Renal agenesis associated with ipsilateral ectopic ureter entering a large seminal vesicle cyst

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We describe a case of right renal agenesis with ipsilateral ectopic ureter opening into a large seminal vesicle cyst in a 24-year old man. The patient presented with left flank pain and the imaging studies showed absent right kidney and a large pelvic cyst possibly arising from seminal vesicle. The cyst was removed together with the right ureter and kidney remnant through a laparotomy.

Key Words: renal agenesis, ectopic ureter, seminal vesicle cyst, congenital anomaly

Introduction

Renal agenesis and ipsilateral ectopic ureter associated with seminal vesicle cyst is an extremely rare congenital anomaly. Most of the cases become symptomatic after puberty due to accumulation of seminal fluid in the obstructed seminal vesicle causing cystic formation.[1,2] The distended cyst may cause abdominal, perineal, testicular pain or painful ejaculation. It also may lead to bladder irritation and storage symptoms. The potential complications include infertility, epididymitis, prostatitis and malignant degeneration.[2-5] We report a case of renal agenesis with ipsilateral ectopic ureter opening into a huge seminal vesicle cyst.

Case report

A 24-year-old patient presented with discomfort in the left flank and gluteal regions with no lower urinary symptoms. On examination there was a large palpable cystic mass in the lower abdomen. Abdominal ultrasound revealed absence of right kidney with normal left kidney. There was a huge pelvic cyst on the right side measuring 10 cm x 15 cm x 10 cm and contained a fluid volume of 914 ml. Magnetic resonant imaging showed a large cyst suggestive of a seminal

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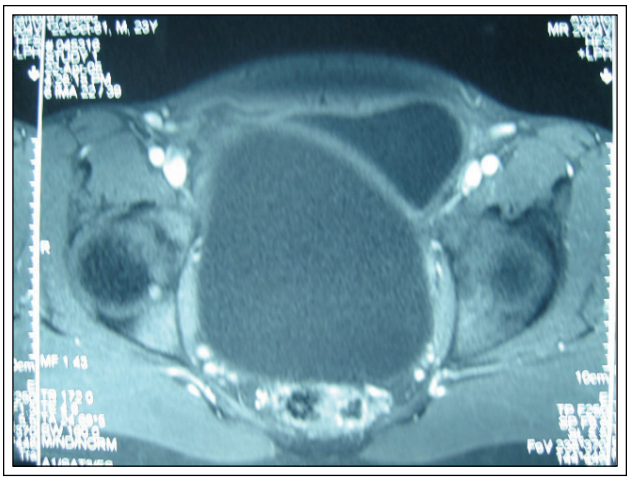

Figure 1. Pelvic MRI revealed large seminal vesicle cyst compressing the urinary bladder.

vesicle cyst associated with renal agenesis, Figure 1. Cystoscopy revealed absence of the right ureteral orifice with displaced bladder by extravesical cyst.

The patient underwent laparotomy and via a midline incision, the seminal vesicle cyst was dissected, Figure 2. The cyst was quite large occupying the patient's whole pelvis. Microscopic examination of aspirated cystic fluid showed plenty of dead sperm confirming the diagnosis of a seminal vesicle cyst, Figure 3. At certain areas the peritoneum was adherent to the cyst, so we decided to open the peritoneum to complete the dissection. The dissection was completed all around the cyst reaching to the level of the prostate and contralateral seminal vesicle. The right vas deferens was followed and was ending in the cyst with no ejaculatory duct on

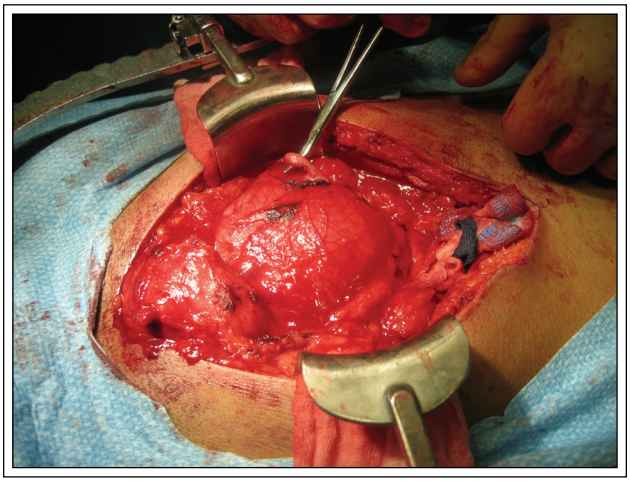

Figure 2. Operative view of the vas deferens opening into the large seminal vesicle cyst.

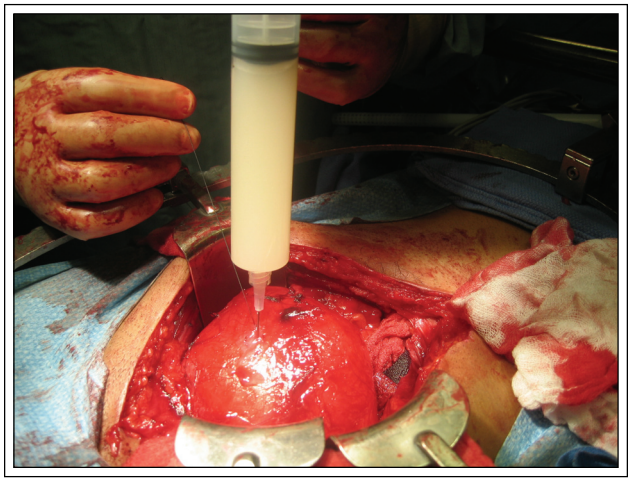

Figure 3. Seminal fluid aspirated from the cyst.

this side. The ectopic ureter was identified on the right side and dissected upwards till its end in small cystic structure, which corresponded to the ipsilateral renal agenesis. The ectopic ureter and the seminal vesicle cyst was excised and sent to pathology, Figure 4. Operative time was 3 hours and 15 minutes, blood loss was 100 cc and the hospital stay was 3 days. The postoperative course was uneventful and the pathological examination showed cystic mass weighing 178 gm. The histopathological examination showed a benign fibromuscular wall lined by simple cuboidal epithelium. The ureter was lined by benign urothelium and terminates in cystic dilatation lined by simple flattened epithelium with no fetal or mature renal tissue. At 6 months follow-up the patient was asymptomatic, and pelvic ultrasound was normal.

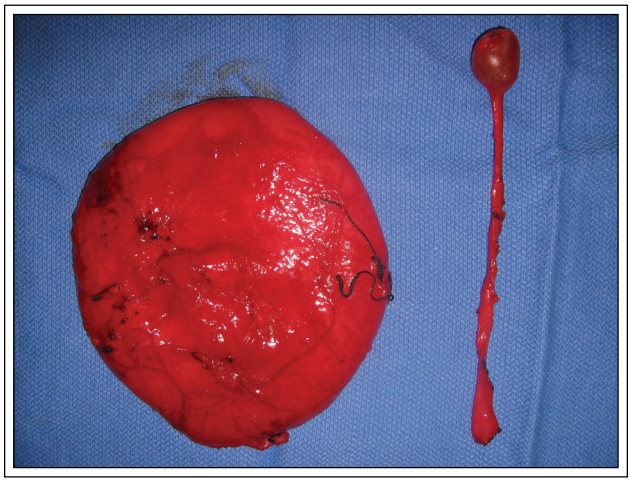

Figure 4. Surgical specimen of the right ectopic ureter with cystic blind end and the cystic seminal vesicle.

Discussion

The embryogenesis of the urinary and reproductive systems is closely related. The ureter arises from the caudal end of the mesonephric duct between the 4th and 7th week of pregnancy. The normal growth of ureteral bud plays the key role in the normal development of the kidney and other Wolffian duct-derived organs. The normal development of the kidney occurs when the ureter meet the primitive kidney in its central portion. The differential growth of the mesonephric duct and the urogenital sinus leads to normal opening of the ureter into the bladder. If the ureteric bud originates too cranially, it results in delayed connection to the urogenital sinus and subsequently ectopic ureteral opening in any structure that arises from the mesonephric duct such as seminal vesicles, ejaculatory duct or vas deferens. In addition, the imperfect development of the ureteric bud results in renal dysplasia or agenesis.[4] The clinical presentation of this congenital malformation is not consistent and the symptoms are not specific and usually related to the size of the cyst, leading to difficult diagnosis.[6] Physical examination may demonstrate normal findings or evidence of chronic epididymitis or palpable cystic mass on digital rectal examination.[4] Laboratory investigation is usually normal, but abnormal semen analysis may be observed in 10% of cases. Abdominopelvic ultrasound and CT are the most useful diagnostic tool in patients in whom such a malformation is suspected.[6,7] Other studies such as excretory urography, MRI and seminovesiculography may be selectively needed for differential diagnosis. The MRI is very sensitive and it can establish a definitive diagnosis .[8] Cystoscopy is necessary to document the presence of a hemitrigone and the presence or absence of a ureteric orifice. The treatment of these cases is indicated in symptomatic patients. The treatment option includes simple cyst aspiration and transvesical drainage, which are associated with risk of recurrence and possible infection.[9]

Laparoscopic approach to the seminal vesicle is minimally invasive and provides excellent visualization and short hospital stay.[4,10] Laparoscopic approach provides meticulous control of blood vessels and clean dissection of the seminal vesicles from the bladder and the prostate. Transperitoneal laparoscopic access is achieved through four ports including the camera placed at the umbilicus, a 12 mm ports at the right lateral border of the rectus muscle, 5 mm port at left lateral border of the rectus and 5 mm port in the midline suprapubic area.[4] The retrovesical peritoneum is opened and the seminal vesicle is dissected to the prostate where it is clipped and divided with the vas deferens. The ureter is dissected cephaled after mobilization of the colon. Then, the seminal vesicle cyst, and the ureter which may terminates blindly or with dysplastic kidney removed altogether en bloc. In our case, we chose open surgery because the diagnosis was uncertain even with MRI, and we do not know how much adhesions surrounded the cyst. Also the cyst was large and abdominally palpable and was easily accessible through open surgery. Surgical excision of the cyst through retropubic, transvesical, transperitoneal or perineal approach is possible surgical options. The aim of the surgery is to remove the cyst and preserve the sexual potency and fertility. Due to the deep pelvic location of the seminal vesicle open surgery may carry the risk of morbidity giving an advantage to laparoscopic surgery, which has minimal postoperative complications. In our case, transperitoneal open surgery allows good exposure and dissection of the seminal vesicle without complications.

Conclusions

This case illustrates a rare congenital anomaly of renal agenesis associated with ectopic ureter opening into a large seminal vesicle cyst which was treated by open surgery. In certain cases the diagnosis is not easy. Surgical excision is the treatment of choice when the patient is symptomatic. Recently most of the authors proposed a laparoscopic approach as a treatment of choice for these cases but in our case open surgery was preferred due to uncertain diagnosis and possibility of adhesions that could make the laparoscopic approach difficult. □

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