Giant hydronephrosis complicated by multiple uroepithelial carcinomas

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ABSTRACT
Giant hydronephrosis is uncommon, and malignant hydronephrosis of unknown origin is even rarer. We report a case of a 43-year-old male patient with giant hydronephrosis without painless carnivorous hematuria. Preoperative imaging and urinary search for exfoliated cells revealed no evidence of renal pelvic cancer. The patient underwent simple nephrectomy and pathology confirmed the diagnosis of multifocal high-grade papillary uroepithelial carcinoma of the renal pelvis.

1. Introduction
Giant hydronephrosis (GH) is variably defined as a kidney containing greater than 1000 ml of urine in its collecting system, or a kidney that accounts for more than 1.6% of total body weight. Giant hydronephrosis with concurrent malignancy is less common clinically and is easily misdiagnosed. We report here a case of Giant hydronephrosis found incidentally with multiple foci of Migratory cell carcinoma and review the current literature.

2. Case report
A 43-year-old man had an incidental finding of a large right abdominal mass with hypothermia, anemia, and hypoalbuminemia without hematuria, lumbar pain, bladder irritation, and weight loss. Other clinical examinations and laboratory findings were within normal limits. Physical examination showed a cystic mass in the right upper abdomen and right lumbar region, which was soft, smooth, pushable, and painless to touch, with percussion pain in the right rib cage. Ultrasound showed right hydronephrosis, measuring approximately 27.0*14.9 cm. Abdominal CT showed multiple cystic hypodense shadows in the right kidney, measuring about 21.5*15.0 cm (Fig. 1). The patient’s creatinine clearance was 0.11 ml/min in the right kidney and 74.59 ml/min in the left kidney. A total of approximately 4200mL of coffee-colored fluid was drained by ultrasound-guided percutaneous puncture of the right nephrostomy. Excretory urography showed no visualization of the right kidney and ureter, and the left kidney was normal (Fig. 2).

The patient’s right kidney was non-functional so a simple nephrectomy was performed under general anesthesia. The mass of the specimen was about 2500g, the size was about 17.0*12.0*4.0cm, the section was multi-cystic dilated, the cortex was thin, the thickness was 0.2–0.5cm, the inner wall has seen multi-focal papilla-like area, the maximum extent was about 3cm*3cm. Histopathological examination using a light microscope with 100x magnification with hematoxylin staining revealed High-grade multifocal papillary uroepithelial carcinoma of the renal pelvis (grade II-III) with the focal invasion of the lamina propria and highly atrophic changes in the surrounding renal parenchyma, with no cancerous invasion of the ureteral cutaneous margin (Fig. 3).

3. Discussion
More than 600 cases of GH have been reported worldwide. Giant hydronephrosis is most commonly seen in children and rarely in adults. Its location is mostly orthotopic, but cases of the ectopic kidney with giant hydronephrosis have been reported. Patients may be relatively asymptomatic or present with symptoms such as chronic back pain, hematuria, bulging abdominal masses, urinary tract infections, pyelonephritis, and renal insufficiency.

The most common congenital cause of giant hydronephrosis is obstruction of the ureteropelvic junction (UPJO). In rare cases, exogenous compression of the UPJO due to ureteral ectasia, duplicated collecting system and anomalous vascular system may result in the formation of a giant hydronephrosis. In adults, acquired disease is more common, namely ureteral stones, trauma, cancer or retroperitoneal fibrosis. Obstruction due to congenital etiology is less common and the
effusion progresses more slowly, so it can reach larger volumes. Potential complications of long-standing giant hydronephrosis include renal failure, hypertension, malignancy, and rupture. In this case, no tumor or stone was found intraoperatively at the renal pelvis-ureteral junction, but rather showed congenital pathological changes such as stenosis, fibroplasia, and fibrosis. Murai et al. suggested that the development of renal pelvic cancer is the result of the carcinogenic effect on the mucosa caused by the long-term retention of carcinogenic urine in the renal pelvis. Secondly, the stimulation of long-term chronic infection can also lead to the development of carcinogenesis.

Ultrasonography is usually the first-line test for massive hydronephrosis, followed by CT and MRI. Excretory urography, retrograde pyelogram and angiography can be used to determine the cause and assess renal function. Giant hydronephrosis combined with renal pelvic cancer mainly presents as an abdominal mass and lacks typical symptoms such as intermittent painless meatus hematuria. Meanwhile, giant hydronephrosis in adults is often confused with large cystic abdominal masses such as intra-abdominal or retroperitoneal cysts, pseudomucinous tumors, pancreatic pseudocysts, ovarian cysts or renal tumors, and therefore has a high rate of misdiagnosis and missed diagnosis in clinical practice. The severity of residual renal function and symptoms determines whether the clinician performs nephrectomy or pyeloplasty. However, in cases where the patient is compromised, percutaneous renal puncture drainage may be performed as a prior or definitive treatment to avoid alterations in hemodynamic balance secondary to sudden abdominal decompression. But if there is a possibility of concurrent malignancy, it is more advantageous to perform nephrectomy with ureterectomy.

4. Conclusion

The prognosis of giant hydronephrosis complicated by urothelial carcinoma montanus is poor and the mortality rate is high. Therefore, when diagnosing giant hydronephrosis, we should be alert to the possibility of malignant lesions, and take a further medical history, improve ultrasound, urine or peel aspiration urine cytology, and CT, and other comprehensive examinations. Kuromoto A et al. concluded that PET-CT can be used to diagnose non-functioning kidneys with concurrent urothelial malignancies. If necessary, careful intraoperative exploration and rapid intraoperative pathological sections can be performed to clarify the diagnosis. Adjuvant postoperative radiotherapy is beneficial for prolonging survival.

Informed consent form

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Declaration of competing interest

The authors has no conflict of interest.

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