

CASE REPORT**BILATERAL TERATOID WILMS' TUMOR IN A CHILD: A CASE REPORT AND REVIEW OF LITERATURE**Amezene Tadesse, MD^{1*}, Tatek Girma, MD², Jakob Schneider, MD³**ABSTRACT**

Teratoid Wilms' tumour is a very rare histopathological variant of the tumour that is characterized by the predominance of heterologous components. It is described by many as a non-aggressive, non-metastatic tumour with a favourable prognosis. We report our experience with a boy who presented with a bilateral flank swelling and significant weight loss. Computed tomography scan demonstrated a large, bilateral intra renal poorly enhancing masses. The patient had neoadjuvant chemotherapy and bilateral nephron sparing surgery for synchronous bilateral Wilms' tumor. The patient had an uneventful recovery. Histopathology report showed bilateral teratoid Wilms' tumor. The management of bilateral teratoid Wilms' tumor depends on individual clinical condition and aims at eradication of the tumor with renal preservation.

Key words: Wilms' tumour; teratoid; case report, Ethiopia.

INTRODUCTION

Wilms' tumour is an embryonic tumour 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, non-metastatic tumour with a favourable prognosis (1-4). 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(4,5). Conversely, Myers et al. (5) 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 reported in those with teratoid Wilms(3,6,7). Tumors of this type behave differently compared to the usual WT because they generally do not respond to chemotherapy or radiation but may be treated effectively by surgery alone (4,8). To date 33 cases of intra renal and 5 extra renal teratoid Wilms tumor have been reported (9). We report a case of a 1-year and 3-month-old boy with this rare variant of Wilms' tumour in addition to the 33 cases reported so far.

CASE REPORT

A one-year and three-month-old boy was referred to our institution with one-month history of bilateral flank swelling and significant weight loss. He had associated poor appetite and low grade intermittent fever. There was no hematuria or abdominal pain. The clinical examination revealed blood pressure of 110/70mmHg (Stage II Hypertension) and mid upper arm circumference (MUAC) of 11cm. He had decreased air entry on the left lower posterior lung field. The abdomen was distended with right flank approximately 12x9 cm hard fixed mass extending to the centre of the abdomen which is bimanually palpable. There was also left flank mass with a size of 3x5 cm, which was fixed and hard in consistency.

Ultrasound revealed huge, well-defined heterogeneous right renal mass arising from the lower pole with some vascularity and calcification on Doppler study. The mass has compressed the calyceal system causing mild dilatation of the pelvi-calyceal system. There was also a heterogeneous well-defined left renal mass arising from the lower pole. It has vascularity and calcification. There was no thrombosis of inferior vena cava (IVC) and renal veins. A computed tomography (CT) scan of the abdomen showed heterogeneously enhancing well defined approximately 9.2x12.7 cm. right renal mass involving the lower pole and mid portion, which caused pelvi-calyceal separation and 3.7x3.7 cm left renal solid mass on the lower pole (Figure1). No metastatic dissemination was registered. The child had normal levels of renal function.

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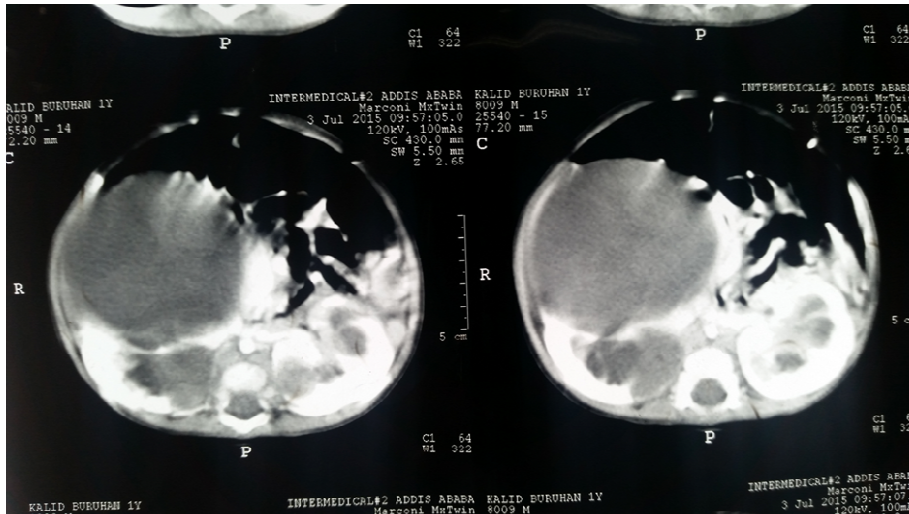


Figure 1. CT scan finding of the abdomen

The child was admitted with the diagnosis of bilateral synchronous Wilms'tumour, severe acute malnutrition (MUAC=11cm), Stage II hypertension and severe pneumonia. On the basis of typical radiological findings, he was started on IV ceftriaxone and nifedipine. The pneumonia subsided after the antibiotic treatment. The patient underwent a preoperative chemotherapy with vincristine and dactinomycin according to the Protocol for localized Wilms' tumor for 10 cycles. He was later given cyclophosphamide and etoposide similar to the protocol for relapse to no avail. Bilateral nephron sparing surgery was

done with a complete tumor excision leaving the healthy renal tissue behind. Gross examination revealed three nodular specimens with smooth surface and solid whitish cut surface on the right side. The diameter of the largest was 11cm; of the smaller was 4.5cm each. One of the smaller nodes was more nodular and cystic on the cut surface. (Figure 2a &2b). The left renal mass was egg shaped with a largest diameter of 4.5cm and a band-shaped necrosis on the cut surface (Figure 3a &3b).

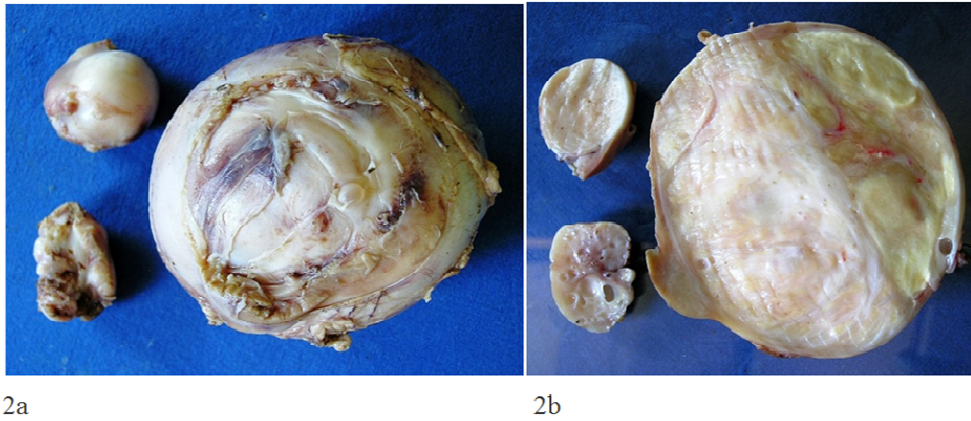


Figure 2a & 2b. Gross feature of right renal masses:

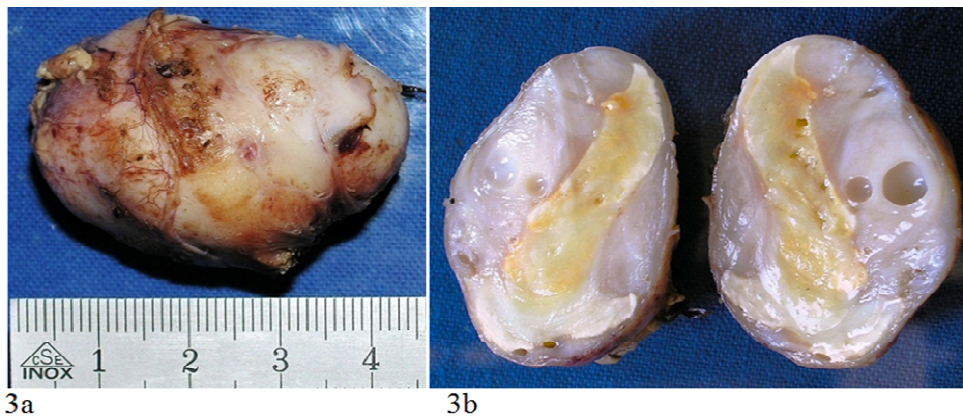


Figure 3. Gross picture of left renal mass

The microscopic features of all four nodes was dominated by intact mature fibrillary mesenchymal tissue with scattered mature renal components: glomeruli, tubuli, some cystic (Figure 4a, 4b,4c) accompanied by heterologous elements: mainly skeletal muscle and foci of squamous epithelium (Figure 4d, 4e,4f).

One of the smaller right sided nodes differs from the others by its prominent cystic component (Figure 5a, 5b). The left sided lesion differs from all others by a band like necrosis, which is demarcated by foamy like macrophages (Figure 6a, 6b)

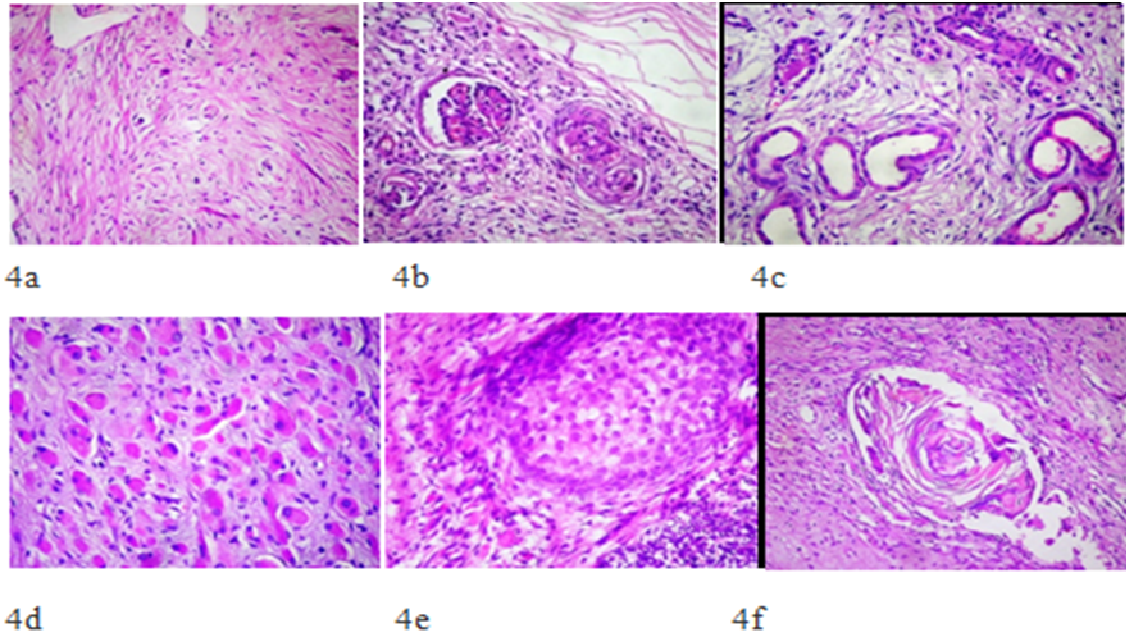


Figure 4a.Fibrillary mesenchyme is the dominant component; 4b. Scattered near normal glomeruli; 4c.Normal looking mature tubuli, some cystic; 4d, skeletal muscle; 4e& 4f: Nests of keratinising squamous epithelium

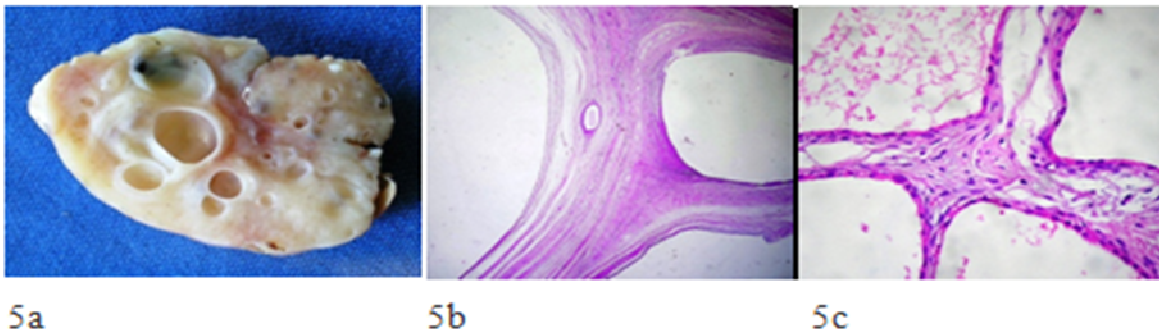


Figure 5. Gross and microscopic feature of the R side smaller node

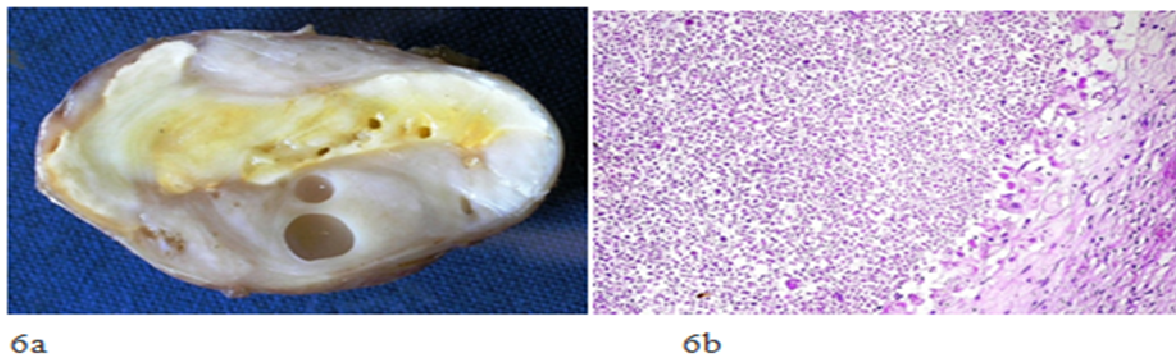


Figure 6. Gross cut surface and microscopic feature of the left sided mass

DISCUSSION

Teratoid Wilms' tumour is a rare histopathological variant of Wilms' tumour, 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 variant of Wilms tumour with a heterologous component of more than 50%.

The microscopic features of all 4 nodes from our patient was dominated by intact mature fibrillary mesenchymal tissue with scattered mature renal components: glomeruli, tubuli, some cystic accompanied by heterologous elements: mainly skeletal muscle and foci of squamous epithelium. These heterologous components are the basis for our diagnosis of Teratoid Wilms in our case.

Teratoid Wilms' tumour has 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 (3,4). This tumour has also been found to have diverse features, such as bilaterality, a tendency to extend into the collecting system, and association with nephroblastomatosis (10). Park et al. described the CT features of teratoid Wilms: It usually appears as a cystic renal mass with multifocal, solid components containing fatty elements and occasional calcifications. The same is true with our case where we got a bilateral tumour with CT finding of heterogeneously enhancing well defined mass.

Although teratoid Wilms has been described as a non-aggressive, non-metastatic tumour with a favourable prognosis (1-4), a total of 4 cases of metastases have been reported (3,7). Teratoid variant of Wilms has been portrayed as chemo-resistant by many (1,2,6,7). Neoadjuvant chemotherapy has been unable to produce a cytoreductive response in most cases (2,6,7). This may be attributed to the well differentiated nature of the teratomatous elements in these cases (2,10).

Despite 11 cycles of chemotherapy, the size of the tumour didn't respond in our case which is similar with the chemo-resistant nature of this pathology reported from other centres. There was a band like necrosis within the left sided tumour demarcated by macrophages which probably represents the effect of the chemotherapy upon an immature component.

The Société Internationale d'Oncologie Pédiatrique (SIOP) guidelines for the treatment of Wilms tumour advises preoperative chemotherapy or radiation therapy, followed by surgery. No specific treatment strategy has been proposed to those with teratoid Wilms. Our patient received 10 cycles of preoperative actinomycin-D (dactinomycin) and vincristine regimen. Cyclophosphamide and etoposide was given later. There was little response to these chemotherapeutic agents. The response of our case to the nephron sparing surgery is excellent. The baby is thriving well with no sign of metastasis with good renal function after two years of follow up. This is in line with the nonaggressive, non-metastatic behaviour of teratoid Wilms' tumour reported from other centres.

Conclusion: We presented a case of rare teratoid Wilms' tumour. Teratoid Wilms' tumour is described as a non-aggressive, non-metastatic tumour with a favourable prognosis. The management of bilateral teratoid WT depends on the individual clinical scenario, the ultimate aim being tumor eradication with renal preservation through surgical intervention. Despite the treatment protocol suggested both by SIOP and National Wilms tumor study group (NWTSG) for a neoadjuvant and adjuvant chemoradiotherapy for a bilateral Wilms tumor, Nephron sparing surgery is the only modality available for teratoid Wilms' tumor and has proved to be effective.

Ethics approval and consent to participate: Ethical clearance was obtained from the IRB of Department of Surgery

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