

**Running Head:** Wilm's tumor and hamartoma with ectogenital deformity in a boy-Li et al.

**Wilms' tumor and benign renal tumor combined with hypospadias and  
incomplete orchiocatabasis appearing simultaneously in a 10 months old boy**

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**Key words:** Wilms' tumor ; benign renal tumor ; bilateral renal tumors ; hypospadias;  
syndrome; boy

## **ABSTRACT**

We herein report a case of left renal Wilms' tumor and right renal hamartoma combined with hypospadias and incomplete orchiocatabasis in a 10-month-old boy. In the literature to date, no case has been reported. The preoperative abdominal computerized tomography (CT) scan was suggestive of bilateral nephroblastomas, and clinical diagnosis was bilateral renal tumors with external genitals malformation, a syndrome? Finally, this case was used by B-ultrasonic guided percutaneous biopsy to help determine the nature of bilateral renal tumors. Afterwards, the boy underwent preoperative chemotherapy, surgery ( a left radical nephrectomy and right wedge excision of the renal tumor) and postoperative chemotherapy. After 3 years of follow-up, there was no evidence of tumor recurrence, the renal function was normal, and the boy's height, weight and intelligence were also within normal range. Owing to no similar cases as a reference, we discussed the preoperative imaging diagnosis, final etiological diagnosis and appropriate treatment of this disease. Long-term follow-up with a sufficient number of cases may be needed to optimize methods of diagnosis and define optimal treatment options for patients with this extremely rare disease.

## INTRODUCTION

Wilms' tumor is the most primary malignant renal tumor in children. Renal hamartoma is a benign neoplasia and is extremely rare in children. We report a case of Wilms' tumor and renal hamartoma combined with hypospadias and incomplete orchiocatabasis appearing simultaneously in a 10-month-old boy. In the literature to date, no identical cases have been reported. Owing to no case as a reference, preoperative imaging diagnosis, final etiological diagnosis and appropriate treatment are very important.

## CASE REPORT

A 10-month-old boy was presented to a palpable abdominal mass with hypospadias and incomplete orchiocatabasis. Laboratory studies showed the blood cell counts, hemoglobin level, blood urea nitrogen, serum creatinine, sex and cortisol hormones were within the reference range. Urinalysis was also normal. Abdominal ultrasound study showed a large solid inhomogeneous retroperitoneal tumor and the urinary system was normal. Computerized tomography (CT) of the abdomen showed a giant heterogeneous mass 11 cm × 10 cm in size, arising from the upper and middle pole of left kidney and crossing the midline of the abdomen. The lesion 2.5 cm × 1.5 cm in size located into the lower pole of right kidney exhibited a heterogeneous and hypodense mass on the contrast-enhanced CT scan (**Figure a**). The CT scan was suggestive of bilateral nephroblastomas. Finally, this case was used by B-ultrasonic guided percutaneous biopsy to help determine the nature of bilateral renal tumors; the result

showed that: the left tumor was a nephroblastoma and the right tumor tended to be a hamartoma. Afterwards, the boy received 6 courses of actinomycin-D and vincristine chemotherapy treatment, there was no significant change in bilateral renal tumors. After chemotherapy treatment, the child was prepared for surgery; a left radical nephrectomy and right tumor wedge excision was performed, and topical cooling of the kidney and vascular control techniques were applied in the right tumor excision (**Figure c**). Postoperative pathology and immunohistochemistry showed that the left tumor was a Wilms' tumor (favorable histology, stage II) and right lesion was a hamartoma (**Figure d,e**). Subsequently, the boy received actinomycin-D, vincristine and adriamycin for approximately 38 weeks in postoperation. The boy underwent cryptorchidopexy and urethroplasty at the age of 1.5 and 3 years. At 3 year follow-up, there was no evidence of tumor recurrence(**Figure b**), the renal function was normal, and the boy's height, weight and intelligence was also normal.

## DISCUSSION

Wilms tumor is the most common pediatric renal tumor, benign renal tumors had been reported infrequently in children, in our case, this boy had two different natures of bilateral renal tumors and congenital ectogenital deformity, these manifestations are related or independent diseases? Is it a syndrome? Such as WAGR syndrome <sup>(1)</sup> (Wilms' tumor, aniridia, ambiguous genitalia, mental retardation), Wiedemann-Beckwith syndrome <sup>(2)</sup> (aniridia, hemihypertrophy, urogenital malformation, Wilms' tumor), Perlman syndrome <sup>(3)</sup> (fetal gigantism,

nephroblastomatosis, hypertrophy of the endocrine pancreas). For this purpose, we examined the boy and his parents' chromosomes and genes in post-operation, the results were normal. From birth to now, the boy's height, weight and intelligence are within normal range. According to the related examination results, we think that the boy's various disease performances may not be related, but we do not rule out the possibility of a kind of syndrome, we need further follow-up and discover similar cases.

As a rare disease, the radiological appearance of renal hamartoma had been reported infrequently in human, especially in children. And the right lesion was once considered a nephroblastoma before biopsy. Common hamartoma exhibited well-circumscribed, round or lobulated nodules 1-3cm in diameter with sharp margin, classical morphologic features such as "popcorn" or fat, a heterogeneous appearance, and identification of calcification within the nodule or fat allows for a confident diagnosis of hamartomas on CT images in lung and reflected the gross patterns ranging from a predominantly cystic mass to a complex solid mass in liver <sup>(4-7)</sup>. And nephroblastoma had a characteristic inhomogeneity with a predominance of hypodense areas and increased inhomogeneity after contrast administration <sup>(8)</sup>. By contrast, we can understand the similarities and differences between these two types of tumors in CT. Therefore, based on the lack of related literature, we can use this case combined with related literatures as summary of renal hamartoma characteristics in CT: it is generally less than 3cm in size and CT showing a heterogeneous, hypodense appearance and a little irregular margin of solid mass which confined

within the renal capsule, this tumor could be considered as a renal hamartoma or benign lesion.

In our case, considering the growing tumor on the right side, and risk of rupture and malignant transformation <sup>(3)</sup>, therefore, we decided to perform a right wedge excision simultaneously. Due to the un conspicuous effect of preoperative chemotherapy, we added the adriamycin into postoperative chemotherapy. In addition, the boy had subcoronal hypospadias with mild curvature, we did tubularized incised-plate (TIP) urethroplasty, the function and cosmetic result was good. As we know, there are many methods to treat hypospadias currently. Such as meatal advancement and glanuloplasty (MAGPI), TIPS, and one-stage or two-stage procedure techniques. Recently some modified techniques are good options for treatment of anterior hypospadias, like anterior urethral advancement technique <sup>(9)</sup>, modified firilit's technique <sup>(10)</sup>, they have the characteristics of one-stage, easy to learn, good appearance and low complications, especially in properly selected case; moreover, "two-stage procedure" has seen increased in popularity for patients of proximal hypospadias and severe curvature, like vascularized preputial island flap technique has been successfully for management of this patients <sup>(11)</sup>.

In conclusion, we may have provided some experiences with this disease, and long-term follow-up with a sufficient number of cases may be needed to define optimal diagnosis and treatment options.

## **CONFLICT OF INTEREST**

There is no conflict interest.

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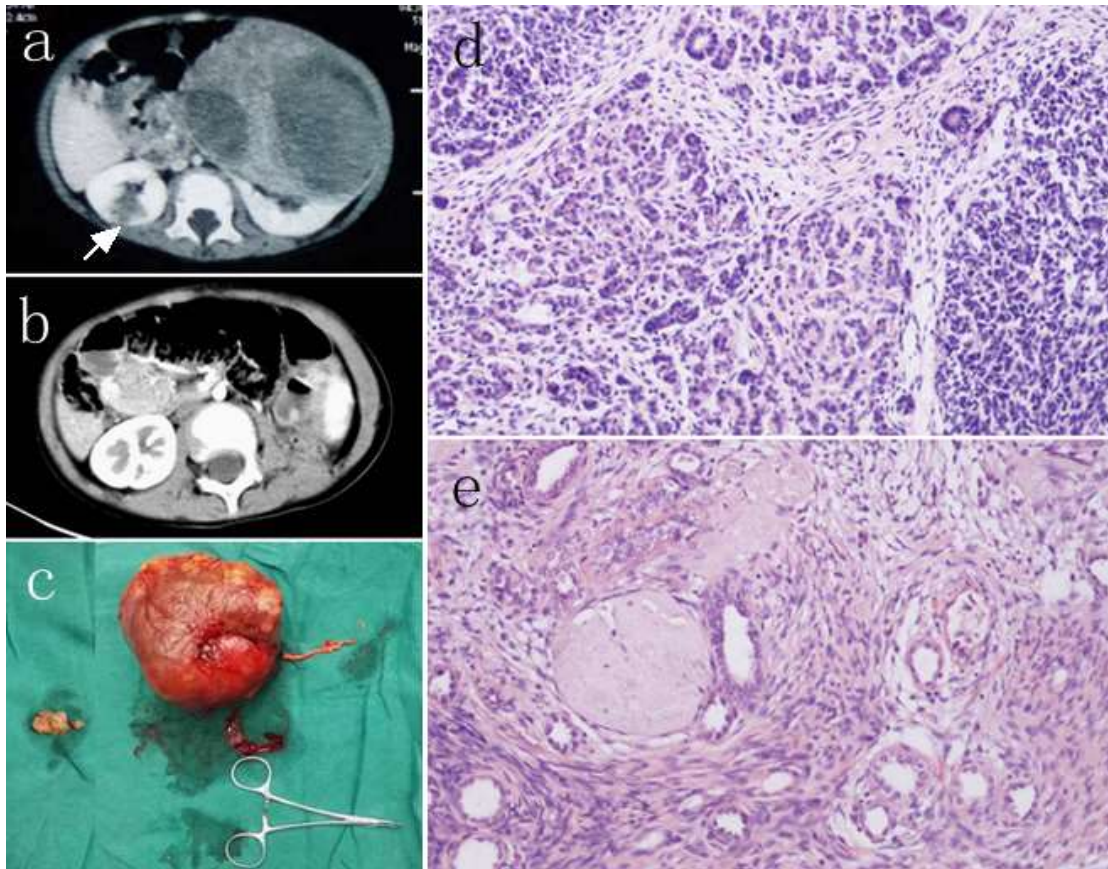
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**Figure a** Preoperation: shows a giant heterogeneous mass was arising from upper and middle pole of left kidney and crossing the midline to the right side of the abdomen on CT scan, the right lesion shows a heterogeneous and hypodense lesion, and its boundary was not clear on contrast enhancement.

**Figure b** Postoperation: 3 years follow up, tumor recurrence was not seen in the primary site.

**Figure c** Tissue specimens: the left ( renal hamartoma ) lesion specimen was harder than the right ( wilm's tumor ) tumor specimen, and the cut surface showed a solid and offwhite appearance.

**Figure d,e** Wilms' tumor ( Stage II, Favorable Histology ) primitive epithelial and rhabdoid differentiation, primitive embryo H&E  $\times 100$ . Hamartoma which contained mature mesenchymal tissues and mature tubular-like structures, cartilages. H & E $\times 100$ .