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# Teratoid Wilms Tumor in a Child: A Case Report

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**ABSTRACT**

Teratoid Wilms is a very rare histopathological variant of Wilms tumors that is characterized by the predominance of heterologous components, and is described by many to be a nonaggressive, nonmetastatic tumor with a favorable prognosis. We report a case of a 4-year-old boy with a rare, aggressive metastatic variant of teratoid Wilms tumor. The boy presented with abdominal pain and a palpable abdominal mass. The computed tomography scan demonstrated a large, cystic, multiloculated left renal mass and a single left pulmonary metastasis. The patient had a 5-week course of neoadjuvant chemotherapy. As a result, the size of the metastatic lesion decreased, but there was no change in the size of the renal mass. Subsequently, a left radical nephrectomy and left pulmonary metastectomy were performed where the pathology report showed metastatic teratoid Wilms tumor. The patient received adjuvant chemotherapy and radiotherapy. One year following the initial surgery, the child developed bilateral pulmonary metastases where he is currently having an aggressive regimen of chemotherapy. Although teratoid Wilms has been described as a nonaggressive tumor with a favorable prognosis, it can present with more aggressive forms, with a tendency for metastasis.

## INTRODUCTION

Wilms tumor is an embryonic tumor of mesodermal origin. It is typically characterized by a display of a triphasic histological pattern of blastemal, stromal, and epithelial cells. Heterologous mesodermal components, such as adipose tissue, skeletal muscle, cartilage, and neurological tissue, may be seen in small foci throughout the neoplasm [1-3]. In 1984, a rare variant with heterologous predominance was described by Variend et al., who introduced the term "teratoid Wilms" [1]. Teratoid Wilms has been described by many as a nonaggressive, nonmetastatic tumor with a favorable prognosis [1-3,5]. Review of 17—including 15 cases without metastases and 2 with metastases—cases showed that 12 of them had no evidence of disease after receiving treatment [3]. Similar outcomes were reported in other cases [5,8]. Conversely, Myers et al. [8] stated that 50% of those with teratoid Wilms presented at stage III or higher. They also reported an incidence of bilateral disease in 38%. According to the available data, a total of 3 cases of metastases and 4 deaths—2 of them linked to progression—have been

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reported in those with teratoid Wilms [3,9,10]. We report a rare case of 4-year-old boy with an aggressive metastatic variant of teratoid Wilms tumor.

## CASE REPORT

A 4-year-old healthy boy presented with abdominal pain and a palpable abdominal mass. A computed tomography (CT) scan of the abdomen showed a huge multiloculated cystic mass involving the left kidney (Figures 1 and 2). A CT chest revealed a single metastasis in the left lung (Figure 3). A diagnosis of Wilms tumor of the left kidney with pulmonary metastasis was made. The patient underwent chemotherapy for 5 consecutive weeks. The patient received carboplatin (450 mg/m², IV, day 1 only) and etoposide (100 mg/m², IV, once daily for 5 days) in week 0, vincristine (0.05 mg/kg, IV, day 1 only), and actinomycin-D (15 mcg/kg, IV, once daily for 5 days) in week 1, and weekly vincristine (0.05 mg/kg, IV, day 1 only) throughout the second, third, and fourth weeks. As a result, the size of the metastatic lesion decreased while there was no change in the size of the renal mass. Subsequently, a left radical nephrectomy in conjunction with a left pulmonary metastectomy was performed. Cut sections of the mass were multicystic in appearance, with areas of necrosis and hemorrhage, and showed no extension to the perinephric fat. Microscopically, sections of the kidney revealed a multicystic neoplastic process with wide areas of necrosis, hemorrhage, and hemosiderin-laden macrophages. The tumor was composed of nests and clusters of embryonal-looking cells, with enlarged hyperchromatic nuclei and occasional mitotic figures (Figure 4). Focal areas of spindle cells and smooth-muscle differentiation were also seen. Few cysts were lined by flattened epithelium while others were lined by stratified squamous epithelium and filled with keratin flakes (Figure 5). The left lung nodule was positive for metastases and had a blastemal component (Figure 6). A panel of immunohistochemical markers was performed on the lung nodule, including CK-Pan, vimentin, and WT-1. The tumor cells were positive for WT-1, focally positive for vimentin, and negative for CK-Pan (Figure 7). Postoperative radiotherapy (to the lung and abdomen) and chemotherapy in the form of actinomycin D (15 mcg/kg, IV, once daily for 5 days on weeks 12, 24, 36, 48, and 60 of initial treatment, and 30 mcg/kg day 1 only of weeks 6, 9, 18, 30, 42, 54, and 66 of initial treatment), vincristine (0.05 mg/kg, IV, day 1 only of weeks 6, 9, 28, 30, 42, 54, and 66, and 0.05 mg/kg, IV, once daily on days 1 and 5 in weeks 12, 24, 36, 48. 60), and doxorubicin (40 mg/m², IV, on day 1 of weeks 6 and 9, and 60 mg/m², IV, on day 1 of weeks 18, 30, 42, 54, and 66) were delivered.

One year following the initial surgery, a routine follow-up CT of the chest demonstrated multiple small focal lesions in both lungs. There were no signs of local recurrence or residual disease in the abdomen.

The patient is currently alive 21 months after initial treatment.

Figure 1. Multicystic, multiloculated left renal mass.
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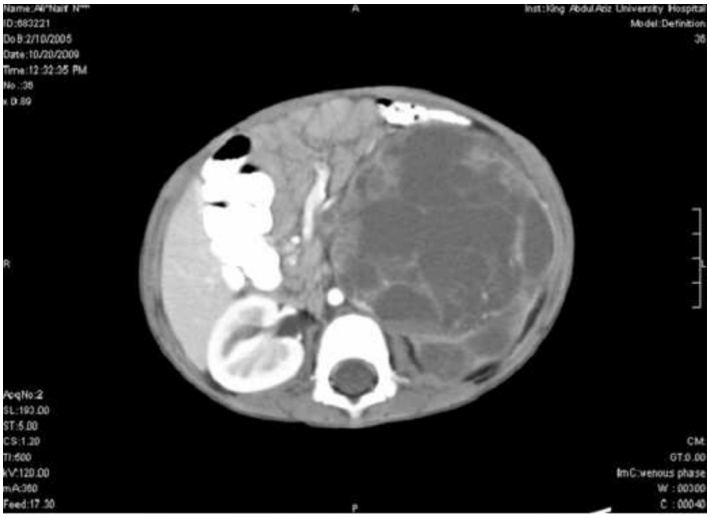

Figure 2. Coronal view of mass.
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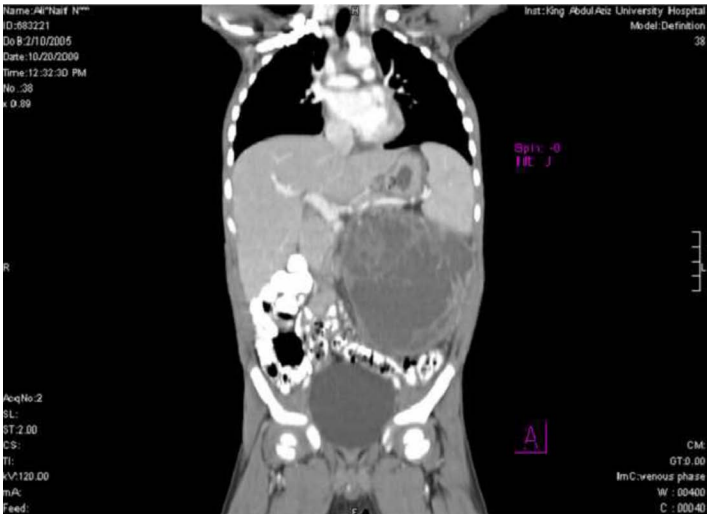

Figure 3. Left pulmonary nodule.
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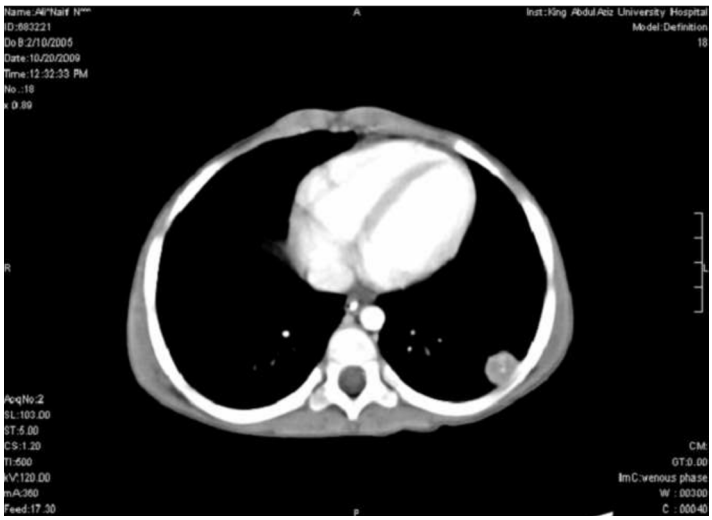

Figure 4. Section from the primary tumor of the kidney showing primitive blue round cells of Wilms tumor. Few tubules are seen showing hyaline casts (X 10, H & E stain).
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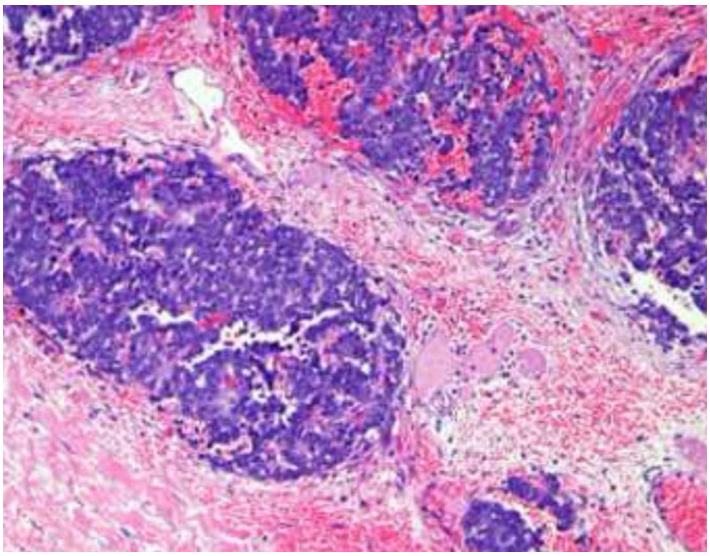

He's had repeated hospital admissions due to febrile neutropenia with chest infections. There has been no cytoreductive response on the metastatic nodules in the lungs so far. There's been no rise to new metastatic lesions at other sites so far. Currently, a regimen of chemotherapy is being given every 3 weeks, for a planned total of 6 courses. It is comprised of ifosfamide (1200 mg/m², IV, once daily for 5 days), carboplatin (450 mg/m², IV, day 1 only), and etoposide (100 mg/m², IV, once daily for 5 days).

## DISCUSSION

Teratoid Wilms is a rare histopathological variant of Wilms tumor, which is characterized by a predominance of heterologous components. A refined definition of teratoid Wilms was introduced by Fernandes et al. in 1988, proposing that the term be used to illustrate versus identify Wilms tumors with a heterologous component of more than 50% [4]. Searching PubMed, we found only 27 cases have been reported.

Teratoid Wilms has had clinical features similar to those of classical Wilms. It affects both sexes, with a mean age of 2.5 years. Abdominal masses and abdominal pain are the usual signs and symptoms [3,5]. This tumor has also been found to have diverse features, such as bilaterality, a tendency to extend into the collecting system, and association with nephroblastomatosis [6]. Park et al. [6] described the CT features of teratoid Wilms: It usually appears as a cystic renal mass with

Figure 5. Squamous component of the tumor (X 20, H & E stain).
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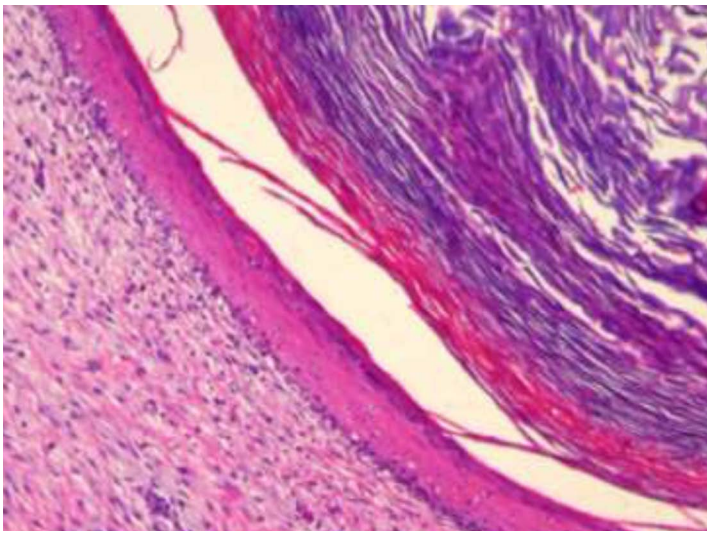

Figure 6. Section from the metastatic tumor to the lung showing sheets of primitive round cells invading the lung parenchyma (X 10, H & E stain).
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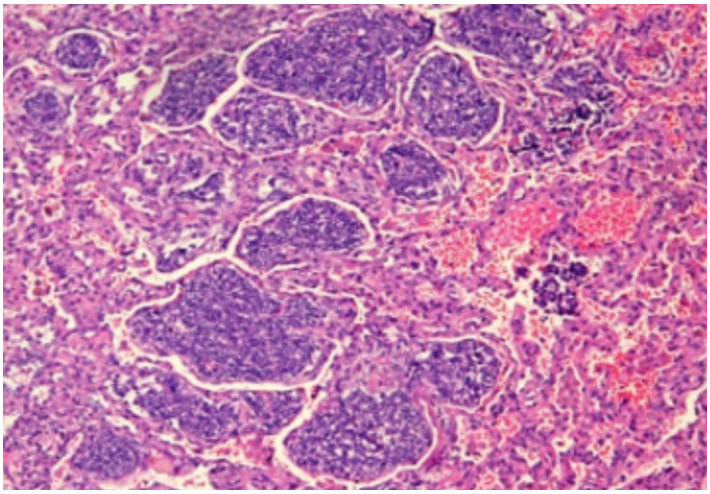

Figure 7. Blastema cells showing positive nuclear staining for WT-1 immunostaining (X 20).
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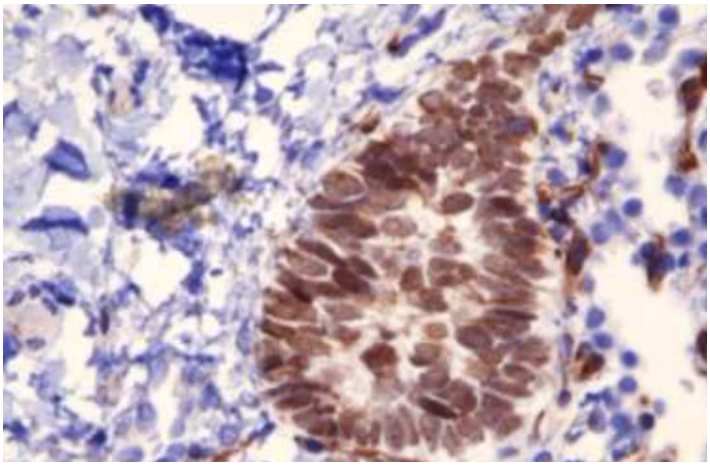

multifocal, solid components containing fatty elements and occasional calcifications.

Teratoid Wilms tumor and renal teratoma have been hisologically similar. Beckwith proposed criteria to differentiate the 2 from each other. He stated that renal teratoids should be intrarenal and show attempts of heterotopic organ formation [7]. Cytogenic analysis for the deletion of the short arm of chromosome 11 has been helpful in complicated cases [2].

Although teratoid Wilms has been described as a nonaggressive, nonmetastatic tumor with a favorable prognosis [1-3,5], a total of 4 cases of metastases have been reported, including our case [3,10]. This rare variant of Wilms has been portrayed as chemo-resistant by many [1,2,10,9]. Neoadjuvant chemotherapy has been unable to produce a cytoreductive response in most cases [10,9]. This may be attributed to the well differentiated nature of the teratomatous elements in these cases [6,2]. In our patient, although downsizing could be achieved in the single pulmonary metastasis, no size reduction was observed in the primary tumor with neoadjuvant chemotherapy.

The American Cancer Society guidelines for the treatment of Wilms tumor advise surgery, followed by chemotherapy with actinomycin-D (dactinomycin) and vincristine for all stages, with the exception of those who are less than 2 years of age, have tumors less than 550 grams, and have a favorable histology. Those with recurrent Wilms tumors may be treated with aggressive chemotherapy, such as the ICE regimen (ifosfamide, carboplatin, and etoposide) or other regimens studied in clinical trials [11]. No specific treatment strategy has been proposed to those with teratoid Wilms. Our patient, as well as 2 other patients who had disease progression, received the ICE regimen, with mortality being the end result of the latter 2 [3].

Taking into consideration the diverse biological behavior of teratoid Wilms tumors, with most of them demonstrating a nonaggressive, nonmetastatic behavior, whereas some variants—including our case—exhibit an aggressive metastatic behavior, a question is raised of whether or not the later variant is truly teratoid Wilms or a different entity. Probably a better description of the clinical, imaging, histopathological, immunohistochemical, and genetic characteristics of these tumors might help distinguish aggressive from nonaggressive variants to predict prognosis.

## SUMMARY

We present a rare, aggressive variant of teratoid Wilms tumor with metastasis. Disease progression was evident, even after neoadjuvant chemotherapy, radical nephrectomy, pulmonary metastatectomy, and adujuvant chemoradiotherapy. Although teratoid Wilms has been described as a nonaggressive, nonmetastatic tumor with a favorable prognosis, it can present

with more aggressive forms, and with a tendency for metastasis. Further investigations are needed to predict tumor behavior and designate treatment protocol for aggressive tumors.

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