

## Case Report

# A rare case of adult Wilms tumor: case report and literature review from a tertiary centre

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## ABSTRACT

Nephroblastoma (Wilms' tumour) in adult population is extremely rare scenario and has a poorer prognosis than paediatric Wilms' tumour with an incidence of about 0.2 per million per year in the USA and Europe. Clinical presentation is also distinct between children and adults. In adults, manifestation is usually hematuria and abdominal pain while paediatric cases frequently present with an asymptomatic abdominal lump. It is arduous to demarcate adult Wilms' tumour from renal cell carcinoma based on radiology alone. The final diagnosis in adult cases is often fortuitous following radical nephrectomy for presumed adult Renal cell carcinoma (RCC). Due to the lack of published discussion, there are no standard protocols for the management of adult Wilms tumour (WT) hence it is managed as per paediatric WT. In view of this, we report a case of adult WT in a 32-years-old female, who was diagnosed propitiously following right radical nephrectomy for an assumed right renal mass.

**Keywords:** Nephroblastoma, Adult Wilms' tumor, Renal tumor

## INTRODUCTION

Renal tumors encompass 7-8% of all tumours in the first decade of life. Wilms' tumor (WT) is the most common (85%) of the lot, followed by renal cell carcinoma (RCC) (3-5%) and mesoblastic nephroma (3%) to mention a few frontrunners. The annual incidence rate in children under 15 years is 7 to 10 cases per million.<sup>1</sup> An impeccable histological report along with an authentic staging of these tumours is invaluable because their management is very complex and different from each other.

Being rare, they represent a management challenge and it's important to deal with them effectively based on deft clinical knowledge and precision via multicentre collaborative trials which include centralised pathology reporting system in order to verify the diagnosis and stage of cases entered on the trials.<sup>2</sup> WT also known as

nephroblastoma, is an extremely uncommon tumor with sparse meaningful mentions in the literature. A few latest reviews mention a number close to 300 cases being reported.<sup>3</sup> WT finds its root in being an embryogenic tumor, derived from nephrogenic blastemal cells, which mimics the renal histology and usually exhibits various patterns of differentiation and it's the most common paediatric renal tumor, occurring preferably in the first 5 years of life with peak incidence between 3 and 4 years of age.<sup>4</sup> WT is extremely uncommon in adults and represents less than 1% of all diagnosed renal tumours. The clinical scenario of adult WT includes abdominal pain and hematuria and it is not uncommon for these to have distant metastasis to the lungs and liver.<sup>5</sup>

Adult and pediatric WT don't have a significant radiological and histological difference but share the classical triphasic histopathological features such as

stromal, epithelial and blastemal features under the lens. The histological pattern is identified by marked structural diversity, and the occurrence of all three types in the same case is uncommon. Adult WT is diagnosed based on well-endowed criteria described by Kilton and associates.<sup>6</sup> They include the following: 1) patient's age should be >15 years; 2) the tumour in focus should have a primary renal origin; 3) presence of primitive blastemic spindle component; 4) spatial confirmation of histology; and 5) formation of embryonal tubules or carcinoma. Management is established on astute surgical resection of the tumor, followed by the risk classification and thus making decision further chemo-radiotherapy.<sup>7,8</sup> A plethora of factors including the benightedness of pathologists and urologist with adult WT, lack of standardized protocols and delay in appropriate risk-adapted therapy leads to a dismal outcome. Adult patients are usually managed on the lines of available treatment protocols used for children, which include protocols developed by the International Society of Pediatric Oncology (SIOP) and National Wilms' Tumour Study Group (NWTG).<sup>9</sup>

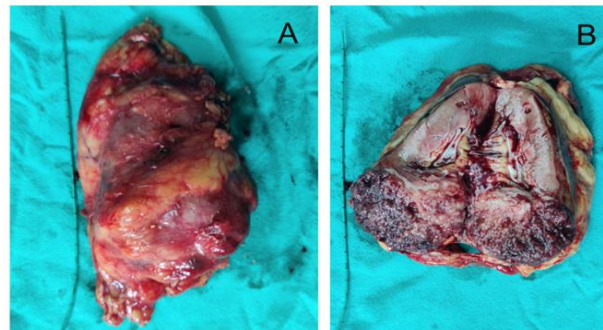
## CASE REPORT

A 32 years old female, having a 3 months history of right upper quadrant pain presented to the urology OPD with no preceding symptoms of hematuria, abdominal trauma, or any other systemic symptoms. The family history was not significant and the patient had no significant past surgical or medical history. She was a non-smoker, non-alcoholic and not on any sort of tobacco consumption. The concerned urology team prescribed an ultrasound of whole abdomen after thorough and detailed physical examination suggestive of right renal angle fullness, which revealed a large right renal mass measuring 7.6×8 cm arising from the lower pole. Proceeding investigation with contrast enhanced computed tomography (CECT) scan of the abdomen with intravenous contrast revealed mass lesion within the right lower pole measuring up to 8.4×8.5×7.2 cm with no evidence of extra renal involvement.

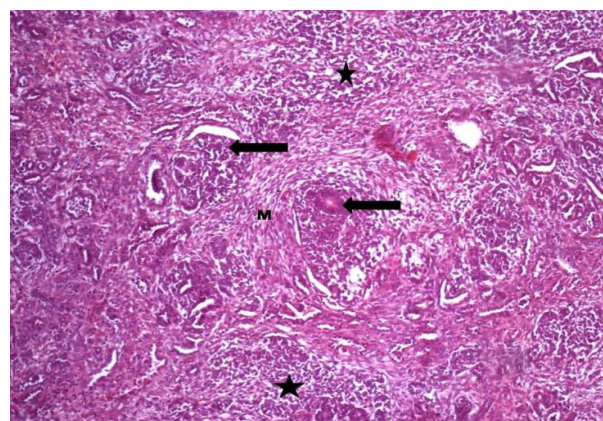
A preliminary interpretation of right renal cell carcinoma was made based on the radiological investigations. The resected right kidney measured 12×10×9 cm (enclosing perirenal Gerota's fascia), (Figure 1) with right ureter of 6 cm of length. Near the lower pole there was a unicentric rounded mass measuring 8×7×7 cm that exhibited, on cut surface, a tan appearance with soft consistency areas, due to the presence of haemorrhage and necrosis and the organ capsule appeared grossly intact. On cut section the tumor was identified in lower part of kidney measuring 8×6.5×7 cm. The tumor showed variegated white grey solid area, few yellowish area and necrosis was also identified.

Tumour was unifocal and seemed to involve the cortex and medulla. Tumor was not invading the perinephric adipose tissue and grossly the renal vessels did not show thrombosis. Histologically, the tumor showed diffuse necrotic and hemorrhagic changes, with presence of undifferentiated blastemal cells and cells of epithelial and

stromal lineages. The epithelial line consisted mainly of tubular structures while the stromal component was made of spindle cells with elongated nuclei and storiform pattern of growth (Figure 2). The IHC=4420 (4456 E) pattern comprised of Pan CK- tumor cells are positive for Pan CK; CK-7- negative; and WT-1- many tumor cells are positive.



**Figure 1: Showing specimen of resected right kidney with longitudinal cut section with inferior polar location of the tumor.**



**Figure 2: Triphasic Wilms tumor displaying epithelial (arrows), blastemal (star marked) and mesenchymal component (M) (H and E stain, at 100X).**

Thus, the final diagnosis was an adult WT. A FISH analysis was made for evaluate the 12p chromosome status which turned up to be negative. The analysis further showed loss of heterogeneity (LOH) for 1p 16q, which is associated with poor outcomes in paediatric WT. The patient was discharged after 8 days with a good kidney function (urea 4.8 mmol/l, creatinine 9 mg/dl, hemoglobin 12.7 g/dl) and no significant complications were observed. Based on chromosomal LOH for 1p, 16q and unfavourable histology the tumour was considered high risk, and chemotherapy was initiated as per the COG protocol, including early radiation to the tumour bed, and multi-agent chemotherapy including doxorubicin, actinomycin D and vincristine. Radiation and chemotherapy were commenced concurrently within 6 weeks of surgery. The patient was followed up at regular intervals with CECT abdomen scans. There has been no clinical and

radiological recurrence at the end of 9 months post-operatively.

## DISCUSSION

Great strides have been made in the treatment of paediatric WT although adult WT still needs further studies and improvement in outcomes. WT is the most common renal tumor and is most prevalent in the first 5 years of life. Clinical scenario such as spontaneous rupture and haemorrhage as presenting face of adult WT are rare. Adult WT is difficult to diagnose preoperatively as there are no specific radiographic tell-tale signs. Management of WT is multifocal involving radical nephrectomy, chemotherapy and occasionally radiotherapy.

Two accepted management protocols are the NWTS and SIOP. The SIOP follows the concept of pre-nephrectomy chemotherapy in order to shrink the tumour. The regimen also prevents intra-operative tumour rupture causing decreased overall tumor spillage and lower tumour stage which will require less overall treatment. Three groups, the South-west Oncology Group (SWOG), The Children's Cancer Study Group (CCSG), The Cancer and Leukemia Group B (CALGB) combined to create National Wilms Tumor Study (NWTS) in 1969. NWTS recommends upfront nephrectomy to define the accurate stage of the tumour and the histology, on which further treatment stratification is decided. The NWTS group has declared an overall survival rate of 82% in adults with favourable histology WT.<sup>10-12</sup>

A previous study group reported their experience regarding adult WT wherein 17 patients with adult WT and more than 16 years were treated according to paediatric WT guidelines. The overall survival in that series was 62.4% at 5 years. Similarly, another group reported their experience with 30 cases of adult WT. A complete remission was achieved in 24 cases with overall survival of 83% and event free survival of 57%.<sup>14-15</sup> There are a number of case series that suggest that the outcome for adult WT, which is both radio and chemo sensitive, has improved when paediatric WT protocols, including multimodality chemo and radiotherapy are adapted. A detailed and research oriented long-term follow up of adults WT should look into the risk of relapse but needs to minutely observe the toxicity profile of the chemotherapy agents.

## CONCLUSION

Adult WT is a rare entity and its diagnosis is generally made on postoperative pathology. Its prognosis is poorer than that of paediatric WT. Management is generally multimodal including radical nephrectomy and chemotherapy. Early relapse in the first year after treatment is a predictor of worse outcome. The need to register such patients to a consortium of international cases series will assist in the research and development of future

policy making and formulating management guidelines for this aggressive pathology.

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