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Case reports

# Teratoid Wilms' tumor: a case report with literature review

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**Abstract** Teratoid Wilms' tumor is an unusual histologic variant of nephroblastoma in which the heterologous tissue predominates. The treatment for this tumor has not been established because of its rarity and varying tumor components. The authors report a case of localized teratoid Wilms' tumor in a 4-month-old female infant successfully treated with nephrectomy only. The tumor originated from the right kidney with multiple cysts and calcifications; renal teratoma was suspected preoperatively, and the operation was performed. The tumor was excised completely, and the specimen weighed 340 g. Histopathologic examination showed that various teratoid elements occupied most of the tumor with sparse islands of triphasic nephroblastoma, which was consistent with teratoid Wilms' tumor. Anaplastic elements, tumor capsule invasion, renal sinus soft tissue invasion, or tumors in intrarenal vessels were not observed. The patient was disease free for 3 years after surgery without receiving chemotherapy. We selected this therapeutic approach because of the chemoresistance of this tumor and the excellent prognosis in small (<550 g) stage I/favorable-histology classical Wilms' tumor diagnosed at less than 24 months of age. Therefore, nephrectomy alone may be an acceptable treatment of this stage of tumor in patients with teratoid Wilms' tumor.


Teratoid Wilms' tumor, a rare variant of nephroblastoma, has a predominance of teratoid elements consisting of more than 50% of the tumor [1]. To date, 15 cases of teratoid Wilms' tumor have been reported in the literature [1-12]. All patients were operated on with combination chemotherapy, and some patients also received radiotherapy, which is the current therapeutic strategy for classical Wilms' tumor. Some problems such as resistance to chemo- and radiotherapy result from the mature heterologous components [1-4]. Treatment of teratoid Wilms' tumor has not yet been established. We describe here an infant with unilateral localized teratoid Wilms' tumor who was successfully treated with nephrectomy only, and we have also reviewed all cases reported in the literature.

## 1. Case report

A 4-month-old boy had a palpable mass on the right side of the abdomen and was admitted for evaluation and management to Mie University Hospital. He was an otherwise healthy child with no remarkable medical history. There was no episode of hematuria before admission.

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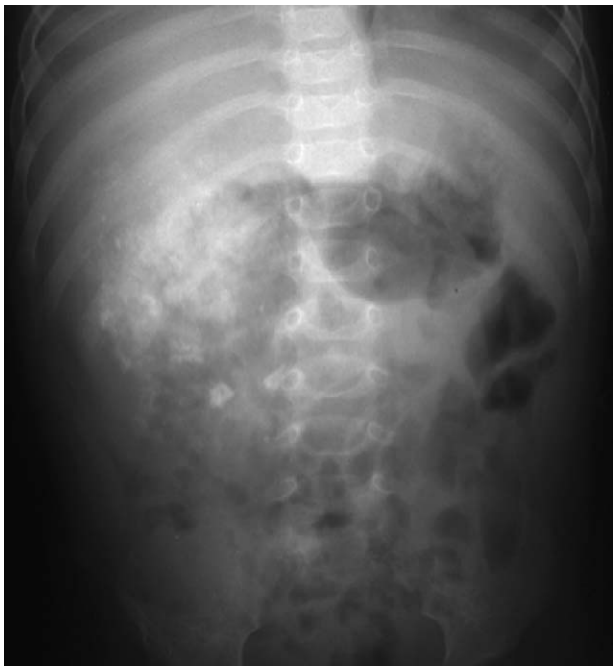

**Fig. 1** Abdominal x-ray showing localized calcification in the right side of the abdomen.

Physical examination revealed an elastic hard mass without tenderness at the right flank of the abdomen. The mass was palpable 8 cm below the right costal margin. A noteworthy laboratory finding upon admission was a human chorionic gonadotropin level of 4.5 ng/mL (normal, <0.5 ng/mL), and a neuron-specific enolase level of 41 ng/mL (normal, <10 ng/mL). Serum levels of ferritin, alpha-fetoprotein, vanillylmandelic acid, and homovanillic acid were within normal limits. Abdominal radiography showed scattered calcification in the mass (Fig. 1). Transabdominal ultrasonography and abdominal computed tomography (CT) revealed a large multicystic mass with multiple calcifications occupying the right kidney. CT scan with intravenous contrast showed that only the encased renal parenchyma was enhanced (Fig. 2). Chest and brain CT findings were normal.

All these findings suggested that renal teratoma was the most probable diagnosis, and thus, right nephrectomy was performed. At the operation, we observed that most of the right kidney was replaced by the tumor, which was enveloped by a smooth, glistening capsule. The inferior vena cava was compressed by the tumor, but vascular invasion was not observed. Some enlarged paraaortic and precaval lymph nodes were also resected. The nephrectomy specimen weighed 340 g and measured 8 × 7 × 5 cm.

Histopathologically, the tumor consisted primarily of various teratoid elements such as stratified squamous epithelium, columnar epithelium, pigment cells, mature adipose tissue, and cartilage or bone tissue. Tendency toward organogenesis was not shown in these tissues. There were also occasional islands of triphasic nephroblastoma with blastematous, epithelial, and stromal components (Fig. 3). Anaplastic elements, tumor capsule invasion, renal sinus soft tissue invasion, or tumors in intrarenal vessels were not observed. There was no lymph node metastasis. These findings were compatible with those of teratoid Wilms' tumor at stage I.

The postoperative course was uneventful, and the child was discharged from hospital without receiving chemotherapy. Preoperatively elevated serum levels of the tumor markers had fallen to a normal range within 8 weeks after surgery. The patient is alive and free of disease 3 years after surgery.

## 2. Discussion

Wilms' tumor is an embryonic tumor with a triphasic histologic pattern represented by blastemal, stromal, and epithelial components [13]. It is recognized that nephroblastoma can include heterologous elements in addition to

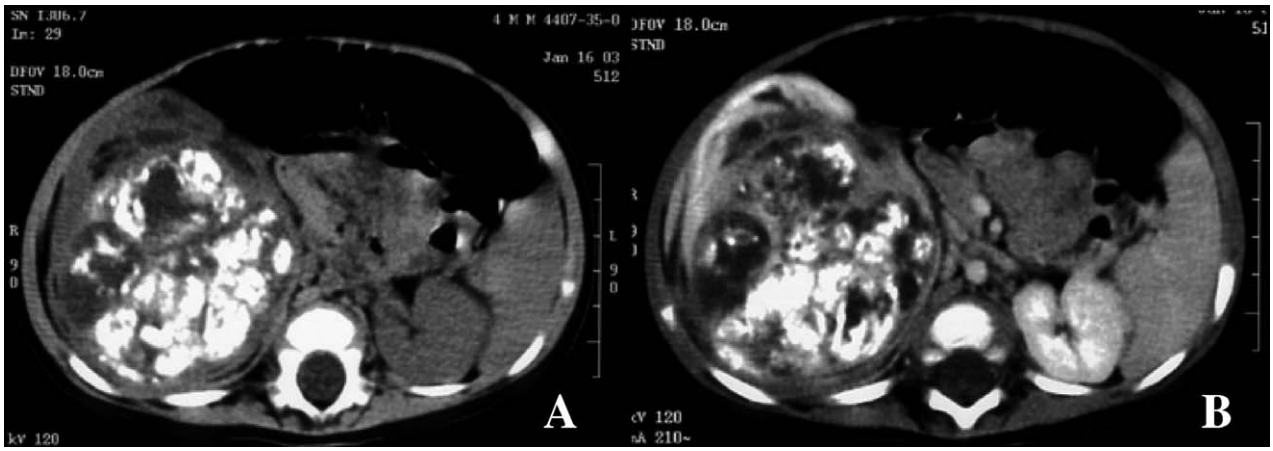


**Fig. 2** CT before operation. A, Noncontrasted study shows the multicystic tumor originating from the right kidney with multiple calcification. B, Contrasted study reveals enhancement of the encased renal parenchyma only.