

Adult Wilms Tumour: Rare case report with an exceptional survival in metastatic disease

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ABSTRACT

Incidence of wilms tumour(WT) in adults is low, the exact number is unknown owing to insufficient documentation or incorrect diagnosis. Compared to their paediatric counterparts, adult wilms tumour has shown poorer prognosis, but with use of multi-modality treatment there is improvement in response and overall survival.

Here in this case report we discuss a case of adult wilms tumour with characteristic triphasic histology & unusual 3 year survival post operatively with irregular treatment in the face of metastatic disease.

Keywords: Adult Wilms Tumour, Triphasic Histology, Poor Prognosis

Key Messages: In view of metastatic disease and perceived radiosensitivity of WT, role of Whole Abdominal Radiation need to be revisited.

INTRODUCTION

Wilms tumour (WT) is the most common malignant kidney tumour in children and accounts for 6% of all childhood malignancies.^[1,2,3] WT is a very rare tumour in the adult age group. Only 3% off all WT are diagnosed in age over 16. It is said outcome for adults is inferior compared with children,^[4,5] here in this case report we discuss a case of adult wilms-tumour with unusual 3 year survival post operatively with irregular treatment which is rare in Indian literature.

Case History

A 32 year old male, presented with complaints of painful lump in abdomen since 1 month with no history of haematuria and other bowel/bladder complaint. Personal history revealed that patient was tobacco chewer for 10 years.

The general examination of the patient was unrevealing with ECOG grade 2. Abdominal examination revealed a palpable mass in the right lumbar region with associated tenderness

CECT Abdomen study showed heterogeneously enhancing mass lesion of size 17.4x 15.3x17 cm arising from right kidney, superiorly abutting and losing fat planes with segment V and VII of liver. There was breach of Gerotas fascia medially. Thrombosis of infrarenal IVC and common iliac veins were noted. Lower pole calyx was compressed by mass causing calyectasis with multiple calculi within, largest measuring 1.12x1.16 cm.

Based on clinical findings and CECT abdomen report provisional diagnosis of renal cell carcinoma

was made and was operated by right radical nephrectomy.

Post operative histopathology though revealed tumor mass of size 21x19x10 cm showing mixed histological pattern comprising predominantly of blastemal, epithelial and stromal elements. Blastemal cells are packed together and arranged in nodules separated by connective tissue stroma. Epithelial element is primitive and show formation of tubules which is suggestive of WilmsTumor (adult type) with tubular differentiation (picture 1,2,3,4,5).

Patient was planned for adjuvant treatment but he defaulted 1½ years and presented with complaints of pain and distension of abdomen for since 1 month. On examination 4 finger breadth hepatomegaly, mild splenomegaly were present, along with presence of omental caking.

USG abdomen and pelvis showed , hypo echoic lesion of size 4.1x3.2 possibly arising from omental wall with internal vascularity. Hepatomegaly with ill-defined lobulated lesion in parenchyma showing internal vascularity with possible extension into gallbladder with moderate ascites.

Patient was started on Ifosphamide + etoposide-based chemotherapy, he tolerated first cycle chemotherapy & showed subjective response in the form of raised appetite and general wellbeing. With reduced hepatomegaly & decreased omental caking on examination.

Patient defaulted after two cycles of chemotherapy for 1 year, only to present with disease progression in form of massive ascites & gross distention of

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abdomen one year later with poor general condition prompting best supportive care.

DISCUSSION

Wilms tumour arises from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules. The preoperative diagnosis of adult WT is extremely difficult because there are no specific radiographic findings that can distinguish it from the more common adult malignant renal neoplasms.^[6] Histopathologically, there is no difference between adult and childhood WTs. Both characteristically have triphasic histology, with components of blastemal, epithelial, and stromal structures, which was noted in our case.

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, & 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.^[7]

Kilton *et al*^[7] have applied following diagnostic criteria for adult WT-

- Primary renal neoplasm
- Primary blastematos spindle- or round-cell component
- Formation of abortive embryonal, tubule-epithelial or glomeruloid structures
- No areas of tumour diagnostic of renal cell carcinoma
- Pictorial confirmation of histology
- Age > 15 years

In the current case, the classic triphasic pattern was clear which is uncommon, with no areas suggestive of renal cell carcinoma. Thus, the final diagnosis was adult WT.

The prognosis for adult WT is significantly worse than that for children^[8] Due to the rarity of WT in adults, no firm treatment guidelines have been established to date. Adult WT is treated according to recent paediatric protocols & warrants aggressive multimodal therapy because of its poor prognosis. The National Wilms Tumour Study Group (NWTSG) recommends that all adult patients with favourable histology should be treated with stage-appropriate combined therapy, as is done for WT in children.^[9]

Prognosis for adult patients with unfavourable histology and Stage IV disease (haematogenous metastases) is poor despite aggressive multimodal therapy.^[10] A considerable lapse of time after primary surgery until the initiation of treatment leads to poor prognosis.^[11] In our case delay in adjuvant management leading to stage IV disease was seen but unusually patient had survival of 3 years with irregular treatment, which is unheard of in literature till date.

CONCLUSION

This case highlights in presence favourable histology treatment protocols can be tailored in less aggressive manner avoiding delay in adjuvant treatment. In view of metastatic disease and perceived radiosensitivity of WT, role of Whole Abdominal Radiation need to be revisited.

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