

A Rare Case of Adult Wilms' Tumour in a Pregnant Female

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Abstract: Wilms' tumor or Nephroblastoma is the most common primary renal malignancy in the pediatric age group. On the contrary, it comprises of only 0.5% of all adult renal neoplasms. The presentation in adults is not very different from that of a renal cell carcinoma. We hereby report a case of WILMS' Tumor in a 23 years old pregnant female with hematuria.

Keywords: Wilms' tumor, Pregnant, Adult.

INTRODUCTION

Wilms' tumor or nephroblastoma commonly originates from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules. It is a childhood tumor and more than 90% cases occur below 7 years of age. Adult Wilms' tumor is a rare neoplasm.

CASE REPORT

A 23 years old pregnant female presented to the Gynaecological outdoor department with complaints of hematuria. Her vitals were stable and her routine work up was within normal limits. There was no gynecological emergency and she was referred to the Department of Surgery.

Patient had no previous complaints of flank pain and no previous episodes of hematuria. She did not complain of burning sensation during micturition. Routine and microscopic examination of urine was within normal limits apart from the presence of RBCs.

Ultrasonography was not helpful and CT scan could not be performed as the patient was pregnant. MRI of the abdomen was done and a mass was noted in the Right kidney. Partial nephrectomy was done and the tissue was sent to the Department of Pathology. On gross examination, the specimen measured 6x5x2.5 cm. It showed a friable variegated tumor mass measuring 5x5x2.5cm. Ureter and vascular line of resection could not be identified. Tissue was submitted for processing and paraffin blocks were made. The prepared slides were stained with Hematoxylin and Eosin and examined

Microscopic examination revealed renal tissue with a tumor composed of cellular areas of small round blue cells in sheets, mesenchymal elements and abortive tubules and glomeruli. The tumor was well

vascularised. The blastemal component was predominant. Mitotic activity was insignificant. The histopathological features were found to be consistent with Triphasic Wilms' tumor with predominance of blastemal element (more than 65% of tumor mass) and features suggestive of favourable histology (absence of Anaplasia). Nephrogenic rests were absent in sections examined [Fig 1, 2, 3].

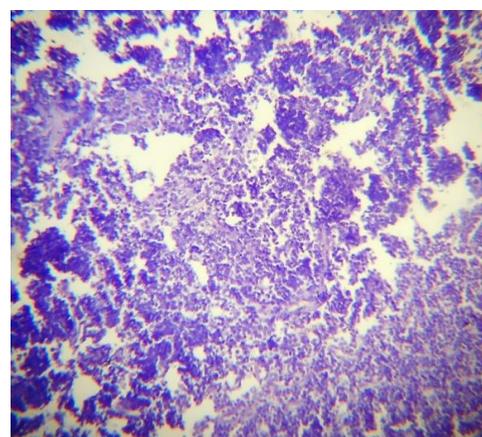


Fig-1: Low power view

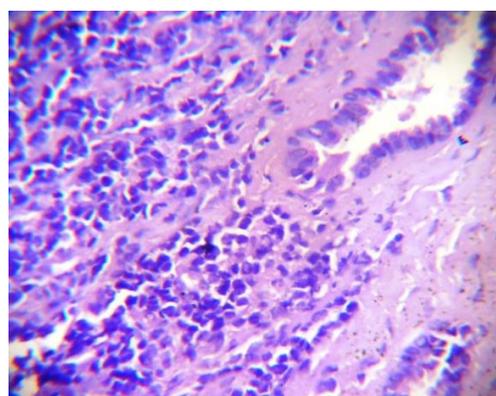


Fig-2: High power view

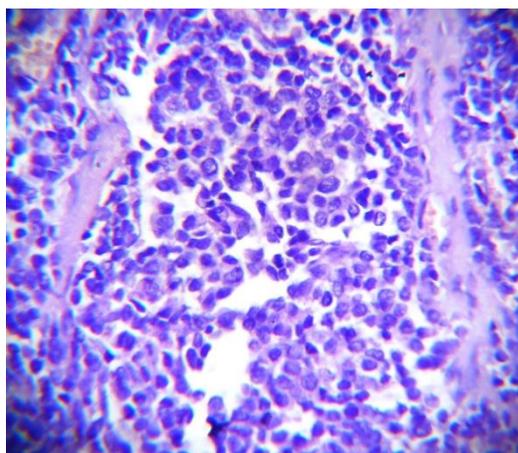


Fig-3: High power view of blastemal component

Fig 1,2,3- Microscopic view(both low and high power) showing histopathological features consistent with Triphasic Wilms' tumor with predominance of blastemal element(more than 65% of tumor mass) and features suggestive of favourable histology (absence of Anaplasia).

On immunohistochemistry, the sections showed WT1 positivity [Fig 4].

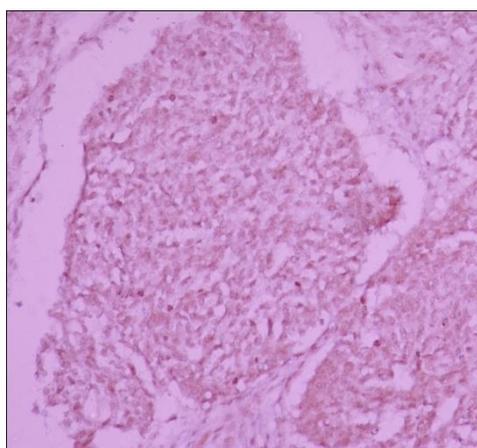


Fig-4: The sections showed WT1 in immunohistochemistry

DISCUSSION

Wilms' tumor comprises of 85% of neoplasms in childhood. Most patients present before 10 years of age. The most common form of presentation is an abdominal lump. They are usually solitary masses but multicentricity and bilaterality have been recorded.

Wilms' tumor is thought to have developed from primitive metanephric blastema [1]. It is said to be composed of three types of cells- blastemal, stromal, and epithelial. The blastemal component may be of diffuse, nodular, serpentine and basaloid patterns. The epithelial component consists of tubular, papillary, glomeruloid, squamous cell, mucinous and neural patterns. The stromal component may show

differentiation towards smooth and skeletal muscles and also towards adipose tissue and cartilage.

Wilms' tumor is known to be associated with a number of syndromes and genetic abnormalities of which mutation in WT1 and WT2 is most common.

Only 0.5% Wilms' tumor occurs in patients more than 15 years of age. By far, mere 200 adult cases have been reported. Adult Wilms' tumor commonly presents with flank pain or abdominal mass and hematuria.

Ultrasonography shows a large complex mass with cystic components unlike renal cell carcinoma which generally shows solid masses [2]. Angiographically, Wilms' shows a hypovascular to moderately vascular mass whereas renal cell carcinomas are generally hypervascular [3]. The disease is difficult to diagnose on radiological basis only. Heterogenous contrast uptake and a pseudocapsule around the tumor in CT are suggestive of Wilms' but are not confirmatory.

Histopathological diagnosis stands confirmatory in the case of adult Wilms' tumour. The diagnosis of Wilms' tumor should not be considered if there is an absence of fetal renal tissue [4]. Nephrogenic blastema is generally located in subcapsular region of renal cortex [5].

Kilton *et al* compiled the following criteria for the diagnosis of adult Wilms' tumor(1)Presence of primary renal neoplasm (2)Presence of a primitive blastemal spindle or round cell component (3) Formation of abortive or embryonal tubular or glomerular structures (4) absence of tumor diagnosis of renal cell carcinoma (5)Age of 15 years [6] Microscopically, adult and pediatric Wilms' shows the same features.

The most common differential is Renal cell carcinoma from which Wilms' tumour can be distinguished by the presence of abortive or embryonic glomerulo-tubular structure with an immature spindle cell stroma [2] which was a definite finding in our case.

The other round cell tumours which are possible in this case are embryonal rhabdomyosarcoma, multicystic nephroma, metanephric adenoma, Ewing sarcoma/peripheral neuroectodermal tumour and neuroblastoma. In the case which we present, Rhabdomyosarcoma was ruled out due to absence of strap cells. Muticystic nephroma is a benign condition showing a lot of cystic spaces without areas of expansile solid areas which was not present in our case. Metanephric adenoma generally has tiny tubular and papillary configuration and bland nuclear features which did not concur with our findings.

To differentiate our case from Ewing sarcoma/peripheral neuroectodermal tumor and neuroblastoma, we performed immunohistochemistry for WT 1 and a diagnosis of Wilms' tumor was arrived at.

Due to the rarity of the case and limited research the treatment protocol is not standardized. Yet, radical surgery, chemotherapy and irradiation of the tumor bed have been agreed upon as necessary steps [7].

The prognosis of adult Wilms' is found to be worse than that of pediatric Wilms' [7]. Hence early diagnosis is important for better outcome.

CONCLUSION

It is always advisable to consider the possibility of Wilms' tumour in adult patients who present with abdominal masses rapidly increasing in size or flank pain. Keeping in mind the rarity and the possibility that this lesion might easily escape a clinician's and a pathologist's mind because of the very improbable age, it is important to consider this diagnosis along with the other most common alternative that is Renal Cell Carcinoma. As the prognosis of Adult Wilms' is worse than pediatric age group, early diagnosis and treatment may bring steady improvements in the outcome of the disease.

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