Ectopic immature renal tissue in gubernaculum associated with undescended testis of a 1-year-old child
A case report
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Abstract
Rationale: Ectopic immature renal tissue (EIRT) is extremely rare in congenital malformations. Moreover, the fundamental pathogenesis of EIRT is still unclear and controversial.

Patient concerns: The right scrotum of a 1-year-old man was found empty for a period of 1 month. B-ultrasound revealed normal bilateral kidneys and a hypoechoic nodule in the right groin.

Diagnoses: Based on B-ultrasonography, surgery and pathological examination, we concluded a case of abnormally located and EIRT in the inguinal canal.

Interventions: After pathological diagnosis, the patient was not treated with drugs.

Outcomes: One year after the operation, the patient recovered.

Lessons: EIRT in gubernaculum is extremely rare. Because of the potential risk of malignant transformation, it is necessary to diagnose and treat it early.

Abbreviations: CD = cluster differentiation, CgA = chromogranin A, CK = cytokeratin, EIRT = ectopic immature renal tissue, HE = hematoxylin-eosin staining, Ki67 = nuclear associated antigen Ki-67, PLAP = placental alkaline phosphatase, Syn = synuclein, WT-1 = Wilm’s tumor protein-1.

Keywords: ectopic immature renal tissue, inguinal canal, metanephric tissue, nephroblastoma, teratoma

1. Introduction
Ectopic immature renal tissue (EIRT) is a metanephric nephrogenic rest found at an extrarenal site.[1] EIRT is also known as ectopic, heterotopic, or extrarenal nephrogenic rest or extrarenal nephroblastomatosis.[2] EIRT occurs primarily due to the failure of kidneys to occupy the normal position in the waist. EIRT is often associated with other malformations, such as skeletal abnormalities, heart valve insufficiency, undescended testicle, and hypospadias in the urogenital system. EIRT occurs mostly in the pelvic cavity resulting in the formation of pelvic kidney.[3,4]

We report a case of 1-year-old man who was diagnosed with EIRT in his inguinal canal. The case illustrates the scope for further research into EIRT including the embryonic and pathological aspects.

2. Case report
The right scrotum of a 1-year-old man was found empty for a period of 1 month. He had a healthy medical history and normal prenatal examination. His parents were healthy, without any family history of genetic disorders. A detailed examination upon admission to the hospital revealed an empty right scrotum; the state of native kidney and renal function are all normal; a nonpalpable testicle, and a palpable tumor in the right inguinal region, measuring 1.2 × 0.5 cm. The tumor was hard and was neither retracted into the abdominal cavity, nor pushed into the scrotum. However, a palpable testis was noticed in the left scrotum, with a fairly developed penis. B-ultrasonography revealed normal bilateral kidneys and a hypoechoic nodule in the right groin with dimensions of 11 × 5 mm. The nodule was identified as an undescended testicle on the right side (Fig. 1). A slightly soft right testis, with dimensions of 1 × 0.5 cm, and mildly separated from epididymis was found during surgery. The ductus deferens was developed fairly and the gubernaculum adhered to aponeurosis of oblique externus abdominis. About 2 cm away from the testis, a solid mass measuring 0.8 × 0.5 cm in size was detected on the gubernaculum testis, with an unclear border. Informed content of patient was obtained. This study was content informed and approved by the local Human Research Ethics Committee in Affiliated Hospital of Nantong University.

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Pathological examination confirmed a solid tumor mass of gray–white tissue measuring $2 \times 1.5 \times 1$ cm without a clear coating, and containing a white-to-tan color cut surface with proper hardness. Microscopically, a few small tube-like structures were found in the glomerulus. The size of cells was not pleomorphic and the mitotic count was zero. The cells contained a small psammoma body. No teratomas or blastemas were found (Fig. 2). Immunohistochemical assays showed positive expression of Wilm’s tumor protein-1, cytokeratin 7, cluster differentiation 10, paired box gene 8, Vimentin and 2% of nuclear-associated antigen Ki-67, and tested negative for cytokeratin 20, synuclein, chromogranin A, cluster differentiation 99, cluster differentiation 117, and placental alkaline phosphatase (Fig. 3).

Based on all the test results, we concluded a case of abnormally located and EIRT in the inguinal canal.

3. Discussion

The kidneys originate in the metanephric tissues of the sacral region.\(^5\) During later embryonic developmental stages, the kidneys usually climb to the waist and develop separately from the abdominal wall. Failure to climb into the waist region results in the development of renal ectopia.\(^6\) Ectopic kidneys are often associated with other malformations, such as skeletal abnormalities, heart valve insufficiency, undescended testicle, and hypospadias in the urogenital system.\(^7\) Normally, during the 7th to 8th month of intrauterine development, the testis is surrounded a double-layered peritoneum, and descends into the scrotum through the inguinal canal. In this case, the patient carried an undescended testicle. We believe that the development of ectopic kidney in the inguinal canal is due to incomplete separation of the metanephric and paramesonephric ducts from the gonads, resulting in remnants of partially developed kidney tissues in the gubernaculum testis. Such cases are rare. However, ectopic kidney tissues in the undescended testicle are related to nephroblastoma outside the kidney.\(^8\) Nephroblastoma originates in the metanephric blastema. However, the origin of extrarenal nephroblastoma is still controversial. A few studies suggest that renal ectopia may be anterior portion of nephroblastoma.\(^9\) Other studies suggest that extrarenal nephroblastoma is derived from metanephric blastema or residual mesonephros.\(^10\) On the other hand, a few researchers also consider teratoma-containing kidney tissues as originating in extrarenal nephroblastoma.\(^8\)

EIRT might occur in the retroperitoneum, sacrococcyx, and inguinal canal.\(^11\) The origin of residual tissues in ectopic kidneys is not clear. The 3 components including mesonephros, metanephros, and teratoma contribute to EIRT. McDougall et al.\(^11\) reported a patient with undescended testicle and ectopic renal tissues in his inguinal canal. He believed that these tissues were derived from mesonephros and not metanephros, without any relationship with the ureter. A similar case was also reported by Shono et al.,\(^8\) who proposed that the remnants of mesonephric duct intrude into the inguinal canal, and grow
along with the testis and spermatic cord. Saito et al.\textsuperscript{13} had argued that tissues derived from both mesonephros and metanephros were left in the inguinal canal during the growth of testis and spermatic cord. An extensive review of the literature showed occurrence of EIRT both associated and unassociated with a teratoma.\textsuperscript{11,11} Suspicious or obvious features of EIRT malignancy have in fact been reported mainly within teratomas, and treated with additional surgery and chemotherapy. On the contrary, some patients unassociated with teratoma had recurrence. They were still alive without adjuvant chemotherapy.\textsuperscript{14}

These rare cases need to be distinguished from other lesions. For example, in metanephric adenoma, benign tumors are generally seen in the young as well as middle-aged women. Microscopically, the metanephric adenoma is composed of fine tubular structures and papillary glands with limited mesenchyme. The overall morphological characteristics suggest that the metanephric adenoma originates in renal tubular epithelial cells. Sertoli trophoblastic tumor and tumor-like lesions are diagnosed by the presence of tubular structures, suggesting a lining of elongated cells resembling Sertoli cells.

In summary, Patients carrying undescended testicles may also contain EIRT in their inguinal canal. Such cases are very rare. However, EIRT developed toward extrarenal nephroblastoma, especially when they contain metanephric tissues and teratomas. These tissues require clear excision, followed by regular monitoring postoperatively.

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