

Teratoid Wilms' tumor – A rare renal tumor

Biswanath Mukhopadhyay, Ram Mohan Shukla, Madhumita Mukhopadhyay¹, Sabitri Mandi¹,
Dipankar Roy, Malay K. Bhattacharya

Department of Pediatric Surgery, NRS Medical College, ¹Department of Pathology,
Institute of Postgraduate Medical Education and Research, Kolkata, India

Abstract Teratoid Wilms' tumor is an extremely rare renal tumor. We report a case of unilateral teratoid Wilms' tumor in a 4-year-old girl. The patient was admitted with a right-sided abdominal mass. The mass was arising from the right kidney. Radical nephrectomy was done and the patient had an uneventful recovery. Histopathology report showed teratoid Wilms' tumor.

Key Words: Chemotherapy, radical nephrectomy, teratoid Wilms' tumor, teratoma, unilateral, Wilms' tumor

Address for correspondence:

Dr. Biswanath Mukhopadhyay, 7E, Dinobandhu Mukherjee Lane Sibpur, Howrah, India. E-mail: drbmukhopadhyay@yahoo.co.in

Received: 09.08.2010, Accepted: 10.10.2010

INTRODUCTION

Variend *et al.* termed teratoid Wilms' tumor in a nephroblastoma with significant heterologous components.^[1] Fernandes *et al.* defined teratoid Wilms' tumor in which heterologous components like adipose tissue, glial tissue, muscles, cartilage or bone were at least 50% of the neoplasia.^[2] According to Beckwith criteria, a renal teratoma should be entirely within the renal capsule, and there should be clear evidence of renal component and other tissues. The pathogenesis of this rare condition is debatable and most probably the heterologous tissue components arise from primitive metanephric blastema.^[3] To the best of our knowledge, 17 cases have been described in English literature and most of them are bilateral.^[4-6] We present an additional case and review the literature.

CASE REPORT

A 4-year-old girl was admitted in the Pediatric Surgery

department with a mass in the right side of the abdomen present since 3 months, intermittent fever for 3 weeks, and hematuria for 1 week. There was no dysuria. Her blood pressure was normal. A right-sided abdominal mass measuring 11 cm × 10 cm was palpable in the right lumbar region. It did not cross the midline. There were no associated anomalies. Urine examination showed microscopic hematuria and culture was sterile. Hemoglobin was 9.7 gm/dl. Chest radiograph, renal, and liver function tests were within normal limit.

Serum levels of alpha-fetoprotein, vanillyl mandelic acid were within normal limits.

Ultrasonography (USG) of the abdomen showed a large heteroechoic mass measuring 11.6 cm × 10.2 cm × 8.9 cm, arising from the lower pole of the right kidney. The left kidney was normal. There was no involvement of the blood vessels. Contrast-enhanced computed tomography (CT) of the abdomen showed heterogeneous mass in the mid and lower pole of the right kidney and measured 11.7 cm × 10.3 cm × 9 cm [Figure 1]. Good excretion of contrast with splaying of pelvicalyceal system was seen. There was no enlargement of local lymph nodes. Laparotomy was done through a supraumbilical transverse transperitoneal incision. Right radical nephrectomy was done. The contralateral kidney was examined and found to be normal. The patient had a smooth postoperative recovery.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/0974-7796.84959



Figure 1: Contrast-enhanced computed tomography (CT) of the abdomen showed heterogeneous mass in the mid and lower pole of the right kidney and measured 11.7 cm × 10.3 cm × 9 cm



Figure 2: Cut open specimen showing the tumor which was involving almost whole of the kidney with cystic changes in some parts

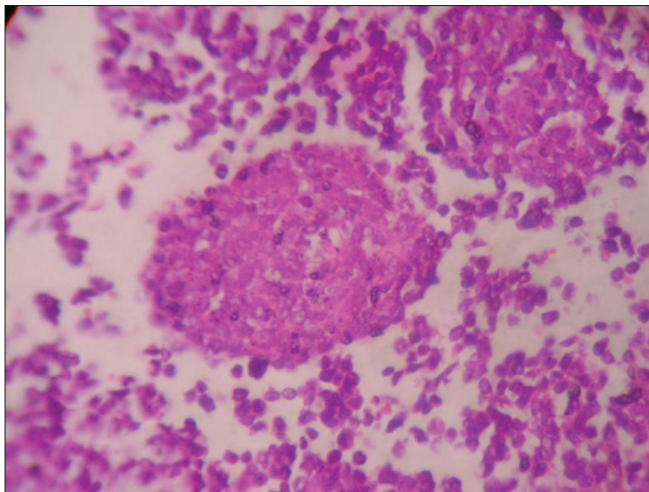


Figure 3: Photomicrograph showing triphasic pattern of Wilms' tumor with blastematos, epithelial and stromal components

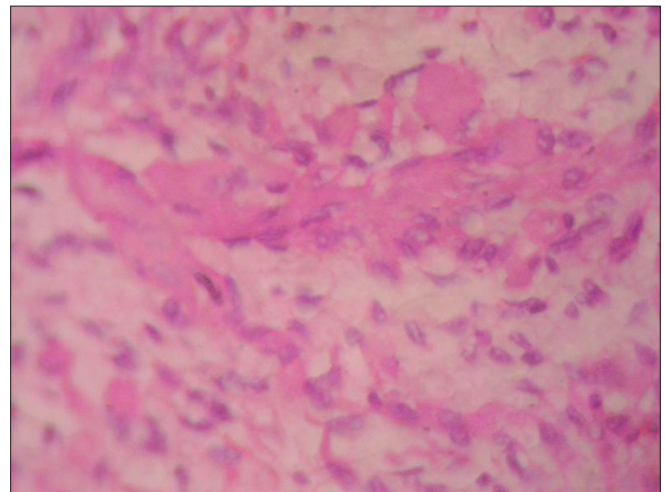


Figure 4: Photomicrograph showing prominent heterologous components which include rhabdomyoblasts

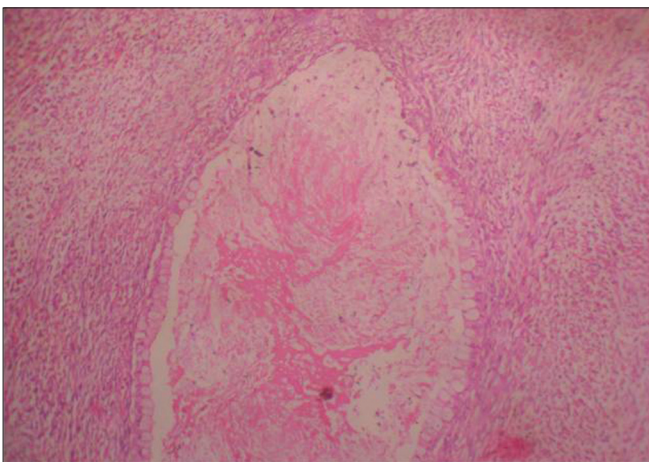


Figure 5: Photomicrograph showing mucinous epithelium in teratoid Wilms' tumor

Pathology

Removed kidney weighed 465 grams and measured 13 cm × 12 cm × 9 cm. The tumor was involving almost whole of the kidney with cystic changes in some parts [Figure 2]. Microscopic examination showed the triphasic pattern of Wilms' tumor with blastematos, epithelial and stromal components [Figure 3], prominent heterologous components, which include rhabdomyoblasts [Figure 4] and different types of mature epithelial tissue showing mucus secreting epithelium [Figure 5].

Treatment

Laparotomy was done through a right supraumbilical transverse transperitoneal incision. Right radical nephrectomy was done. The contralateral kidney was found to be normal. The patient

had a smooth recovery and was started on chemotherapy (Vincristine and Actinomycin D).

DISCUSSION

Teratoid Wilms' tumor, a rare histological variant of classical Wilms' tumor, shows a predominance of teratoid elements.^[2,4] This tumor has been reported mainly in pediatric patients.^[7] Only one adult patient with teratoid Wilms' tumor has been reported by Seo *et al.*,^[7] Literature search revealed 17 cases of teratoid Wilms' tumor in children. In a review,^[4] most of the patients were diagnosed between 2 to 4 years of age and 6 bilateral cases were observed. Wilms' tumor, retroperitoneal tumor, neuroblastoma, hydronephrosis, and renal cysts must be included in the pre operative differential diagnosis. Characteristic radiological features include calcific densities and stippling areas of attenuation indicating adipose tissue.^[5] Ruchika Gupta *et al.*,^[6] showed stroma predominant Wilms' tumor with teratoid features. They have concluded that the pediatric surgeon and the pathologist should be aware of this appearance of nephroblastoma. There are isolated case reports where alpha-fetoprotein was elevated in teratoid Wilms' tumor.^[8] Alpha-fetoprotein level was normal in our patient. Teratoid Wilms' tumor is usually not aggressive and prognosis is comparatively good if the tumor is excised completely.^[4] For this reasons, surgery seems to be the best treatment.^[9,10] Chemotherapy has been recommended in the cases of teratoid Wilms' tumor regardless of the tumor size, stage, age at diagnosis and histology. Our patient was 4 years old and had stage II/favorable histology weighing less than 500 gms. Microscopy showed the triphasic pattern of Wilms' tumor and prominent heterologous components and different types of mature epithelial tissue. Histopathology did not show capsular invasion, invasion of renal sinus or intrarenal vessels and there

was no anaplastic element. Treatment of teratoid Wilms' tumor has not yet been established because of its rarity and presence of varying tumor components. Resistance to chemotherapy and radiotherapy has been reported due to the presence of mature heterologous components.^[4,11]

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How to cite this article: Mukhopadhyay B, Shukla RM, Mukhopadhyay M, Mandi S, Roy D, Bhattacharya MK. Teratoid Wilms' tumor - A rare renal tumor. *Urol Ann* 2011;3:155-7.

Source of Support: Nil, **Conflict of Interest:** None.

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