# Extrarenal teratoid Wilms' tumour

**Chowhan A K, Reddy M K, Javvadi V, Kannan T**


## ABSTRACT

**We report an unusual case of extrarenal teratoid Wilms' tumour in a 15-month-old male child. The tumour was retroperitoneal in location and consisted of triphasic Wilms' tumour elements, along with the presence of heterologous components. The heterologous teratoid elements were composed of predominantly glandular epithelium with the presence of focal skeletal muscle, adipose and neuroglial tissues. Although extrarenal Wilms' tumours have been documented in the literature, only a few cases have been noted to date. We present the relevant clinical, radiological, histomorphological, histochemical and immunohistochemical features of this rare tumour, and discuss the various theories of its histogenesis.**


**Keywords: kidney neoplasms, retroperitoneal space, teratoma, urologic neoplasms, Wilms' tumour**

*Singapore Med J 2011;52(6):e134-e137*

## INTRODUCTION

Wilms' tumour is a malignant neoplasm of primitive metanephric cells that retain their embryonic potential.[1] The classical location for Wilms' tumour is the kidney. However, cases with typical morphologic features of Wilms' tumour have been recorded in extrarenal sites, including the retroperitoneum, sacrococcygeal region, testis, uterus (sometimes presenting as a cervical polyp), inguinal canal and mediastinum.[2-6] Teratoid Wilms' tumour is a rare histologic variant of the classical Wilms' tumour, containing predominantly heterologous tissues.[7] Our patient presented with an extrarenal teratoid Wilms' tumour, an even rarer entity, located in the retroperitoneum. The first case of teratoid Wilms' tumour was diagnosed in 1984 by Variend et al.[8] Up till now, few cases of extrarenal teratoid Wilms' tumour have been reported in the literature. To our knowledge, a few cases of teratoid Wilms' tumour of renal origin have been reported from India, but none involving extrarenal sites has been noted so far.[7,9]

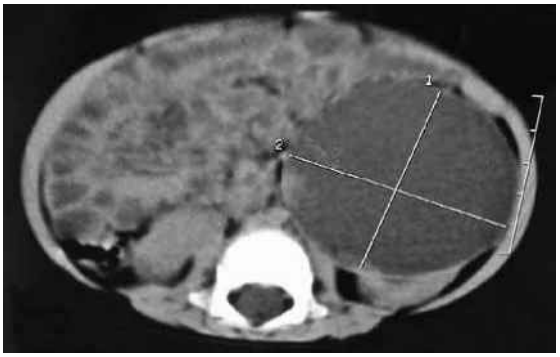

**Fig. 1** CT image shows the solid-cystic mass located anteriorly and overlying the lower pole of the left kidney with hydronephrosis.


**Fig. 2** Photograph of the cut section of the gross cystic globular mass with solid polypoidal projections on the slimy inner wall.

## CASE REPORT

A 15-month-old boy presented with mild abdominal distention, which had slowly increased in size since the past two months. On physical examination, a palpable, non-tender mass was noted in the left lumbar region. The rest of the clinical examination was unremarkable. No congenital anomaly was detected. The laboratory findings, including haematological, biochemical and urine examinations, were within normal limits. Serum alpha-foetoprotein, measured by radioimmunometric assay, was 10 ng/ml (normal < 15 ng/ml). Ultrasonography of the abdomen revealed a round hypoechoic lesion measuring 6.6 cm × 5.9 cm in the retroperitoneum, located just below and close to the left kidney, along with moderate

**Department of Pathology, Sri Venkateswara Institute of Medical Sciences, Tirupati 517507, Andhra Pradesh, India**

Chowhan AK, MD
Assistant Professor

Reddy MK, MD
Professor

**Department of Medical Oncology**

Kannan T, MD, DM
Professor

**Department of Paediatric Surgery, Sri Venkateswara Medical College, Alipuri Road, Tirupati 517520, Andhra Pradesh, India**

Javvadi V, MS, MCh
Professor

**Correspondence to:**
Dr Chowhan Amit Kumar
Tel: (91) 0949 0398 668
Fax: (91) 0877 2286
Email: chowhanpath@gmail.com

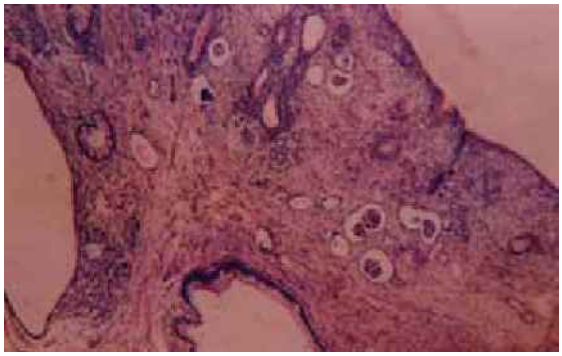

**Fig. 3** Photomicrograph shows abortive glomeruli and primitive tubules present in association with the mucinous columnar-lined cystically dilated glandular elements (Haematoxylin & eosin, × 100).

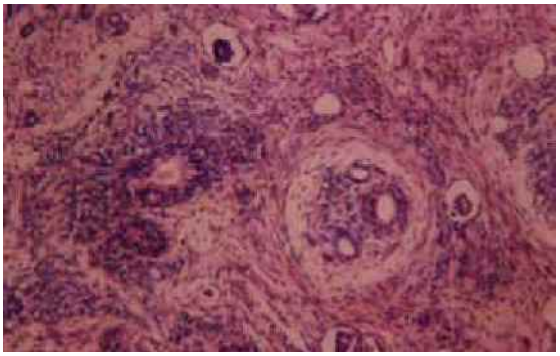

**Fig. 5** Photomicrograph shows islands of blastemal elements, abortive glomeruli and primitive tubules (Haematoxylin & eosin, × 200).

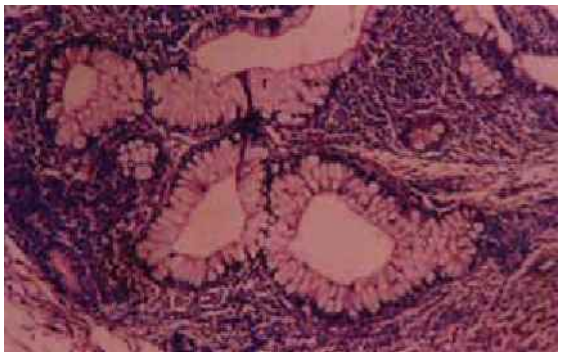

**Fig. 4** Photomicrograph shows islands of blastemal elements with neoplastic glandular structures lined by tall columnar mucinous epithelium (Haematoxylin & eosin, × 400).

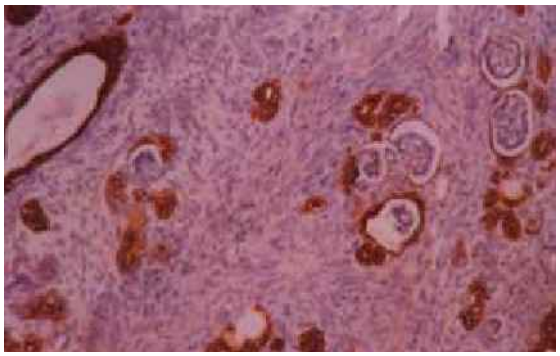

**Fig. 6** Photomicrograph shows epithelial membrane antigen displaying strong positivity for primitive tubular elements and Bowman's capsule of abortive glomeruli (IHC stain, × 200).

hydronephrosis of the left kidney. Contrast-enhanced computed tomography (CT) imaging showed a large cystic mass measuring 6.9 cm × 5.7 cm in close approximation and anterior to the lower half of the left kidney, with small areas of solid components present toward the inner peripheral portion of the mass (Fig. 1). Radiologically, the possibility of a retroperitoneal teratoma compressing the left ureter leading to hydronephrosis of the left kidney was suggested. Intravenous pyelogram displayed delayed filling and excretion of contrast with hydronephrotic changes in the left kidney. The chest radiograph was normal.

Perioperatively, the retroperitoneal mass was found to be adherent to the outer surface of Gerota's fascia of the left kidney. It was overlying and causing pressure on the left ureter, resulting in mild hydronephrosis. The left renal vessels were the feeding vessels for the tumour. The surgically resected specimen revealed a thin-walled globular cystic mass that measured 6 cm × 6 cm, with smooth, mildly transparent capsule showing engorged prominent blood vessels on its outer surface. The cut section showed a loculated cyst filled with mucinous

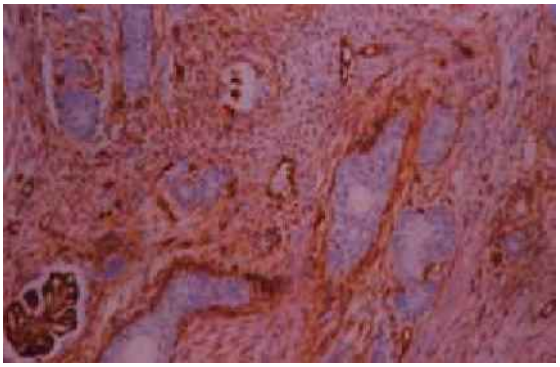

**Fig. 7** Photomicrograph shows vimentin antigen displaying strong positive mesenchymal elements, glomerular tuft and conspicuously negative for primitive tubular elements and Bowman's capsule of abortive glomeruli (IHC stain, × 200).

material, making the inner surface of the cyst wall slimy and glistening, with the presence of solid polypoidal projections seen in some areas (Fig. 2).

Multiple bits taken from the encapsulated solid cystic tumour showed an extensive (approximately 70%) presence of heterologous structures comprising predominantly glandular structures lined by mucinous

columnar epithelial cells, some of which were abnormally dilated with retained mucin (Figs. 3 & 4). The cystically dilated glands were lined by a single layer of flattened cuboidal epithelium. There was focal presence of adipocytes, neural and skeletal muscle tissues. Triphasic Wilms' tumour elements formed the remaining part of the tumour, displaying islands of blastemal elements, whorls of undifferentiated spindle to plump cellular mesenchymal elements and a fair number of abortive glomeruli and primitive tubules representing the epithelial element (Fig. 5). With these microscopic features, a histopathological diagnosis of teratoid Wilms' tumour was made, and the tissue was subjected to various histochemical and immunohistochemical stains. Mucicarmine stain prominently displayed mucinous secretions and the mucinous columnar lining epithelium of the glands. The mesenchymal tissue was well highlighted by Massons Trichrome stain. Immunohistochemical stain for cytokeratin and epithelial membrane antigen (EMA) showed strong positivity for columnar epithelial cells lining the glandular elements, focal positivity for primitive tubules and Bowman's capsule of abortive glomeruli. Vimentin showed positivity for the mesenchymal tissue, along with the vascular tuft of the abortive glomeruli. An interesting finding was focal, mild positivity for the blastemal elements with cytokeratin, EMA, vimentin, nonspecific enolase (NSE), S-100 protein and desmin (Figs. 6 & 7).

According to the National Wilms' Tumour Study (NWTS), the tumour was stage II, as it was located outside the kidney and excised completely. Apart from excision of the tumour, our patient was put on a two-drug chemotherapeutic regimen comprising vincristine and actinomycin D, as per the NWTS protocol of management.[14] Six months after the therapy, our patient was doing well, and close follow-up has been instituted in order to monitor any future recurrence or metastasis.

## DISCUSSION

The classical Wilms' tumour has a triphasic histologic pattern, with blastemal, mesenchymal and epithelial components. The heterotopic mesodermal elements are commonly seen but usually involve a minor part of the neoplasm. According to Variend et al's[8] criteria for diagnosing teratoid Wilms' tumour, the heterotopic elements should have dominant presence (> 50% of the total area). As the tumour in our patient displayed mainly (approximately 70%) mature teratomatous elements comprising predominantly mucinous columnar epithelial-lined glands and cysts, along with classical triphasic elements of Wilms' tumour and was extrarenal in location, the most appropriate diagnosis of extrarenal teratoid Wilms' tumour was made.

Various theories have been put forth by different authors to explain the origin of this complex neoplasm. Wilms postulates that the classical triphasic tumour arises from the remnants of undifferentiated mesodermal tissue before the mesodermic derivatives of myotome, sclerotome and nephrotome develop.[2] The explanation proposed by Variend et al for teratoid Wilms' tumour of the kidney is a direct origin of the diverse epithelia, together with mesenchymal elements from totipotent primitive nephrogenic blastema, probably occurring at a stage in development when totipotency of the metanephric blastema is likely to be appreciably greater.[8] Hou and Azzopardi considered a possible metaplasia of collecting tubules giving origin to the diverse teratoid heterologous elements.[10] The finding of extrarenal Wilms' tumours outside the course of germ cell migration, such as those arising in the inguinal area, would suggest that they may not necessarily be teratoid but may have originated from misplaced primitive nephrogenic blastema.[11] The extrarenal location of teratoid Wilms' tumour has led to the debate on whether the origin is embryonic or neoplastic, but most authors believe the origin of this tumour to be the embryonic remnants of mesonephric tissue.[12,13]

As the location of the tumour in this case was consistent with the embryological mesonephric remnant, we strongly support the theory proposed by Variend et al[8] that the diverse teratomatous elements, together with the triphasic elements of Wilms' tumour, are differentiated from the primitive nephrogenic blastema, as evidenced by focal positivity shown by the blastemal cells for the various immunohistochemical stains like cytokeratin, EMA, vimentin, NSE, S-100 protein and desmin, as was the case in our patient.

We present this unusual case of extrarenal teratoid Wilms' tumour so that paediatric clinicians as well as pathologists are made aware of the rare location of this tumour, and because of the strong immunohistochemical support that it gathers in favour of the pathogenetic theory by Variend et al.[8]

## REFERENCES

1. Ashworth MT, Pizer BL, Oakhill A, Spicer RD, Berry PJ. A teratoid Wilms' tumour with raised serum alpha-fetoprotein level. Pediatr Pathol Lab Med 1996; 16:853-9.
2. Tebbi K, Ragab AH, Ternberg JL, Vietti TJ. An extrarenal Wilms' tumor arising from a sacrococcygeal teratoma. Clin Pediatr (Phila) 1974; 13:1019-21.
3. Ho J, Ma L, Wong KC. An extrarenal Wilms' tumour arising from

an undescended testis. Pathology 1981; 13:619-24.
4. Lüchtrath H, de Leon F, Giesen H, Gök Y. Inguinal nephroblastoma. Virchows Arch A Pathol Anat Histopathol 1984; 405:113-8.
5. Benatar B, Wright C, Freinkel AL, Cooper K. Primary extrarenal Wilms' tumor of the uterus presenting as a cervical polyp. Int J Gynecol Pathol 1998; 17:277-80.
6. Badillo AT, Kreiger PA, Schmitz KR, et al. Wilms' tumour arising within a mediastinal teratoma. J Pediatr Surg 2006; 41:e1-5.
7. Bakshi N, Mansoor I, Venkataramu NK, Katariya S. An unusual renal malignancy of childhood: unilateral teratoid Wilms' tumour. Pediatr Pathol Mol Med 2003; 22:435-41.
8. Variend S, Spicer RD, Mackinnon AE. Teratoid Wilms' tumor. Cancer 1984; 53:1936-42.
9. Parikh B, Trivedi P, Shukla K. A unilateral teratoid Wilms' tumor with raised serum alpha-fetoprotein level. Indian J Pathol Microbiol 2007; 50:317-9.
10. Hou LT, Azzopardi JG. Muco-epidermoid metaplasia and argentaffin cells in nephroblastoma. J Pathol Bacteriol 1967; 93:477-81.
11. Akhtar M, Kott E, Brooks B. Extrarenal Wilms' tumor: report of a case and review of the literature. Cancer 1977; 40:3087-91.
12. Koretz MJ, Wang S, Klein FA, Lawrence W Jr. Extrarenal adult Wilms' tumour. Cancer 1987; 60:2484-8.
13. Park HR, Park CY, Choi NK, Park YE. A case of retroperitoneal teratoma with nephroblastoma. J Korean Med Sci 1991; 6:95-102.
14. Bhatnagar S. Management of Wilms' tumor: NWTS vs SIOP. J Indian Assoc Pediatr Surg 2009; 14:6-14.