



A Case of Primary Paratesticular Wilms Tumor in an Undescended Testis

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Extrarenal Wilms tumor is rare. We describe the first reported case of primary paratesticular extrarenal Wilms tumor with regional metastasis in 8-month old male with left undescended testis. Patient underwent left radical orchiectomy with regional lymph node excision. The inguinal node and paratesticular mass demonstrated the classic Wilms triphasic pattern, stained positively for WT-1 and demonstrated no loss of heterozygosity of chromosomes 1p and 16q. Work-up was negative for primary renal Wilms tumor or distant metastasis. Patient underwent adjuvant chemoradiation therapy for stage III disease. Patient is currently 2 years of age with no evidence of recurrence or metastatic disease. *UROLOGY* 129: 197–199, 2019. © 2019 Elsevier Inc.

INTRODUCTION

Wilms tumor is the most common primary malignant renal tumor of childhood. Extrarenal Wilms tumor (ERWT) is rare and accounts for 3% of all Wilms' tumors.¹ There have been 4 previously reported cases of primary paratesticular ERWT. Here we describe the first reported case of primary paratesticular ERWT with regional metastasis.

CASE REPORT

The patient was an 8-month old male with a palpably normal left undescended testis in the upper inguinal canal. At the time of left inguinal orchiopexy, the testis was noted to be grossly abnormal with 2 firm, tan lesions located in the upper aspect of the testis. The epididymis was thickened and hard in consistency. A possible regional lymph node was noted to be grossly enlarged. This possible node or tissue was found within the inguinal canal. Given these findings, a left radical orchiectomy with excision of regional lymph node was performed.

Pathologically, the left inguinal nodule was encapsulated by a thin fibrous membrane with submembranous lobules of fat. Though this nodule had the gross appearance of a lymph node, this nodule was found to have no characteristic of normal lymphatic tissue microscopically. It demonstrated the characteristic Wilms triphasic pattern (blastemal, epithelial, and stromal elements) with adjacent foci of nephrogenic rests. The larger mass containing the testis had nodules of tumor containing focal complex tubules and blastema

admixed with areas of nephrogenic rests (similar to those seen in the inguinal nodule). The mass was noted to be infiltrating the peritesticular soft tissue, penetrating the tunica albuginea and interdigitating with the benign seminiferous tubules. The scattered nephrogenic rests suggested origin from juxta-gonadal mesonephric duct remnants.

The testis was immature consistent with patient's age. Tumor did not involve the surgical margins in either specimen. Both specimens stained positively for WT-1 and demonstrated no loss of heterozygosity of chromosomes 1p and 16q. The specimens stained negatively for P53, CD99, Myogenin, and Desmin. No testing was done for gain of 1q.

Oncological work up was negative for primary renal Wilms' tumor or distant metastasis. The patient was treated as Wilms tumor stage III due to the regional metastasis to the inguinal canal. Given this, patient was not considered for retroperitoneal lymph node dissection as we would have in the case of primary testicular malignancy. In anticipation of adjuvant radiation therapy, the patient underwent surgical relocation of his contralateral right testicle into an abdominal pouch along with Mediport placement. Patient completed chemotherapy with Vincristine, Doxorubicin, and Dactinomycin over a 6-month period (COG protocol AREN0532). Patient completed external beam radiation to the left inguinal canal with 6 treatments for a total radiation dose of 10.8 Gray.

After completion of adjuvant chemoradiation therapy, patient underwent relocation of the right testicle to the scrotum at 15 months of age. Patient is currently 2 years of age and is doing well with no evidence of recurrence or metastatic disease on surveillance imaging.

Conflict of interest: None.

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DISCUSSION

ERWT was first described by Moyson et al in 1961. ERWTs are rare and the majority of the reported cases

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originate in the retroperitoneum and inguinal regions along the spermatic cord followed by sacrococcygeal region, thorax, chest wall, and uterus.^{2,3} The prevalence of primary paratesticular ERWT is even more rare, with only 4 reported in the literature. ERWTs occur mostly in childhood.⁴

Clinical presentation of ERWTs depends on the location and stage of the tumor. Most presenting symptoms are nonspecific and vary according to the extrarenal location as well as the presence or absence of mass effect on surrounding structures. Given this, diagnosis of ERWT is usually made after surgical resection of the tumor and pathological evaluation of the specimen. Pathologically, ERWTs share the same classic primary renal Wilms tumor triphasic pattern with epithelial, tubular, and stromal components. ERWTs are also histopathologically classified as favorable and unfavorable. The majority of reported ERWT cases are of favorable histology. Teratomatous elements must be excluded during pathological analysis of the entire specimen as teratoid Wilms tumor (defined as having >50% teratomatous elements) represent a different class of tumors with a different embryological origin.⁵

Embryogenesis of ERWTs is controversial but they are thought to arise from immature kidney remnants (primitive metanephric cells).³ These cells can be found anywhere along the craniocaudal migration line of mesonephrones and metanephros cells. The urogenital ridge and mesonephrones are in close proximity during embryologic development. Renal embryonic rests, metanephric blastema or parts of the mesonephric duct might be displaced to the adjacent gonad or Wolffian duct, which can persist in postnatal. This may explain the cases of ERWT found the inguinal regions along the spermatic cord and testis, uterus and sacrococcygeal region. Interestingly, ERWTs have been seen in association with horse-shoe kidneys in 13% of the reported cases so far.³

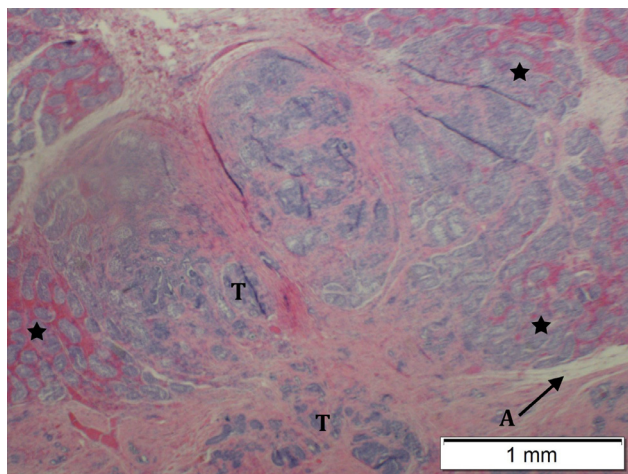


Figure 1. Primary paratesticular ERWT (T) invading the testis—H&E stain. A = tunica albuginea; ★ = seminiferous tubules. (Color version available online.)

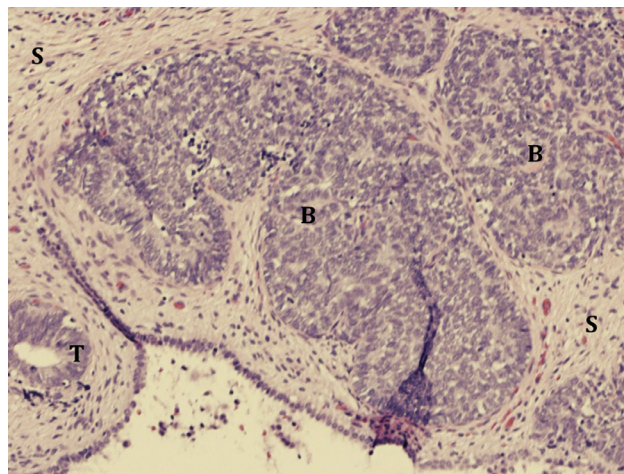


Figure 2. Primary paratesticular ERWT tumor demonstrating the classic Wilms triphasic pattern with blastema (B), tubular epithelial (T) and stromal components (S)—H&E stain. (Color version available online.)

EWRTs are treated with radical surgical excision along with regional lymph node sampling. After the diagnosis of ERWT has been established, patients must undergo radiographic evaluation to rule out primary renal Wilms tumor with a secondary extrarenal renal metastasis as well as to determine metastasis status. According to Shojaeian et al, ERWTs rarely metastasize but 3% of reported ERWT cases were metastatic. Lungs and liver are the most common locations for distant metastasis. Local recurrence has been observed in 11% of the reported cases.

Staging of ERWTs has been challenging. There is currently no accepted staging for ERWT. However, the National Wilms Tumor Study staging system can be applied to EWRT to guide treatment. Adjuvant chemotherapy is recommended for all patients with ERWT

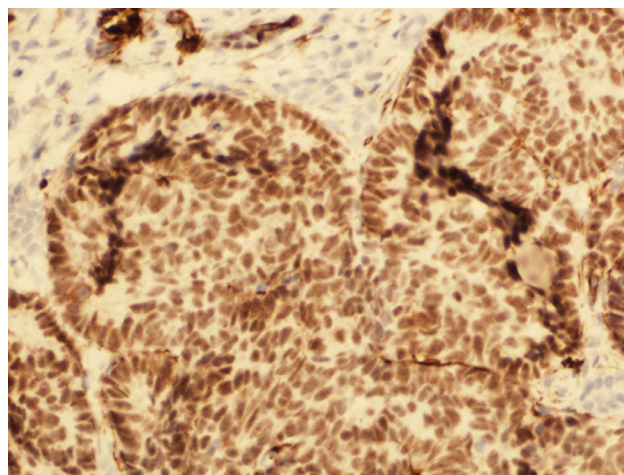


Figure 3. Primary paratesticular ERWT with strong nuclear staining with WT-1. (Color version available online.)

despite of favorable histopathology. Stage II ERWT (ie, completely excised with negative surgical margins), adjuvant chemotherapy with Vincristine and Actinomycin D is recommended. Doxorubicin is added to this adjuvant regiment for those with stage III ERWTs. Radiotherapy is recommended for those with unresectable disease or those with gross residual disease following excision, and for metastasis. In a retrospective review of 34 cases, the radiation dose used range from 2,000 to 5,000 cGy.²

Mutations of the WT1 gene on chromosome 11p13 are observed in approximately 25% of ERWTs.³ Just like with primary renal Wilms tumor, ERWT should be evaluated for tumor-specific loss of heterozygosity for in chromosomes 1p and 16q. These are associated with an adverse outcome even with favorable histology (Figs. 1-3).

According to Andrews et al, the prognosis of ERWT is comparable to that of a primary intrarenal Wilms tumor.⁴

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