

## Wilms' Tumor and Benign Renal Tumor Combined with Hypospadias and Incomplete Orchioctabasis Appearing Simultaneously in A 10 Months Old Baby

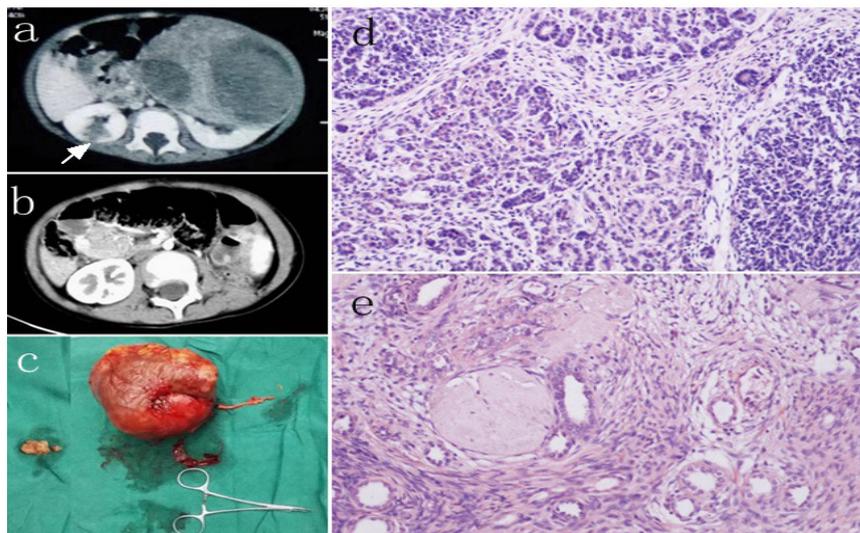
Chun Li<sup>1</sup>, Wei-Song Li<sup>1\*</sup>

We herein report a case of left renal Wilms' tumor and right renal hamartoma combined with hypospadias and incomplete testis descent 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. Finally, we used B-ultrasonic guided percutaneous biopsy to help determine the nature of bilateral renal tumors. Afterwards, the boy underwent preoperative chemotherapy, surgery (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.

**Keywords:** Wilms' tumor ; benign renal tumor ; bilateral renal tumors ; hypospadias; boy

### 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 cryptorchidism 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.



**Figure 1.** 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. b) Postoperation: 3 years follow up, tumor recurrence was not seen in the primary site. 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 off white appearance. 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  $\times 100$ .

Department of Pediatric Surgery, Department of General Surgery, The First Affiliated Hospital of Anhui Medical University, 218 JiXi Avenue, Hefei 230022, Anhui, PR China.

\*Correspondence: Department of Pediatric Surgery, Department of General Surgery, The First Affiliated Hospital of Anhui Medical University, 218 JiXi Avenue, Hefei 230022, Anhui, PR China.

Tel: +86 551 62923764. Fax: +86 551 62923770. E-mail: leiforget1970@hotmail.com.

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## CASE REPORT

A 10-month-old boy was presented to a palpable abdominal mass with hypospadias and cryptorchidism. 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 1a). The CT scan was suggestive of bilateral nephroblastomas. We used B-ultrasonic guided percutaneous biopsy to help determine the nature of bilateral renal tumors. Pathology 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 1c). Postoperative pathology and immunohistochemistry showed that the left tumor was a Wilms' tumor (favorable histology, stage II) and right lesion was a hamartoma (Figure 1d,e). Subsequently, the boy received actinomycin-D, vincristine and adriamycin for approximately 38 weeks in postoperation. The boy underwent orchidopexy and urethroplasty at the age of 1.5 and 3 years. At 3 year follow-up, there was no evidence of tumor recurrence (Figure 1b), 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. Syndromic approach could also be thought of like WAGR<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 (8). 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>, we decided to perform a right wedge excision simultaneously. Due to the unobvious effect of preoperative chemotherapy, we added the adriamycin into postoperative chemotherapy. In addition, the boy had subcoronal hypospadias with mild curvature. We performed tubularized incised-plate (TIP) urethroplasty and the function and cosmetic result was good. 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.

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