and hepatitis B and C surface antigens.

Funduscopic was normal. The results of echocardiography and magnetic resonance angiography of his carotids and intracranial vessels were normal. A nuclear perfusion scan demonstrated normal function in the left kidney, with very little residual kidney function in the right. Meanwhile, the patient’s hospital course was significant for progressively worsening hypertension that required three antihypertensive medications for good control.

Surgical exploration through a midline transperitoneal approach revealed a large right RAA with fistula formation to the renal vein, and an infrarenal AAA ending above the bifurcation (Fig 2). Given the largely infarcted, nonfunctional status of the right kidney, no attempt at partial preservation was made, and it was resected by using standard techniques. Next, AAA exposure and control were obtained. After weight-based systemic heparinization, standard aneurysmorrhaphy was performed with a 10-mm expanded polytetrafluoroethylene tube graft using an interrupted suture technique with nonabsorbable monofilament sutures.

The patient’s postoperative course was uneventful. His hypertension resolved and he was discharged home on postoperative day 4 on pain medication and an inhaled bronchodilator.

The pathologic examination of the aortic and renal arterial tissue revealed intimal and medial fibroplasia with fragmentation of the elastic fibers and increased mucopolysaccharides, with few Giant cells but no inflammation (Fig 3). Immunofluorescent stain...
ing of the aortic wall for immunoglobulin (Ig)-G, Ig-M, Ig-A, and C-3 was negative. Culture of the aneurysm wall for bacteria was also negative.

At 1-year follow-up, his physical exam is unremarkable and he remains normotensive with a normal creatinine; CT scan reveals a well-incorporated graft without evidence of other abdominal or pelvic aneurysms.

**DISCUSSION**

Aneurysms in the pediatric age group are uncommon and their etiologies are variable. A clinicopathologic classification proposed by Sarkar1,3 includes nine categories, with idiopathic aneurysms being the least common. The more common etiologies are infection, trauma, arteritides,
Kawasaki disease, vascular dysplasias, noninflammatory medcial degeneration, and others. Presentation can vary, ranging from asymptomatic to rupture. Etiologic evaluation must exclude all other possibilities before a diagnosis of idiopathic aneurysms in children. A history of febrile illness with rash and mucous membrane inflammation may indicate autoimmune connective tissue diseases. Similarly, a history of sepsis or umbilical artery catheterization may indicate infected aneurysms. A careful examination of the skin for hyperelasticity and the joints

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Gender</th>
<th>Vessels involved</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Miguel¹</td>
<td>1834</td>
<td>14</td>
<td>M</td>
<td>AA, upper extremity</td>
<td>None</td>
<td>Rupture/death</td>
</tr>
<tr>
<td>Williams²</td>
<td>1975</td>
<td>5</td>
<td>M</td>
<td>AA, bilateral common iliac</td>
<td>Aortic reconstruction, left nephrectomy</td>
<td>—</td>
</tr>
<tr>
<td>Short³</td>
<td>1978</td>
<td>7</td>
<td>F</td>
<td>AA, bilateral renal, right brachial common iliac and anterior tibial, bilateral popliteals</td>
<td>Extremity saccular aneurysms serially resected aortic and renal aneurysm left in situ</td>
<td>Good (14 yrs)</td>
</tr>
<tr>
<td>Schiller⁴</td>
<td>1983</td>
<td>8</td>
<td>M</td>
<td>AA, right renal, internal iliac, brachial, circumflex humoral, popliteal and posterior tibial, bilateral superficial femoral</td>
<td>1. Aortic reconstruction</td>
<td>Good</td>
</tr>
<tr>
<td>O’Hara⁵</td>
<td>1985</td>
<td></td>
<td></td>
<td></td>
<td>2. Redo aortic reconstruction with ligation of right hypogastric and ex-vivo repair of right RAA</td>
<td></td>
</tr>
<tr>
<td>Fee⁶</td>
<td>1983</td>
<td>3</td>
<td></td>
<td>Left subclavian, ectasia of right common carotid, splenic and phrenic</td>
<td>Left subclavian aneurysm exclusion left carotid/axillary bypass</td>
<td>Good (6 mon)</td>
</tr>
<tr>
<td>Bordeaux⁷</td>
<td>1990</td>
<td>7</td>
<td>F</td>
<td>AA, left internal iliac, left renal, left brachial, left popliteal and tibial</td>
<td>Ruptured AAA reconstruction</td>
<td>Satisfactory (3 yrs)</td>
</tr>
<tr>
<td>Lanfermann⁸</td>
<td>1990</td>
<td>6</td>
<td>M</td>
<td>AA, bilateral renal and common iliac, inferior mesenteric, right hepatic, bilateral internal carotid</td>
<td>None</td>
<td>—</td>
</tr>
<tr>
<td>Halpern¹⁰</td>
<td>1997</td>
<td>6</td>
<td>M</td>
<td>AA, right brachial, radial and internal carotid</td>
<td>1. Aortic reconstruction</td>
<td>Good (3 mon)</td>
</tr>
<tr>
<td>Checinski¹¹</td>
<td>2000</td>
<td>6</td>
<td>F</td>
<td>AA, bilateral renal</td>
<td>2. Brachial artery reconstruction</td>
<td>—</td>
</tr>
<tr>
<td>English¹²</td>
<td>2004</td>
<td>10</td>
<td>M</td>
<td>AA, bilateral iliac and renal, left vertebral, brachial and radial</td>
<td>1. Left vertebral embolization</td>
<td>Good (3 mon)</td>
</tr>
<tr>
<td>Current case</td>
<td>2002</td>
<td>6</td>
<td>M</td>
<td>AA and RRA</td>
<td>2. Left brachial reconstruction</td>
<td>Good (1 yr)</td>
</tr>
</tbody>
</table>

AAA, Abdominal aortic aneurysm; AA, abdominal aorta; RAA, renal aortic aneurysm; RRA, right renal artery.

Published reports of multiple idiopathic aneurysms in children.

Callicutt et al
for hypermobility may lead to the suspicion of Marfan’s syndrome. An early rheumatologic consultation is well advised in these cases.

Although not assayed in this patient, elevated serum elastase levels were reported in one of these 12 multiple idiopathic aneurysms cases and in another child with an isolated thoracoabdominal aneurysm. Pediatric aneurysm secondary to Behçet’s disease has also been recently reported, however none of the clinical or pathologic findings were present in this case.

The results of histologic examinations of idiopathic pediatric aneurysms have varied but consistently show intimal fibroplasia, which probably reflects a secondary process rather than an etiologic factor. There is also lack of inflammation or cystic medial degeneration, thus excluding other known etiologies. In this case and in the one reported recently by English et al, Giant cells were encountered on other known etiologies. In this case and in the one reported in inflammation or cystic medial degeneration, thus excluding inflammation, negating the diagnosis of arteritis.

To date, including this current case, only 12 cases of multiple idiopathic aneurysms in pediatric patients have been reported (Table). The locations of the aneurysms in these cases most commonly involve the aortoiliac system in 10 (83%), renal arteries in 9 (75%), and upper extremities in 9 (75%). Other locations in decreasing frequency include the iliac artery or its branches (58%), the carotid/vertebral circulation (33%), the mesenteric vessels (25%), and lower extremity arteries (25%). Because of the small number of cases reported, no conclusive trends or associations can be made; however, one can note the following observations:

1. Sixty-six percent of the patients were boys.
2. The mean age of presentation was 8 years (median, 6 years).
3. The association between AAA and RAA (unilateral or bilateral) appears very strong, with only 1 (11%) of 9 cases of RAA occurring without an AAA, and 1 (10%) in 10 cases of AAA occurring without an RAA. Although upper extremity aneurysms are as common as RAA, their association is not as strong (7 of 10 AAA had upper extremity aneurysms).
4. Bilateral involvement is common, with 7 (58%) of 12 cases involving bilateral structures.
5. In the three cases with reported lower extremity aneurysms, the tibial and popliteal arteries were involved in each patient.
6. The clinical outcome appears to be generally good with surgical repair, with variable length of reported follow-up.
7. The development of new aneurysms at other locations during follow-up was reported in two cases, highlighting the need for long-term observation.

The aim of surgical repair is to prevent aneurysm-related complications such as rupture, distal embolization, and death. The surgical principles are similar to adults, with few modifications.

The choice of vascular conduits for the aorta is particularly difficult given the need for expansion as the child grows. Although autogenous conduits such as the right internal iliac artery are appealing, their future integrity is unknown in patients with idiopathic aneurysms. Most of the reported cases used prosthetic grafting as in our case. We used a slightly oversized graft (2 mm) and an interrupted suture technique to minimize the purse-string effect of the running suture. There is still concern, however, about long-term integrity of the prosthetic graft and suture line as well as hemodynamic compromise as the child grows. This, along with the potential of developing new aneurysms, underscores the need for periodic follow-up.

**REFERENCES**


Submitted Sep 11, 2004; accepted Feb 7, 2005.