Renal arteriovenous aneurysm in a 4-year-old patient

Joe Potchanard Chauvapun, MD, Michael G. Caty, MD, and **Linda M. Harris, MD,** *Buffalo, NY*

We report a case of a symptomatic renal arteriovenous aneurysm in a 4-year-old pediatric patient. We were able to diagnose the lesion by means of a Doppler renal sonogram with color duplex interrogation. The diagnosis was confirmed by digital subtraction angiography. On the basis of the angiographic findings, the aneurysm was resected, and the renal arteriovenous fistula was repaired. (J Vasc Surg 2005;41:535-8.)

Renal arteriovenous fistula is an uncommon lesion. The peak incidence occurs in patients between 30 and 40 years old.[1] Rarely is it found in the pediatric population. There are 2 types of renal arteriovenous fistula: congenital and acquired. The latter is more common.[2] Many renal arteriovenous fistulas are asymptomatic; hence, the true incidence of the disease is unknown.[3] Rarely, renal arteriovenous fistula may be associated with renal artery aneurysm. In such cases, the term *arteriovenous aneurysm* may be correctly applied to such a lesion. Clinical manifestations of renal arteriovenous fistula include hypertension, left ventricular hypertrophy, cardiac failure, hematuria, and abdominal pain.[4] Treatment options include vascular ligation, subtotal excision of a renal arteriovenous aneurysm with direct suture ligation of their communication, nephrectomy, selective embolization, balloon catheter occlusion, and endovascular repair.

CASE REPORT

A 4-year-old boy was evaluated for a well-child visit and was found to be hypertensive. He was referred to the renal clinic for further evaluation. There was no history of trauma and no hematuria, and he had no complaints of abdominal pain. On physical examination, the patient weighed 22.3 kg (>95th percentile), with a height of 104.2 cm (50th percentile) and a blood pressure of 144/90 mm Hg (>95th percentile). No abdominal bruit or mass was identified. His laboratory tests were within normal limits, including a urinalysis that revealed a negative dipstick and a specific gravity of 1.020. Renin plasma level and 24-hour urine for catecholamine were within normal limits. The patient was started on 2.5 mg of enalapril daily. A renal sonogram with duplex study was obtained and demonstrated the left kidney to be 7.3 cm long and the right kidney to be 8.1 cm long. The contour of the kidney was noted to be of normal shape and echo texture. Of note, there was a lesion just external to the left kidney hilum which measured 3.3 × 1.8 × 1.7 cm. Color Doppler interrogation showed a mixed flow signal composed of venous flow with some arterial overlay. A renal angiogram was obtained that revealed an aneurysm with a narrow neck arising 1 cm from the origin of the left renal artery (Fig 1).

The patient was taken to the operating room for repair of the vascular lesion. A midline incision was made, and the abdomen was explored. The descending colon was mobilized to allow access to the renal vessels. The aneurysm was identified. The proximal renal artery and vein were dissected free and looped. There was significant inflammation around the aneurysm. Before further dissection, the left hypogastric vessel was mobilized in case it was needed for bypass. Systemic heparin and mannitol were given for renal protection. The renal vessels were controlled with vessel loop and vascular clamps. The aneurysm was then opened. Upon entry into the aneurysm, a single pinpoint opening of blood flow was noted. This was confirmed by proximal clamp relaxation. The inflow was repaired by oversewing from within the aneurysm. The outflow was noted to be venous. This was confirmed by placing a sound probe and a Fogarty catheter into the outflow tract. The venous outflow was ligated with a figure of 8 stitch from inside the aneurysm. Aneurysmal tissue was removed and sent to pathology. The lesion was irrigated, and the abdomen was closed.

After the operation, the patient recovered uneventfully. A postoperative arteriogram revealed no evidence of stenosis or aneurysmal dilatation. A radiologic "beak" was noted at the site of the previous arteriovenous fistula (Fig 2). The pathology result was consistent with arterial dysplasia. Immediately after the operation, the patient's blood pressure remained increased. However, on 6-month follow-up, his blood pressure remained controlled, requiring only 1.0 mg of captopril daily.

DISCUSSION

A renal arteriovenous fistula was first described by Vorela[5] in 1928. This lesion is uncommon; however, its incidence has been described with increasing frequency, with more than 200 reported cases.[6] Renal arteriovenous fistula may be congenital or acquired. The exact cause of congenital renal arteriovenous fistula is unknown; however, it is thought to be present at birth or to be a result of a congenital aneurysm that erodes into an adjacent vein.[7] The congenital form is rare with an incidence of 0.04%, encompassing only 25% of renal arteriovenous fistulas. Cho and Stanley[1] studied a series of 9500 angiograms of the

From the University at Buffalo School of Medicine and Biomedical Sciences and Kaleida Health.

Competition of interest: none.

This article won the award for the Junior Resident Paper at the Seventeenth Annual Meeting of EVS in New York, May 1-4, 2004.

Reprint requests: Linda M. Harris, MD, Department of Surgery, Millard Fillmore Hospital, 3 Gates Circle, Buffalo, NY 14209 (e-mail: lharris@kaleidahealth.org).

0741-5214/$30.00

doi:10.1016/j.jvs.2004.12.032

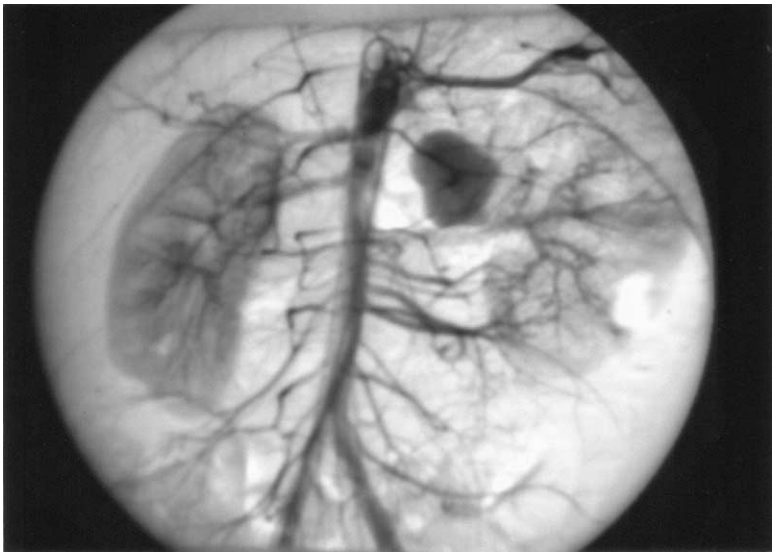

Fig 1. Preoperative renal angiogram demonstrating left kidney arteriovenous fistula. The lack of opacification of the inferior vena cava is due to the low flow within the fistula.

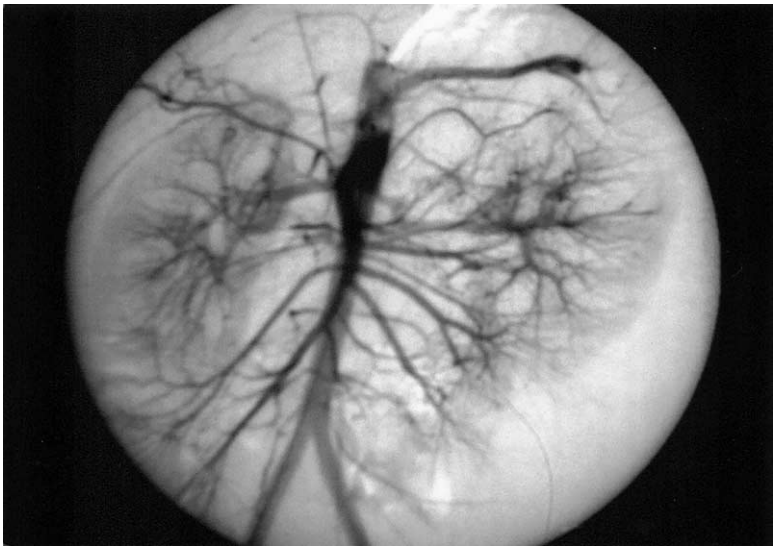

Fig 2. Postoperative renal angiogram revealed no evidence of stenosis or aneurysmal dilatation.

renal artery and found only 4 cases of congenital origin. Acquired renal arteriovenous fistula is more common and occurs secondary to trauma, inflammation, renal surgery, renal angioplasty, or percutaneous biopsy.[8-13]

The most common symptom of renal arteriovenous fistula is hematuria.[8] It is reported in 72% of the reported cases. The patient often presents with microscopic hematuria. However, cases of massive hematuria have been reported.[14] In our case, the patient did not have hematuria.

The pathophysiology of cardiovascular derangement is due to the shunting of blood.[15] The blood flow bypassed the renal parenchyma and promoted rapid venous return. As blood is shunted from the parenchyma, it causes relative renal ischemia and renin-mediated vasoconstriction and fluid retention. With a long-standing renal arteriovenous fistula, increased venous return leads to high cardiac output. This can result in left ventricular hypertrophy and, eventually, cardiac failure. The central overload also depends on the size of the arteriovenous fistula and the proximity to the heart. In our case, because of the early detection of this lesion and the small size of the fistula, cardiac derangements were not appreciated.

Color duplex imaging has become the noninvasive test of choice for screening patients for renal arteriovenous fistula.[16,17] The use of color Doppler ultrasound imaging for detecting renal arteriovenous fistulas is limited by the difficulty in distinguishing an arteriovenous fistula from an aneurysm. Dynamic computed tomography can demonstrate prompt venous filling and can identify the renal arteriovenous fistula.[18] The improvement in ultrasound 3-dimensional technology may make this the diagnostic modality of choice. The 3-dimensional reconstruction provides information regarding the location, orientation, and spatial relationship to other vascular structures. Mohaupt et al[17] have demonstrated that 3-dimensional ultrasound angiography may be as effective as magnetic resonance imaging angiography in identifying and localizing renal arteriovenous fistulas. However, selective renal arteriography or digital subtraction angiography remains the definitive diagnostic modality for renal arteriovenous fistula.[8] In our case, the color duplex scan suggested evidence of a renal arteriovenous fistula. Our angiographic study was limited by the low venous return; thus, the rapid opacification of the inferior vena cava was not noted.

Patients diagnosed with renal arteriovenous fistula should be closely observed for the development of symptoms. Intervention should be reserved for symptomatic patients. Individuals with renal arteriovenous fistula discovered after percutaneous biopsy should be followed up closely with periodic duplex surveillance, because most fistulas will close spontaneously.[19]

Surgical options for symptomatic renal arteriovenous fistula include ligation of feeding vessels, nephrectomy, partial nephrectomy, and ex vivo repair for deep intrarenal fistulas.[20-30] The goal of renal arteriovenous fistula treatment is the preservation of functioning renal parenchyma and the eradication of symptoms.[8] The evolution of surgical management is summarized in Table. Many authorities believe that surgery is the best treatment for renal arteriovenous fistula. Our patient underwent surgical exploration because of symptomatic hypertension that was not resolved with medical treatment. With the aneurysm dissected, the venous outflow was ligated with a figure 8 from the inside of the renal arteriovenous aneurysm. This was the simplest and most effective means of repairing this lesion. Renal function was preserved, as evidenced by a stable blood urea nitrogen and creatinine. Follow-up renal angiography revealed no stenosis or aneurysmal dilatation. Moreover, the patient's hypertension was better controlled and required less antihypertensive medication.

Experience has been gained over the past 20 years with percutaneous arterial embolization therapy for congenital and acquired renal arteriovenous fistula. Fibrin glue, coils, and balloons have been used to occlude fistulas.[31-33] Complications of embolization include pulmonary and peripheral arterial embolization.

In recent years, advances in endovascular technique have made possible the use of stents in the treatment of renal arteriovenous fistula.[34-36] This modality of treatment adequately closes the fistula with minimal morbidity. In cases in which the aneurysm is large and the risk of central or pulmonary embolization is high, the use of a covered stent may be advantageous, as demonstrated by Sprouse and Hamilton.[34] As advances continue in this field, endovascular embolization or stenting may become the treatment of choice for renal arteriovenous fistulas. In our case, had the identification of renal arteriovenous fistulas been made before the operation, renal arteriovenous fistula embolization may have been considered. However, in this case, the aneurysm was readily identified by the diagnostic test, and definitive identification of the renal arteriovenous fistula occurred only at the time of operation.

Furthermore, in the pediatric population, even when embolization is feasible, operative repair may remain the optimal treatment, depending on the patient's age and the size of the child's vessels. Our experience with pediatric vascular trauma demonstrated that stenting children with growing vasculature that has not reached its full maturity may lead to future stenosis. Interrupted suture repair allows for vascular growth as the child grows.

The development of surgical management for renal arteriovenous fistula

Author	*Year*	*Surgical management*
Edsman[20]	1957	Renal salvage via partial nephrectomy
Boijsen[22]	1962	Ligation of the arterial branches of a renal arteriovenous fistula
Palmer[21]	1966	Segmental nephrectomy under selective renal hypothermia
Merkel[24]	1970	Obliteration of renal arteriovenous fistula via venotomy and direct suture from the venous vessel
Merritt[26]	1972	Subtotal excision of a renal arteriovenous aneurysm via direct suture of their communication
Ehrlich[25]	1975	Renal arteriovenous fistula treated via endofistulorrhaphy with direct suture from the arterial side
Stoney[29]	1978	Ex vivo renal artery reconstruction with the use of hypothermic perfusion preservation
Morgan[27]	1981	Intrarenal vascular reconstruction for arteriovenous fistula
Boyce[28]	1981	Use of ultrasonic velocimetry in resection of renal arteriovenous fistulas

REFERENCES

1. Cho KF, Stanley JC. Non-neoplastic congenital and acquired renal arteriovenous malformations and fistula. Radiology 1978;129:333-43.
2. Maldonado JE, Sheps SG, Bernatz PE, et al. Renal arteriovenous fistula. Am J Med 1964;37:499.
3. Saliou C, Raynaud A, Blanc F, et al. Idiopathic renal arteriovenous fistula: treatment with embolization. Ann Vasc Surg 1998;12:75-7.
4. Messing E, Kessler R, Kavaney RB. Renal arteriovenous fistulas. Urology 1976;8:101-7.
5. Vorela ME. Aneurisma arteriovenoso de las vas renales y asidolia consecutive. Rev Med Latin Am 1928;14:32-44.

6. Motta J, Breslin DS, Vogel F, et al. Congenital renal arteriovenous malformation in pregnancy presenting with hypertension. Urology 1994;44:911-4.
7. Thompson WB, Ross M, Radwin HM, et al. Intrarenal arteriovenous fistula. J Urol 1972;108:526-9.
8. Crotty K, Orihuela E, Warren M. Recent advances in the diagnosis and treatment of renal arteriovenous malformation and fistulas. J Urol 1993;150:1355-9.
9. Huppert P, Duda S, Erley C, et al. Embolization of renal vascular lesions: clinical experience with microcoils and tracker catheters. Cardiovasc Intervent Radiol 1993;16:361-7.
10. Oleaga JA, Grossman RA, McLean GK, et al. Arteriovenous fistula of a segmental renal artery branch as a complication of percutaneous angioplasty. AJR Am J Roentgenol 1931;136:988-9.
11. Robinson D, Teitelbaum G, Pentecost M, et al. Transcatheter embolization of an aortocaval fistula caused by residual renal artery stump from previous nephrectomy: a case report. J Vasc Surg 1993;17:794-7.
12. Steffens J, Defreyne L, Kramann B, Hoffmann W, Ziegler M. Selective transcatheter embolization of a pediatric postnephrectomy arteriovenous fistula. Urol Int 1994;53:99-101.
13. van der Zee J, van den Hoek J, Weerts J. Traumatic renal arteriovenous fistula in a 3-year-old-girl, successfully treated by percutaneous transluminal embolization. J Pediatr Surg 1995;30:1513-4.
14. Gopalakrishnan G, Al-Awadi K, Bhatia V, Mahmound A. Renal arteriovenous malformation presenting as hematuria in pregnancy: case report. Br J Urol 1995;75:110-1.
15. McAlhany JC Jr, Black HC Jr, Hanback LD Jr, Yarbrough DR III. Renal arteriovenous fistulas as a cause of hypertension. Am J Surg 1971;122:117-20.
16. Gainza F, Minguela I, Lopez-Vidaur I, et al. Evaluation of complications due to percutaneous renal biopsy in allografts and native kidneys with color-coded Doppler sonography. Clin Nephrol 1995;43:303-8.
17. Mohaupt MG, Perrig M, Vogt B. 3D ultrasound imaging: a useful noninvasive tool to detect AV fistulas in transplanted kidneys. Nephrol Dial Transplant 1999;14:940-3.
18. Yokoyama H, Tsuji H. Color Doppler ultrasound for detection of renal arteriovenous fistulas. Nippon Hinyokika Gakkai Zasshi 2002;93:615-20.
19. Merkus J, Zeebregts C, Hoitsma A, et al. High incidence of arteriovenous fistula after biopsy of kidney allografts. Br J Surg 1993;80: 310-2.
20. Edsman G. Angiography and suprarenal angiography. Acta Radiol 1957; 155(Suppl):110-3.
21. Palmer JM, Connolly JE. Infrarenal arteriovenous fistula: surgical excision under selective renal hypothermia with kidney survival. J Urol 1966;96:599-605.
22. Boijsen E, Kohler R. Renal arteriovenous fistula. Acta Radiol 1962;57: 433-45.
23. Cosgrove MD, Mendez R, Marrow JW. Branch artery ligation for renal arteriovenous fistula. J Urol 1973;110:632-8.
24. Merkel FK, Sako Y. Surgical treatment for traumatic renal arteriovenous fistulas. Arch Surg 1970;101:438-41.
25. Ehrlich RM. Renal arteriovenous fistula treated by endofistulorrhaphy. Arch Surg 1975;110:1195-8.
26. Merritt BA, Middleton RG. Repair of a huge renal arteriovenous aneurysm with preservation of the kidney. J Urol 1972;107:521-3.
27. Morgan DA, McLoughlin MG. The role of intrarenal vascular reconstruction for arteriovenous fistula. J Urol 1981;125:674-5.
28. Boyce WH. Ultrasonic velocimetry in resection of renal arteriovenous fistula and other intrarenal surgical procedures. J Urol 1981;125:610-3.
29. Stoney RJ, Silane M, Salvatierra O Jr. Ex-vivo renal artery reconstruction. Arch Surg 1978;113:1272-8.
30. Novick AC, Straffon RA, Stewart BH. Surgical management of branch renal artery disease: in situ versus extracorporeal method of repair. J Urol 1980;123:311-6.
31. Hubert JP Jr, Pairolero PC, Kazmier FJ. Solitary renal artery aneurysms. Surgery 1980;88:557-65.
32. Kearse W Jr, Joseph A, Sabanegh E Jr. Transcatheter embolization of large idiopathic renal arteriovenous fistula: case report. J Urol 1994; 151:967-9.
33. Wikholm G, Svendsen P, Herlitz H, et al. Superselective transarterial embolization of renal arteriovenous malformations of cryptogenic origin. Scand J Urol Nephrol 1994;28:29-33.
34. Sprouse LR II, Hamilton IN Jr. The endovascular treatment of a renal arteriovenous fistula: placement of a covered stent. J Vasc Surg 2002; 36:1066-8.
35. Donayre CE. Endovascular treatment of traumatic arteriovenous fistulas and pseudoaneurysms. In: White RA, Forgarty TJ, editors. Peripheral endovascular interventions. St Louis: Mosby-Year Book Inc; 1996. p. 391-6.
36. Rikimaru H, Sato A, Hashizume E, et al. Saccular renal artery aneurysm treated with an autologous vein covered stent. J Vasc Surg 2001;34: 169-71.

Submitted Jul 23, 2004; accepted Dec 6, 2004.