

ADULT WILMS' TUMOUR: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Wilms' tumour is a common tumour in children but it occurs occasionally in adults. We present a case report of adult Wilms' tumour in a 17 year female.

Key words: Adult, Wilms' tumour

INTRODUCTION

Wilms' tumour is extremely rare in adults, representing only 0.5% of all renal neoplasms.¹ However it is the most common abdominal tumour in children accounting for approximately 5-6% of the neoplasms in children.² Patients with certain congenital defects, including anomalies of the male genitourinary system, aniridia, Deny-Drash syndrome, hemi hypertrophy and Beckwith-Wiedemann syndrome are at increased risk of developing this cancer.³

CASE REPORT

A 17 year old female presented to surgical outpatient department with haematuria, right flank pain, and fever. There was no history of trauma and patient did not have any congenital anomaly. On examination she had a lump in the right lumbar region.

Haemogram and biochemical parameters were within normal limits. Urine examination showed presence of RBCs in the sediment, there were no malignant cells. Her chest X-Ray was unremarkable.

B mode ultrasonography revealed enlarged right kidney with moderate hydronephrosis, renal pelvis was filled with a hypoechoic mass, and there was a heterogenous mass in lower pole of the kidney with septate sub capsular collection.

Contrast enhanced computed tomography (CT) showed a 6x6 cm round, heterogenous, minimally

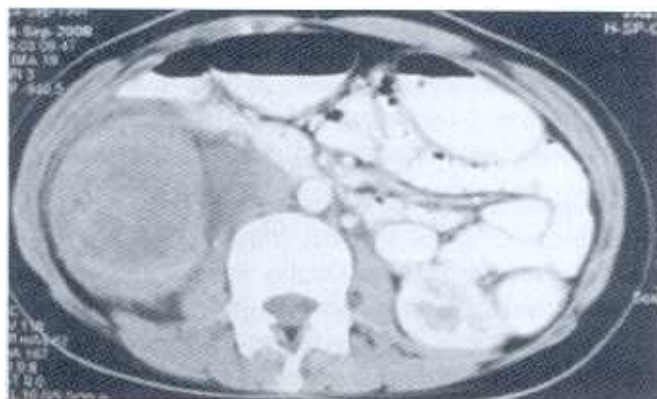


Fig. 1: Contrast enhanced CT scan showing hypodense in the lower pole of the right kidney.

enhancing mass in lower pole of right kidney, and minimally enhancing heterogenous mass in sub capsular region (Fig 1). There was no significant lymphadenopathy in retro-peritoneum. Right renal vein and IVC were unremarkable.

Ultrasound guided FNAC from lower pole mass of right kidney was suggestive of tumour, with recommendation for biopsy to ascertain detailed histopathology.

On surgery, tumour was seen infiltrating the whole of the renal parenchyma, hemorrhagic collection in subcapsular space and blood clots filling up the pelvicaliceal system were also seen. A total right nephrectomy was performed.

Gross cut section of kidney revealed that almost whole of renal parenchyma was replaced by a tumour measuring 12.5x6x6cm. The growth was solid, grey brown with focal grey yellow areas. The pelvis was dilated and showed presence of tumour deposits. The hilar vessels were unremarkable. Histopathology revealed a triphasic

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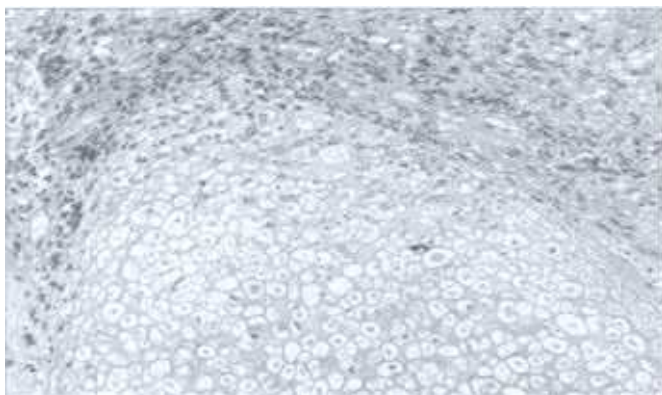


Fig. 2: Wilms' tumour showing cartilage differentiation (H&E, 100X)

tumour composed of blastemal and stromal components with foci of epithelial component which were seen as abortive tubules. The blastemal cells were seen as nodular aggregates of closely packed cells with high N/C ratio and coarse chromatin. The stromal component was dispersed as fascicles of anaplastic spindle shaped cells lying in a myxoid stroma. Focal cartilage differentiation was noted along with large areas of haemorrhage and necrosis (Fig 2). Many multinucleated tumour giant cells, multipolar atypical mitotic figures (Fig 3) and focal intra and extracellular hyaline globules were also seen (Rhabdoid differentiation). The diagnosis was Wilms' tumour (Nephroblastoma) with anaplastic nuclear features, confirmed on histopathology.

DISCUSSION

Wilms' tumour, named after the German surgeon Dr. Carl Max Wilhelm Wilms, is also referred to as nephroblastoma. The tumour probably develops from primitive metanephric blastemal remnants of kidney. The histological appearance is characterized by marked structural diversity. Classic Wilms' tumour is composed of three types of cells – blastemal, stromal, and epithelial cells. It is rare in adults, but it is the most common renal tumour in children under 6 years of age. Adult Wilms' tumour may have a more aggressive clinical course and a higher tumour stage at presentation compared with that in children³. Most adults present with local pain and haematuria, in contrast to the palpable boggy mass which is more common presentation in children. In adults, Wilms' tumour is larger and ill-defined, with areas of necrosis and haemorrhage as seen in the index case. About half of the patients have stage 3 or 4 disease at presentation. A child with Wilms' tumour may present with vomiting and fever. The diagnostic criteria defining adult nephroblastoma were described by Kilton et al.⁴ This disease is difficult to differentiate from

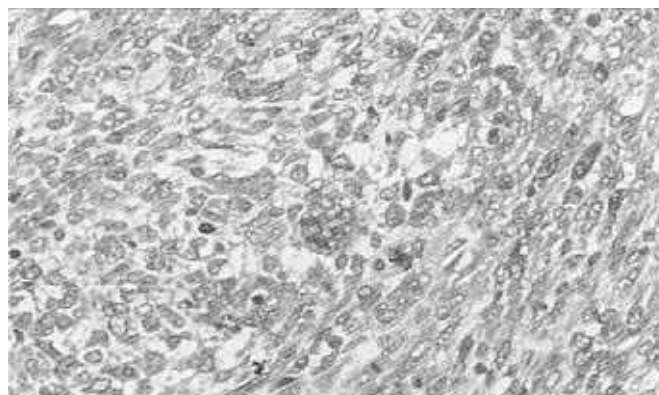


Fig. 3: Wilms' tumour showing anaplastic cells and tumour giant cells (H&E 200X)

renal cell carcinoma based only on imaging techniques, though preoperative diagnosis by imaging may be suggestive in about 75-80% of cases. There are about ten cases of extrarenal Wilms' tumour in adults documented in literature; four of them were in the retroperitoneal region, two each in the ovary and endometrium, and one each in ovo-testis and prostate⁴.

Ultrasonography is the recommended first-line imaging modality because it provides a panoramic view of the abdomen. CT can also visualize pelvic and abdominal structures as well as lymph nodes. Ultrasound observation of a rapidly growing abdominal mass in a young patient, with heterogeneous contrast uptake and surrounded by a pseudocapsule on CT is suggestive of Wilms' tumor¹. Calcification may be a sign of slow tumour growth and possibly a favourable prognostic sign in cases of adult Wilms' tumor⁵.

Arteriography characteristically shows a hypovascular mass with neoformed blood vessels exhibiting a zigzag pattern¹. Magnetic resonance imaging (MRI) is noninvasive, does not employ ionizing radiations and has the potential for providing the same information as computed tomography, sonography, liver spleen radionuclide scanning and excretory urography. Although expensive, magnetic resonance will be cost-effective if it can replace all the above techniques⁶. The definitive diagnosis of Wilms' tumour can only be made by surgical resection or biopsy.

Adult Wilms' tumour is diagnosed based on criteria given by Kilton, Mathews and Cohen.⁴ These include (a) the tumour under consideration should be a primary renal neoplasm; (b) presence of primitive blastemic spindle or round cell component; (c) formation of abortive or embryonal tubules or glomerular structures; (d) no area

of tumour diagnostic of renal cell carcinoma ; (e) pictorial confirmation of histology and (f) patient's age >15 years. In 1980 they reported a series of 35 cases of adult Wilms' tumour with all the above said criteria⁴.

Aggressive treatment, including radical surgery, chemotherapy and irradiation of the tumour bed, is considered necessary. The routinely used chemotherapeutic agents are vincristine, actinomycin-D, doxorubicin and ifosamide. Satisfactory results have also been published with cisplatin and etoposide in stage IV patients and patients in progression after conventional chemotherapy⁷. In 2004, Reinhard reported their experience with 30 cases of adult Wilms Tumor⁸. A complete remission was achieved in 24 of their patients. Event-free survival was 57%, and overall survival was 83%. It was concluded that adults can be cured in a high percentage by a multimodal treatment according to paediatric protocols.

National Wilms' Tumour Study (NWTs) was established in USA in 1969. An update from NWTs group about treatment outcomes in adults with favourable histology Wilms' tumour (FHWT) described 45 patients treated in the modern era. The overall survival rate was 82%. NWTs and other studies have recommended multimodal therapy for the disease with surgery, chemotherapy for 15 months and tumour bed irradiation in the case of stage 3 disease.⁹

To conclude in an adult patient presenting with flank pain, and a renal mass on imaging, though rare, possibility

of Wilms' tumour should also be considered. Although the prognosis in adult Wilms' tumour is poorer as compared to that in children, the outcome for adult patients is improving steadily with multimodal therapy.

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