

Case Report-IV

Adult Wilms' Tumour

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ABSTRACT

Wilms' tumour is most common malignant renal tumour of childhood. Rarely, it may be seen in the adults. We report a 35 year old male presenting with flank pain. Abdominal CT scan revealed a right renal mass and a clinical diagnosis of renal cell carcinoma was made. Nephrectomy was performed, and a final diagnosis of adult Wilms' tumour was made. This case is being reported on account of its rarity. Pertinent literature is being reviewed.

INTRODUCTION

Wilms' tumour, or nephroblastoma, is a malignant renal tumor that arises from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules. More than 90% of all Wilms' tumor cases occur before age 7, with peak incidence between age 3 and 4.¹

Case : A 35 year old male, presented with history of abdominal lump and pain of one month duration. CT scan abdomen showed 13x11x12mm well-defined lobulated heterogeneously enhancing mass lesion with internal necrotic areas involving the upper pole of right kidney indenting the inferior surface of right lobe of liver, displacing the second part of duodenum, head pancreas and IVC medially. Right renal vessels were displaced but were patent. Left Kidney was normal (Fig. 1). Fine Needle aspiration cytology from the mass

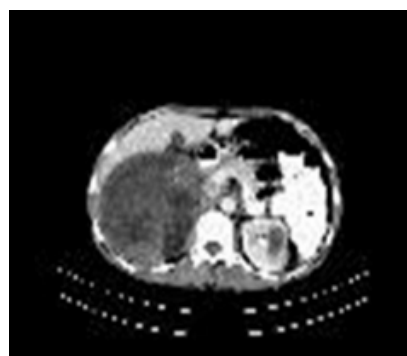


Fig 1: Lobulated heterogeneously enhancing mass lesion involving upper pole of right kidney

revealed round cell tumour. The hematological and biochemical parameters were within normal limits Chest x-ray-normal. Patient underwent right radical nephrectomy and histopathology revealed Adult Wilms' tumour, measuring 13.5 cm, triphasic tumour showing predominantly blastemal component with

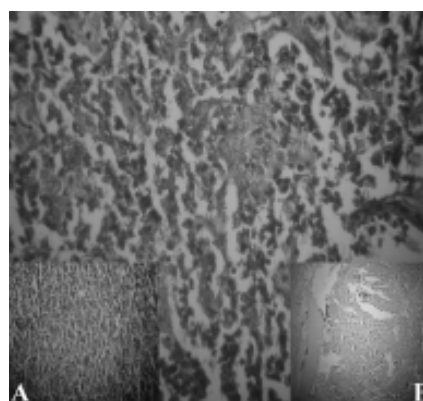


Fig 2: Histopathology (40X)of nephrectomy showing blastemal component with variable mesenchymal (inlet A) and epithelial component (inlet B).

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variable mesenchymal and epithelial component (Fig. 2), necrosis+++ , heterogeneous components i.e. bone, cartilage, squamous epithelium with keratin also seen (seen as minor tumour component). Vascular invasion was absent. Tumour was present in peritoneal fat and showed metastatic foci in one perirenal lymph node. Cut end of renal vessels was free of tumour. Post operatively patient received chemotherapy using Actinomycin - D, vincristine followed by adjuvant radiotherapy as on lines of pediatric Wilms' tumour protocol.

DISCUSSION:

Wilms' tumour, or nephroblastoma, is a malignant renal tumour that arises from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules. More than 90% of all Wilms' tumour cases occur before age 7, with peak incidence between ages 3 and 4.¹

There is no significant difference between the radiological appearance of Wilms' tumour seen in either children or adults. About 75-80% of the cases have similar clinical and radiological findings.² Plain radiograph, intravenous pyelogram (IVP), angiography, USG, CT, or magnetic resonance imaging (MRI) should be performed to establish the diagnosis. The hypo vascular nature on angiography is typical appearance of this malignancy. MRI better delineates renal capsule and extension of the tumour. It is possible to confuse a Wilms' tumour with renal cell carcinoma because of the similar radiological appearances. Even in adult patients, the possibility of a Wilms' tumour should be taken into consideration in cases of flank pain, large tumour mass, fast tumour growth, and young age.

Adult Wilms' tumour is diagnosed based on the criteria given by Kilton, Mathews, and Cohen³.

These include:

- 1) The tumour under consideration should be a primary renal neoplasm;
- 2) Presence of primitive blastemic spindle or round cell component;
- 3) Formation of abortive or embryonal tubules or glomerular structures;
- 4) No area of tumour diagnostic of renal cell carcinoma;
- 5) Pictorial confirmation of histology and
- 6) Patient's age >15 years.

The differential diagnosis of an adult Wilms' tumour with mainly epithelial differentiation includes metanephric adenoma. A predominant blastemic Wilms' tumour has a strong resemblance to lymphoma, peripheral neuroectodermal tumour and rhabdomyosarcoma; and rarely metastatic small cell tumours from lung, immature teratoma, and primary renal cell sarcoma. Extensive search for any other components is needed as a poorly differentiated renal carcinoma can have large sarcomatous areas resembling blastema.³

Most adults present with local pain and haematuria, in contrast to the palpable boggy mass which is more common in children. In adults, about half of the patients have stage 3 or 4 disease.⁴ The most frequent places of metastasis are lung, liver, bones, skin, bladder, colon, brain and contralateral kidney. Metastasis rates for children and adults are 10% and 29%, respectively.¹ The cytogenetic changes of isochromosome 17q are frequently seen with these types of tumour in adult age group⁵

There is no standard therapy for treating patients with adult Wilms' tumour due to the rarity of this subset and paucity of literature available. The National Wilms' Tumour Study- 3 (NWTS) showed that for low risk patients (favorable histology [FH], Stage I/II), less aggressive therapies were not demonstrably worse than more intensive procedure.¹⁰ Wilms'

tumour in adults has a worse prognosis than in the pediatric population, a phenomenon for which there is no adequate explanation.¹⁶

However, results of treatment for adult patients classified with Stage I/II disease published more recently are promising, with an outlook similar to that for children. An update from the NWTS group about treatment outcomes in adults with favorable histology Wilms' tumour (FHWT) described 45 patients treated in the modern era. The overall survival rate was 82%.¹⁴

The aggressive pattern of Stage III adult Wilms' tumour has been demonstrated by several case reports. Treatment should consist of multimodal therapy with surgery, chemotherapy (dactinomycin plus vincristine plus doxorubicin) for 15 months and tumour-bed irradiation according to the NWTS and the experiences associated with adult Stage III disease.^{6, 8} Prognosis for adult patients with unfavorable histology and Stage IV disease (hematogenous metastases) is poor despite aggressive multimodal therapy.⁸ However, durable complete remissions were achieved in 12 of 34 detailed cases of advanced adult Wilms' tumour reported in the literature as demonstrated by Dawson et al.⁹ The recent SIOP study of 963 patients included 30 patients greater than 16 years of age. A complete remission was achieved in 24 patients; four patients relapsed after complete remission; and three of them reached a second remission after further treatment. Event-free survival was 57%, with an overall survival of 83% (median observation time 4 years).¹⁵ They concluded that adults can be cured in a high percentage by a multimodal treatment according to pediatric protocols.

Adults with Wilms' tumour reported in the National Wilms' Tumour Study (NWTS) from 1968 to 1979 and from 1979 to 1987 showed an improvement in the overall 3-year survival rate from 24 to 67%.^{7, 8}

Furthermore, for patients with recurrent disease, encouraging results have been reported recently: long term remissions have been achieved using high dose chemotherapy, radiotherapy, and allogeneic bone marrow transplantation or combination chemotherapy with cisplatin and etoposide.¹¹⁻¹³

Thus adult Wilms' tumour, may be treated with the similar protocols as those used in children,.

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MEETINGS

(i) Symposium on Ovarian Cancer, AIIMS, New Delhi.

27-28th March, 2009. Registration fees Rs 200=00

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(ii) Biennial National Conference of Indian Society of Medical & Pediatric Oncology and Indian Society of Oncology at Taj Coromandel, Chennai.

13th to 15th February, 2009

Information: www.oncon2009.org

(iii) 3rd Biennial Conference of Indian Society for the study of Lung Cancer (NALCON 2009), Shimla (HP) 25-26th April 2009.

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