Retroperitoneoscopic Nephrectomy for a Horseshoe Kidney with Unilateral Severe Hydronephrosis and Ureteral Hypoplasia

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A horseshoe kidney is the most common renal fusion anomaly. It is well known that horseshoe kidneys may be associated with many urological problems, including calculi, vesicoureteral reflux, and ureteropelvic junction obstruction. However, a horseshoe kidney with unilateral severe hydronephrosis and ureteral hypoplasia is very rare. We report an 11-year-old female who underwent a retroperitoneoscopic nephrectomy for a horseshoe kidney with severe hydronephrosis and unilateral ureteral hypoplasia. **(Korean J Urol 2009;50:512-515)**

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A horseshoe kidney is the most common renal fusion anomaly. Several associated anomalies, including ureteropelvic junction obstruction, vesicoureteral reflux, and a duplicated ureter, can occur in patients with this anomaly. The common presenting symptoms are urolithiasis, infection, and hydronephrosis.[1] However, a horseshoe kidney with unilateral severe hydronephrosis and ureteral hypoplasia is very rare. Here we report an 11-year-old female who underwent a retroperitoneoscopic nephrectomy for a horseshoe kidney with severe hydronephrosis and unilateral ureteral hypoplasia.

CASE REPORT

An 11-year-old female presented with left flank pain and gross hematuria after a fall. Her past medical and family history were nonspecific. There was a distended, palpable mass and direct tenderness in the left upper quadrant on the physical examination. There were no abnormal findings on the blood tests; however, there were many red blood cells on the urinalysis. Computed tomography imaging showed a horseshoe kidney with severe hydronephrosis and renal cortical thinning of the left kidney (Fig. 1A, B). A MAG3 renal scan confirmed the left kidney function to have a 24% decrease in uptake (Fig. 1C). The cystoscopy findings were normal. One day before surgery, we performed a percutaneous nephrostomy for reduction of the hydronephrosis, and 4,000 ml of brownish fluid was drained (Fig. 1D).

The patient was placed in the lateral position, and a lateral incision was made longitudinally at 2 cm below the left 12th rib; the nephroscope was inserted with an attached surgical glove at the tip; 800 ml of saline was injected; and the required space was secured for the surgical procedure. After inserting a 12 mm Hasson trocar, CO_2 gas was perfused to form a pneumoperitoneum, and then a flexible camera was inserted. In addition, 5 mm trocars were inserted at the anterior and posterior axillary lines, at the level of the umbilicus, after examination of the surrounding structures. The position of the previously placed nephrostomy catheter was confirmed, and we then opened Gerota's fascia, dissected the kidney from the surrounding tissues, and then dissected by the psoas muscle, noting the thread-like hypoplastic ureter (Fig. 2A-C). A total of 3 arteries and veins were clipped, and the isthmus was incised by using electrocautery and the Sonosurg® (Olympus, Japan) for the resection of the left kidney (Fig. 2D, E). The resected left kidney was removed from the body in a LapBag® (Sejong Medical, Korea) through a 12 mm Hasson trocar (Fig.

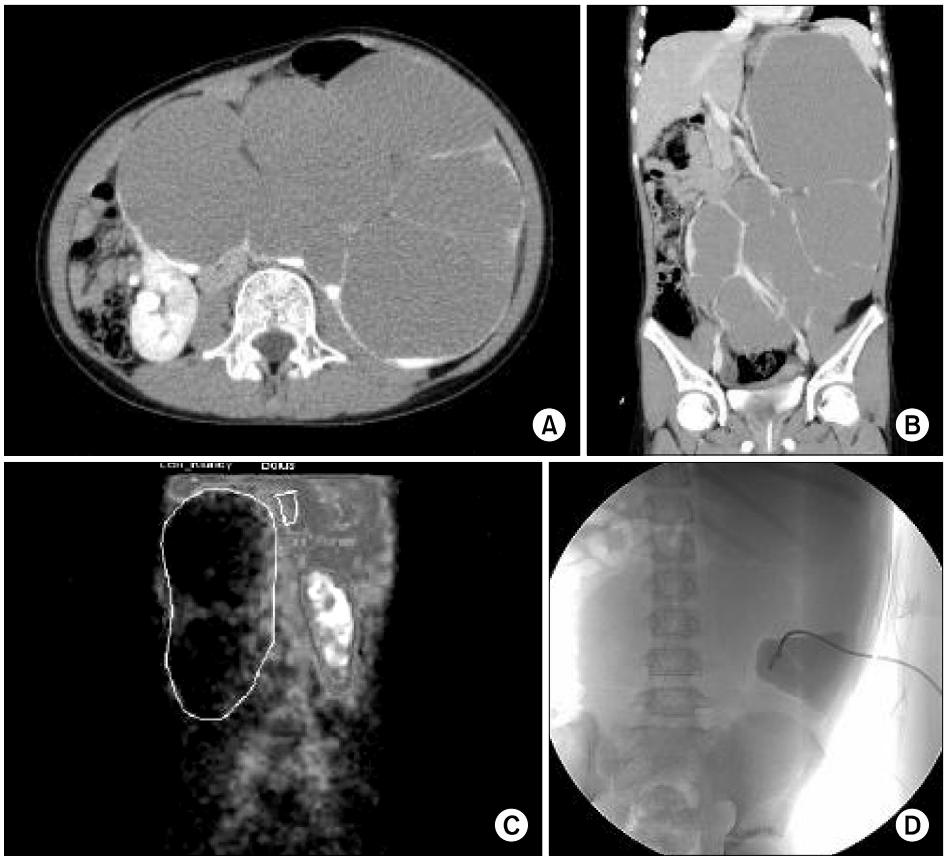

Fig. 1. Imaging studies. (A) and (B) enhanced computed tomography scan, transverse and coronary images, showing a horseshoe kidney with very large hydronephrosis of the left kidney. (C) The 99mTc-MAG3 renal scan showed decreased renal uptake. (D) Insertion of a percutaneous nephrostomy was performed to reduce the size of the hydronephrosis and mark the site so that it could be easily seen during the operation.

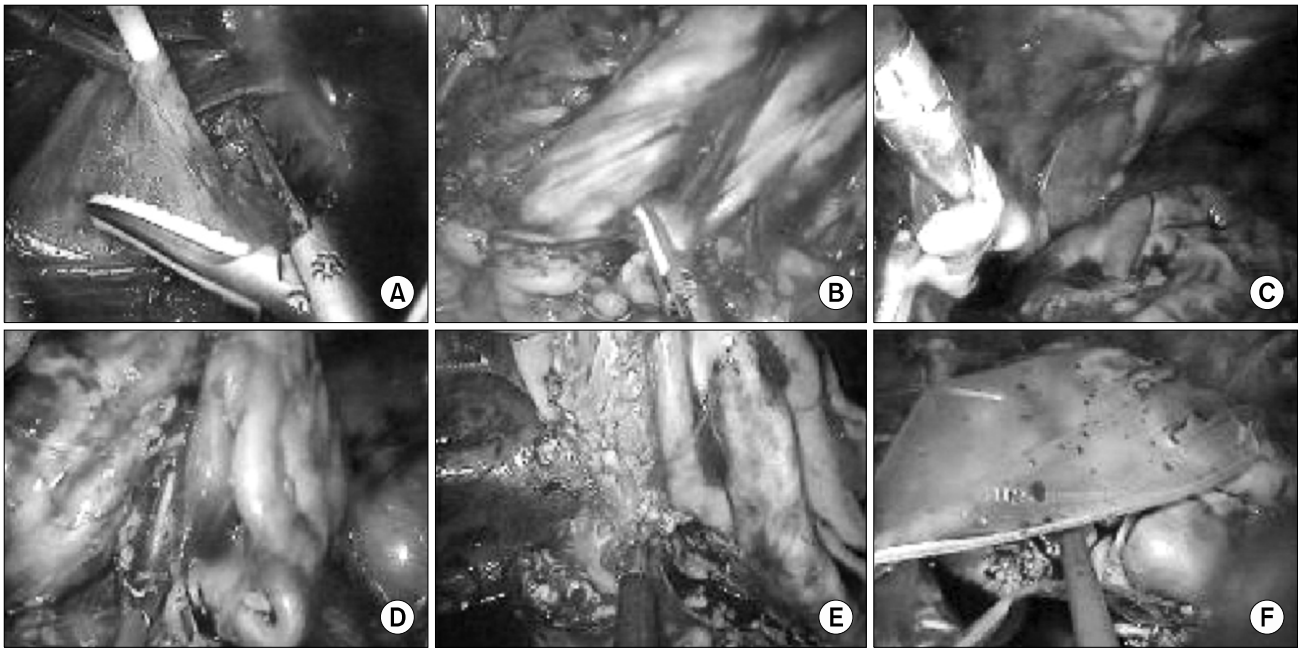

Fig. 2. Intraoperative retroperitoneoscopic nephrectomy procedure for a horseshoe kidney with severe hydronephrosis due to unilateral ureteral hypoplasia. (A) The percutaneous nephrostomy tube was checked. (B) The Gerota's fascia was opened and the kidney was freed. (C) Thread-like hypoplasia of the ureter was seen. (D) Renal arteries and veins were clipped and cut. (E) The isthmus was divided by ultrasonic scissors (Sonosurg®). (F) The specimen was put into a LapSac® and extracted.

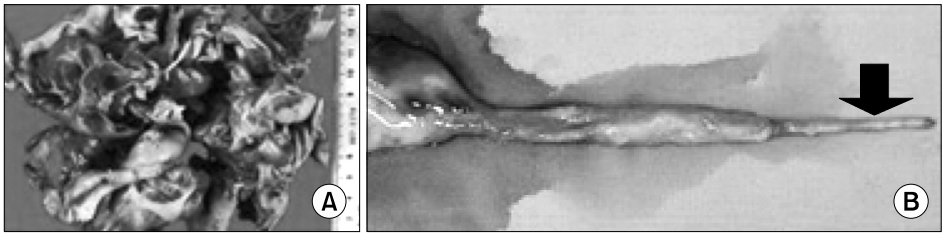

Fig. 3. Gross findings. (A) Excised kidney showing severe hydronephrosis. (B) Distal ureteral hypoplasia (arrow).

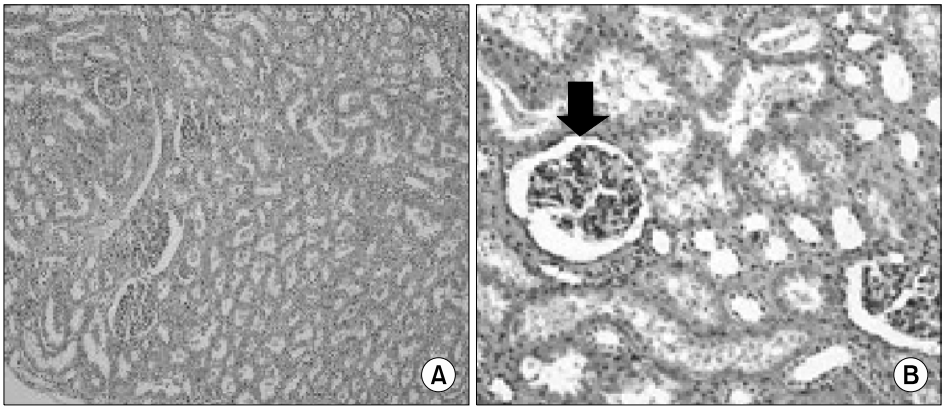

Fig. 4. Pathologic findings. (A) H&E, x100, Nonfunctioning kidney with chronic inflammation. (B) H&E, x200, A few normal nephrons (arrow).

2F). The procedure was completed after inserting a Jackson-Pratt drainage tube, removing the trocar, and suturing all of the incised parts. The surgical procedure took 70 minutes, and the estimated blood loss was less than 50 ml. The cortex was hardly observed by gross examination of the resected left kidney; a hypoplastic ureter that was expanded in the proximal area and tapered distally was noted (Fig. 3). Pathological evaluation showed that the shape of the nephrons was normal; however, the kidney had chronic inflammatory hypoplasia with decreased overall numbers of nephrons (Fig. 4). There were no pre- or postoperative complications. Oral feeding and ambulation were started on day 1, the drainage tube was removed on day 4, and the patient was discharged on day 7. One week and 6 months after the procedure, the ultrasound follow-up showed no complications.

DISCUSSION

A horseshoe kidney is the most common congenital renal fusion disorder, with a prevalence of approximately 1 out of 400. Vesicoureteral reflux, ureteropelvic junction obstruction, and ureteral duplication are commonly associated malformations that cause complications such as hydronephrosis, infection, and calculus. Hydronephrosis occurs more frequently in a horseshoe kidney than in a normal kidney. According to a report by Shimkus and Mekhanna,[2] 64% of patients had hydronephrosis among 170 cases of horseshoe kidneys. Hydronephrosis is common with a horseshoe kidney because the ureter is connected to the upper pelvis of the kidney and passes in front of the isthmus, and the development of the vessels is very dynamic. Although most horseshoe kidneys are asymptomatic, urolithiasis and ureteropelvic junction obstruction represent the most common complications requiring surgical intervention. Riedl et al[3] reported in 1995 that laparoscopic nephrectomy in patients with a horseshoe kidney was a safe and effective minimally invasive surgical option. However, the laparoscopic surgical technique is difficult because of the abnormal vessel distribution, abnormal kidney location, and isthmus; therefore, transperitoneal surgical procedures are preferred.[4] On the other hand, the retroperitoneoscopic procedure can directly approach the retroperitoneal organs, similar to open surgery, and is preferred in patients who have had previous intraperitoneal surgery or peritonitis. In addition, the risk of injury to other organs such as the intestines is reduced, and fewer trocars are needed because the liver, spleen, and intestines do not have to be pulled away for the dissection. Furthermore, the recovery is much faster because of the absence of the development of intestinal adhesions in the peritoneum.[5] Hence, we selected the retroperitoneoscope method for the procedure in the patient

reported here. The surgery was performed with only 3 trocars and the isthmus could be managed with electrocautery and Sonosurg® (Olympus, Japan) without bleeding. The patient recovered quickly and started an oral diet and ambulation on day 1, and the drainage tube was removed on day 4.

The presence of a hypoplastic ureter is very rare and on the developmental spectrum is considered to be between ureteropelvic junction obstruction and polycystic dysplastic kidneys. In patients with a hypoplastic ureter, the ureter does not transport urine, which is the main function of the ureter; urine obstruction occurs during early development in the fetus. The extent and timing of obstruction are critical factors in determining the function of the kidneys. For example, the occurrence of obstruction in earlier stages of development in the fetus tends to be associated with dysplastic kidneys, whereas the occurrence during later stages results in simple fetal hydronephrosis.[6,7] Therefore, a hypoplastic ureter may be associated with a wide range of renal pathologies. Frozen section biopsy of the kidney might help to determine the associated renal pathology; however, in most cases, a nephrectomy will be the best option.[8]

In a case report by Allen and Husmann[8] on 3 children with ureteropelvic junction obstruction and hypoplastic ureter, all had fiber-like thin hypoplastic ureters; an Anderson-Hynes pyeloplasty was performed after expansion of the ureter with a lacrimal probe and insertion of a thin silicon ureteral stent. In addition, saline was injected from time to time to facilitate peristalsis of the ureter. However, peristalsis of the ureter was not present in these cases, and the obstruction persisted, resulting in a nephrectomy.

In conclusion, a horseshoe kidney with severe hydronephrosis was found in the patient reported here; however, the computed tomography did not detect the hypoplastic ureter. The function of the left kidney was decreased by 24% on the renal scan. Therefore, a retroperitoneoscopic nephrectomy was successfully performed.

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