Association of abdominal aortic aneurysm, horseshoe kidneys, and left-sided inferior vena cava: Report of two cases

Marc A. Radermecker, MD, PhD, Hendrik Van Damme, MD, Arnaud Kerzmann, MD, Etienne Creemers, MD, and Raymond Limet, MD, PhD, Liège, Belgium

Surgery for abdominal aortic aneurysm may be challenging when rare renal or venous anomalies are present. This article reports two similar cases of aortic abdominal aneurysm associated with horseshoe kidney and left-sided inferior vena cava treated with a transperitoneal approach. Preoperative knowledge of the anatomic situation enabled appropriate aneurysm repair. Operative strategy is discussed. This report describes an uncommon venous vascular malformation complex and stresses the importance of computed tomography imaging not only in assessing the characteristics of the aneurysmal disease but also in detecting variations in pertinent vascular or parenchymal anatomy. (J Vasc Surg 2008;47:645-8.)

Aortic abdominal aneurysm (AAA) is most often an asymptomatic disease until rupture, which leads to death in 65% of patients.1 The disorder is approximately four times more frequent in men than women, with a prevalence rate between 1.3% and 8.9% depending on population risk factors and age.1 A horseshoe kidney is found in approximately 0.1% of autopsy series2 and in 0.1% to 0.6% of aortic operations.3 The presence of a left-sided inferior vena cava (LIVC) is the least frequent malformation at the level of the IVC and has a reported prevalence of 0% to 0.5%.4,5 Accordingly, the occurrence of AAA with coexistent horseshoe kidney and LIVC is rare and has been described only once.6

Although horseshoe kidneys and anomalies of the retropitoneal veins are usually asymptomatic, they add real complexity to the standard technique of open AAA repair, including the risk of ureteral injury, intraoperative major venous hemorrhage, and renal ischemia.5 Ideally, these anomalies should be recognized preoperatively with contrast-enhanced spiral computed tomography (CT).

This article reports two additional cases of this rare association, with special emphasis on the developmental link between horseshoe kidney and venous vascular malformations. These observations illustrate decision making and surgical management, which are based on two consensual principles. Optimal correction by open5 or endovascular aneurysm repair (EVAR)7 should be provided and minimal loss of renal parenchyma, if any, should be consented.4,8,9

CASE REPORTS

Patient 1. A 64-year-old woman was admitted for treatment of a 55-mm AAA. A CT scan showed a noninflammatory fusiform infrarenal AAA extending to the proximal iliac arteries. A CT angiography (CTA) revealed a horseshoe kidney characterized by a 5-cm isthmus. The IVC was situated to the left side of the AAA and crossed the aorta anteriorly to the right, at the level of the neck of the aneurysm. It received the left renal vein before merging with the right renal vein to form the subhepatic IVC (Fig 1). Both congenital anomalies were asymptomatic incidental findings. The horseshoe kidney was vascularized by a normally positioned right renal artery, two left renal arteries, and a significant aberrant renal artery originating from the right iliac artery, supplying the fused isthmus of the horseshoe kidney.

Surgery was performed with a midline xiphopubic laparotomy. The neck of the aneurysm was partially hidden by the LIVC, which was dissected, mobilized, and retracted laterally to the left. The dissection was extended caudally to reach the upper border of the renal isthmus. The posterior peritoneum was then opened in the midline, carefully avoiding the ureters. The aortic bifurcation was exposed, and the dissection was continued to the lower border of the isthmus. No additional renal artery was found. The LIVC had a retro-isthmic course.

Both femoral arteries were exposed. After heparin administration, the aorta was cross-clamped below the renal arteries and both iliac arteries were ligated. The aneurysm was incised above and below the isthmus. There was no back bleeding from lumbar arteries or the inferior mesenteric artery. A 16 - 8-mm Dacron graft was placed behind the ilium. After proximal anastomosis, the aberrant renal artery was reimplanted into the graft with a Carrel patch technique. The patient’s postoperative recovery was uneventful, and she is doing well 4 years later.

Patient 2. A 69-year-old man consulted for a 51-mm infra-renal aneurysm. The CT incidentally disclosed a coexistent horseshoe kidney and a LIVC. The aneurysm was fusiform and did not involve the bifurcation. A CTA showed two renal arteries at the level of the aneurysm neck. The LIVC crossed the midline at the level of the renal arteries to join the right-sided suprarenal IVC (Fig 2, A). There was no anomaly of the left renal vein. The LIVC was posterior to the renal isthmus. Both ureters were anteriorly located to the isthmus (Fig 2, B). The angulation of the neck was considered as a relative contraindication to stent grafting.
Intraoperative dissections disclosed an accessory renal artery for the isthmus originating at the anterior surface of the aneurysm. After opening the aneurysm, we faced a significant back bleeding from lumbar arteries and the inferior mesenteric artery. These arteries were controlled from inside after lifting up the isthmus. Continuity was restored by the interposition of an 18-mm tube graft in which the aberrant renal artery was reimplanted. The patient’s postoperative course was uneventful. At the 18-month follow-up, the patient is in good general health.

DISCUSSION

Repair of AAA in the presence of renal or venous anomalies is challenging. In the perioperative assessment of AAA, CT scan, CTA, and magnetic resonance angiography are the most useful and reliable investigations, not only to document the extent of the aneurysm but also to rule out renal anatomic anomalies and to assess in detail the renal vascularization and venous anatomy. Intra-arterial angiography in case of aneurysmal disease has nowadays only an indication in view of endovascular aneurysmal stent grafting. Routine arteriography is rarely necessary and has the inherent risk of nephrotoxicity and atheroembolism. Its indication probably remains in horseshoe kidney when aberrant arteries are poorly identified by noninvasive imaging.

A horseshoe kidney is found in 0.25% of the general population and is observed twice as frequently in men as in women.
women. \(^3\) The ectopic kidney is less common, at 0.2% to 0.03%. \(^1\) Both are the consequence of an anomalous renal rotation concomitant to the embryonic ascent from their pelvic position during gestational weeks 4 to 6. In the case of horseshoe kidney, the metanephric buds fuse, which blocks their cephalic migration. Depending on the moment of the dysembryogenesis from the fourth to eighth week of gestation, varying degrees of fusion and incomplete migration may occur.

In most patients, the urinary collecting system is placed anteriorly and can be assessed on preoperative CT scan. \(^12\) Renal vascularization is abnormal in two-thirds of the cases, with accessory renal arteries originating from the abdominal aorta or the iliac arteries. \(^13\) The classification of Crawford et al, \(^14\) which is based on the site of origin of the renal arteries, is useful. In type I, the kidney arises from the normal position, whereas in type III, they all have an ectopic origin. In type II, the vascularization is supplied from two normal renal arteries and one or more accessory renal arteries originating from the aorta or the iliac arteries. An attempt should be made to preserve any significant accessory renal artery. \(^3\) Division of the isthmus should be avoided, considering the inherent risk of urinary leakage. \(^15\)

The IVC is formed from a complex network of three parallel pairs of embryologic veins—posterior cardinal, subcardinal, and supracardinal veins—that anastomose among themselves (retroaortic anastomotic channels) and later undergo partial regression. The right supracardinal vein provides most of the infrarenal IVC, whereas the posterior cardinal veins almost involute and only provide the iliac veins. \(^10,16\) Faults in this development will result in five anomalies of the IVC and left renal vein that are relevant to abdominal aortic surgery: duplication of the IVC (0.5% to 1%), persistence of normally involuted left supracardinal vein, \(^4\) preaortic confluence of the iliac veins, transposition of the IVC namely LIVC (0.5%), \(^6,11\) retroaortic left renal vein (1.5% to 3.7%), \(^4,5,11\) and circumaortic renal vein collar or duplication of the left renal vein (6%). \(^4,5,10,11\)

Because the embryogenesis of the renal parenchyma and its venous drainage in the IVC occur simultaneously from gestational weeks 4 to 10, it is plausible that horseshoe kidney (anomalous embryonic renal rotation and ascent) and LIVC (anomalous involution of right supracardinal vein) are the consequence of a shared disturbed signal during development of these retroperitoneal structures. Another explanation could be that horseshoe kidney and its related failure of cephalic ascent and abnormal rotation has a direct impact on the locoregional development of the complex venous cardiac network. Compression of segments of cardinal veins by the abnormal kidney mass could induce their regression and involution. This explanation would account for the different variants of IVC anomalies reported so far in horseshoe kidney, such as duplication of the IVC, LIVC, and preaortic right IVC.

The present association of horseshoe kidney and LIVC has been reported only three times. \(^4,17,18\) Boyd\(^{17}\) first reported the anatomic association in 1931. Billiet et al\(^{18}\) documented the association of horseshoe kidney and LIVC in a case of right hydronephrosis on a pelvic junction stenosis that was operated on. The only case associated with an AAA was reported by Sonneveld et al\(^9\) and was successfully operated on by the transperitoneal open approach, with reimplantation of a significant anomalous artery for the isthmus.

A surgical transperitoneal approach was used in the two cases reported here because of significant aberrant arteries and proved to be efficient and safe. A left retroperitoneal access has been advocated for endovascular aneurysmal grafting in case of associated horseshoe kidney and is certainly useful in case of “pancake kidneys,” because it prevents complications at the level of the isthmus. \(^19,20\)

We, however, favored the transperitoneal route in this combined pathology. This access usually permits direct control of the proximal aorta and adequate exposure of the anterior aspect of the aneurysm to evidence short aberrant renal arteries supplying the isthmus, which have been missed on preoperative screening. \(^3,6\) In our particular patients, the LIVC would have been a barrier to exposure of the aneurysm of the aorta by a left retroperitoneal approach. Some authors have reported a right retroperitoneal approach as an alternative when left retroperitoneal route is contraindicated by the presence of LIVC, retroperitoneal fibrosis, or colostomy. \(^21\)

The presence of a LIVC has been considered a formidable challenge. \(^22-24\) The LIVC in the absence of situs inversus is a mirror image of a normal IVC: it receives the left gonadal veins and lies on the left lateral wall of the aorta. As it courses cranially, it crosses anteriorly the aorta at the level of the renal arteries and reaches, after receiving both renal veins, its right-sided subhepatic location. In this subhepatic setting, dissection of the lower and medial borders of the LIVC and gentle upward and leftward retraction achieve sufficient exposure of the neck of the aneurysm. In case of poor exposure, division of the proximal right renal vein is conceivable to facilitate exposure of the underlying aneurysmal neck because venous drainage of the right kidney will be supplied by gonadal and adrenal veins. \(^5,23\)

The ureters are at risk during the transperitoneal approach. Their course, as predicted by embryology, is anterior to the horseshoe kidney isthmus, as illustrated in Fig 1. This fact implies strictly median and careful dissection of the aortic bifurcation. The insertion of a ureteral JJ catheter may be exceptionally indicated when section of the horseshoe kidney isthmus is anticipated. \(^7\)

In complex and difficult situations, endovascular treatment may be proposed after considering the usual feasibility criteria and the anatomic variations of renal vascularization. Crawford type I can be suitable for stent grafting, whereas type III should be viewed as a contraindication. In Crawford type II, stent grafting may be undertaken if the patient is not an appropriate candidate for open repair and minor parenchymal loss is anticipated. Current evidence suggests that vessels ≤3.0 mm can be sacrificed in the presence of two main renal arteries and normal kidney function. \(^9\) When a more liberal definition of accessory vessel is adopted, \(^25\) segmental renal infarction occurs in
21% of the cases with unknown impact on long-term renal function.

CONCLUSION

This report discusses the surgical management of a rare association of AAA with a malformed complex comprising horseshoe kidney and LIVC. This congenital anomaly is described and debated within the framework of the classical renal and venous malformations. The importance of preoperative diagnosis and investigation with CT is stressed, and decisive arguments for transperitoneal grafting are given. Awareness of such venous and renal anomalies is crucial to reduce the risk of inadvertent intraoperative venous or ureteral injury.10,11

REFERENCES