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Report of a rare testicular teratoid Wilms Tumor in an adult patient

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ABSTRACT

Although primarily a pediatric disease, nephroblastomas (also known as Wilms tumor) occur in adults at a rate of less than 0.2 cases per million per year. Rarer still are teratoid Wilms tumors, which arise from teratomas and therefore can be extrarenal. We describe the sixth recorded case of a testicular teratoid Wilms tumor in an adult patient with accompanying histological images of the specimen. Following the case, there is a brief discussion of the current literature.

1. Introduction

Nephroblastoma (also known as Wilms tumor) is the most common form of pediatric renal, and second most common cause of pediatric intraabdominal malignancy.[1] Mutations most commonly associated with nephroblastoma are WT1 and WT2, although other genes and loci are likely involved.[1,2] The classic presentation is an asymptomatic abdominal mass in a pediatric patient, however hypertension, malaise, pain and hematuria may also be seen.[1] Although once a quite deadly disease, advances in chemotherapy, radiation, imaging, and surgical oncology have increased the overall survival rate to over 90% in recent studies.[1]

While primarily a disease of children, nephroblastomas are known to occur in adults as well at a rate of less than 0.2 cases per million per year.[2] Renal presentations are overwhelmingly the most common in the adult population, however extrarenal tumors have been observed arising from ovaries, uteri and testicles.[2] Although data are limited, there is some evidence that adults may suffer from a more symptomatic presentation than their pediatric counterparts.[3] Owing to their rarity, adult nephroblastoma do not have true treatment guidelines and are instead treated with regimens identical to pediatric nephroblastoma.[2,3]

In general, the prognosis is worse for adults with nephroblastoma than it is for children, however the overall 5-year survival rate is still above 80%.[3] There are several potential reasons for the discrepancy in survival rates. Adults tend to present with more advanced disease, with 45–70% of adults and 30% of children having stage III or IV disease at time of nephrectomy.[2] Chemotherapy toxicities, while expected, are more severe in adults being treated for nephroblastoma than children.[2] Noncompliance, due both to the aforementioned toxicities as well as due to unfamiliarity with nephroblastoma regimens may also contribute.[3] Finally, there is an increase in the time between surgery and chemotherapy in adults being treated for nephroblastoma. One study reported significant survival benefits for those receiving adjuvant therapy less than 30 days after surgery, however in the same study the mean time between surgery and adjuvant therapy was 59 days.[2]

2. Report

Extrarenal, "teratoid" Wilms tumors of the testicle are an exquisitely rare occurrence, with only 5 prior cases described in the literature. We report the case of a 28-year-old with nephroblastoma occurring within a testicular teratoma. Four months prior to presentation, the patient noticed a persistent, progressive swelling of the right testicle that was not associated with pain. Upon presentation, a right testicular mass was noted, with ultrasound concerning for testicular tumor. Abdominal X-ray at that time was negative for obvious abdominal disease, tumor marker labs showed AFP 26.8 ng/mL, LDH 128 U/L, and HCG <3 mIU/mL.

Two weeks after initial presentation, the patient underwent abdominal CT, which showed multiple left periaortic and right lower quadrant mesenteric lymph nodes ≤7 mm in size. These lymph nodes were thought to be reactive and not pathological in nature. No renal lesions were noted on radiography. Approximately three weeks after presentation, the patient was taken to the OR for right radical orchiectomy. A tumor measuring 5.5 × 5.0 × 4.0 cm was removed with associated epididymis and spermatic cord (Fig. 1). Pathology revealed pT1Nx malignant mixed germ cell tumor composed of teratoma (85%) with associated nephroblastoma, yolk sac tumor (10%), and seminoma

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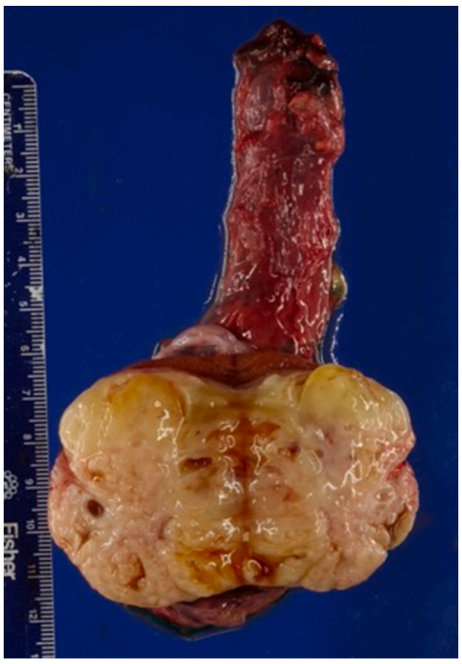

Fig. 1. Radical Orchiectomy Gross Specimen: Tan-white mass measuring 5.5 × 5.0 × 4.5 cm. The mass is seen to replace almost the entirety of the testicular parenchyma with no gross signs of necrosis.

(5%) (Fig. 2). The nephroblastoma was noted to make up 25% of the teratoma. Staining of the nephroblastoma element of the tumor was positive for cytokeratin, OSCAR and glypican 3, weakly focally positive for WT1, chromogranin, synaptophysin, CD56, NSE, CD99 and PAX8, and negative for beta-catenin, desmin and myoD1 (Fig. 3). The immunophenotype and morphology of this tumor were felt to strongly support the diagnosis of nephroblastoma.

CT chest taken one month after surgery was negative for metastatic disease. Serum tumor markers taken at this time showed AFP 2.3 ng/mL, LDH 153 U/L and HCG <3 mIU/mL. After consultation with multiple oncologists, RPNLD was agreed upon to guide administration of adjuvant chemotherapy. Right-sided, modified template RPNLD performed 2 months after initial orchiectomy showed no evidence of tumor in any of the 13 paraaortic lymph nodes removed. As there was no sign of residual tumor after orchiectomy, the patient was not started on adjuvant chemotherapy.

3. Discussion

Teratoma with nephroblastoma elements, also known as teratoid Wilms tumor is a poorly characterized disease due to its rarity.[4] A 2017 literature review revealed only 54 cases in the more than 55 years between the first reported case and the review's publish date.[4] Staging and treatment of these tumors remain controversial, as guidelines state that tumors outside of the kidney are considered stage II and therefore require adjuvant chemotherapy.[1] As stated previously, however, the adjuvant agents (vincristine and dactinomycin) used in stage II Wilms tumor have significant toxicities in adults.[3] There is additional mounting evidence that teratoid Wilms tumors may have substandard responses to chemotherapy, further calling into question the use of adjuvant therapy in these cases.[5]

Although many genes, notably WT1 and WT2, have been implied in the pathogenesis of Wilms tumor in general, there remain many questions regarding teratoid Wilms tumor. The consulting geneticist on this case believed the tumor to be sporadic due to lack of other syndromic features and negative family history. Although offered a full genetics panel, the patient did not follow up for specimen collection, leaving any genetic etiology to remain unknown.

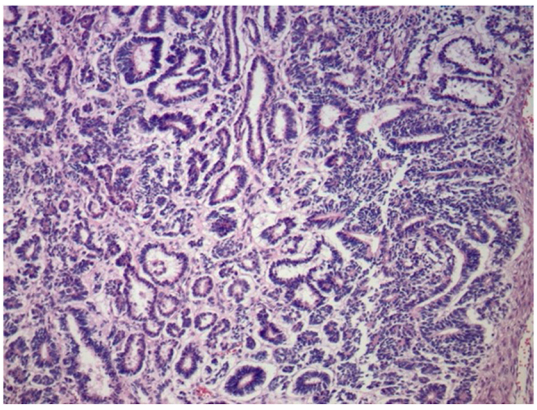

Fig. 2. H&E Histology of Wilms Tumor component: Epithelial tubular structures, small blue blastemal cells, and pale mesenchymal stroma can be seen. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

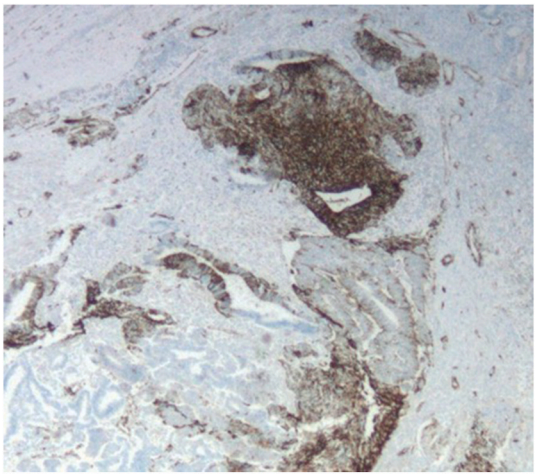

Fig. 3. WT1 Staining of Wilms Tumor component: Patchy WT1 expression can be seen on immunohistochemistry. Although not shown in this image, the Wilms Tumor component also stained positive for cytokeratin.

4. Conclusion

Limited data exist regarding the treatment of teratoid Wilms tumor, especially in the adult population. Owing to this, the decision of when and if to proceed with chemotherapy, especially in the setting of high toxicities, is a difficult one. In our case, the decision not to use adjuvant chemotherapy was made somewhat easier by negative CT, normalizing tumor markers, and a negative RPLND. Further data are needed to establish a treatment protocol for teratoid Wilms Tumor, although owing to this disease's rarity the effort would likely need to be multicentered and highly collaborative.

Consent

The patient provided informed consent prior to the submission of this case report.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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