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Research Article

ADULT WILMS TUMOR: CASE REPORT AND REVIEW OF LITERATURE

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A R T I C L E I N F O

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A B S T R A C T

Wilms' tumor is childhood's most common renal tumor, and its presentation in the adult age is extremely rare. It is difficult to differentiate adult Wilms' tumor from renal cell carcinoma based on radiological findings alone. The diagnosis in adults is often made following nephrectomy for a presumed renal cell carcinoma. We report a case of 28year old female diagnosed as Adult Wilms tumor on the basis of histopathological and immunohistochemical findings. Finally, a literature review is performed to assess diagnosis of adult wilms tumor.

INTRODUCTION

Wilm's tumour (or nephroblastoma), is a malignant renal tumor that arises from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules.[1] It is the most common abdominal tumor in children while in adults, it is extremely rare, representing only 0.5% of all renal neoplasms.[2] In children, the tumour is frequently a painless, rapidly enlarging abdominal mass that is often readily palpable while adults normally present with abdominal pain and hematuria. Distant metastasis is seen to the lungs and liver.[1] The standard pediatric treatment is proposed by two collaborative groups: the National Wilms Tumor Study (NWTS) and the International Society of Paediatric Oncology (SIOP). The treatment includes multimodal management with radical nephrectomy as the cornerstone of management, along with exclusive chemotherapy or with concomitant radiotherapy in most patients.[3] Due to its low frequency, it is rarely suspected in older patients, leading to delayed diagnosis and management, presenting with later stages at the initial diagnosis.[4]

We present a case report of a 28year old female treated in the institute Pt. B.D. Sharma, PGIMS, Rohtak. The patient presented with the complaints of a dull aching pain and history of weight loss for 6 months.

Case Report

A 28 years old female presented with complaints of a dull aching pain in abdomen and weight loss for 6 months without any history of fever /trauma/ hematuria /hypertension.
The general physical examination of the patient was unrevealing with no lymphadenopathy or bony tenderness. Laboratory investigations revealed a haemoglobin level of 10.2g/dL, Blood urea nitrogen (BUN) of 50 mg/dl, serum creatinine of 1.6 mg/dl and eGFR was 49.5 ml/min/1.73m^2. Albuminuria was present and coagulation profile was normal. An ultrasound examination of the abdomen revealed a bulky left renal mass with ill defined corticomedullary differentiation and internal hyperechoic areas with echogenic content. Contrast enhanced computed tomography (CECT) abdomen showed a large hypo to isodense exophytic mass lesion measuring 10.7x9.2x12.8 cm with contrast enhancement, cystic component and enhancing septation, noted in lower and middle pole of left kidney. (Figure1) Left radical nephrectomy was done on provisional clinical diagnosis of? renal cell carcinoma /? Pheochromocytoma and the nephrectomy specimen was sent for histopathological examination.

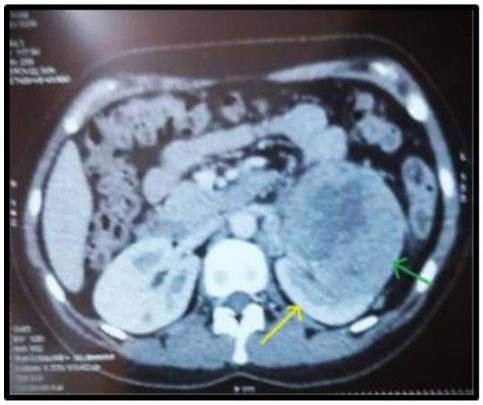

Fig 1 Axial CECT: There is a large heterogenous mass arising from the left kidney. The yellow and green arrows point to normal renal parenchyma and tumor mass respectively.

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Gross examination

Received a left nephrectomy specimen weighing 450gms and measuring 10 X 7X 7cm.The external surface was fibrofatty tissue covered, congested and hemorrhagic at places. The cut surface showed a well circumscribed tumor measuring 8.6x7.5x6.6 cm. The tumor was grey brown to yellow, with occasional foci of haemorrhage and necrosis. (Figure2) The adjacent renal parenchyma appeared unremarkable. The renal capsule was intact. There was no involvement of the renal pelvis or vein.

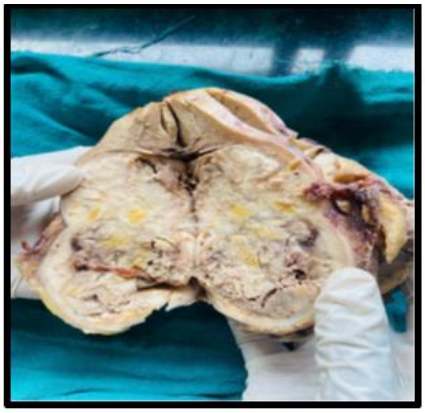

Figure 2 Gross examination of the renal mass revealed a well-delineated white-tan, nodular-appearing mass with hemorrhage replacing the lower pole of the kidney.

Microscopic examination

Histopathological examination showed a cellular tumor comprising of epithelial and blastemal elements. The epithelial component comprised of tubules admixed with blastemal cells. The tubular pattern resembled pseudo-rosettes at places. The blastemal component was composed of sheets of small blue round to oval primitive cells with scanty cytoplasm. Areas of necrosis and frequent mitotic figures were present but no areas of anaplasia were seen. (Figure3)

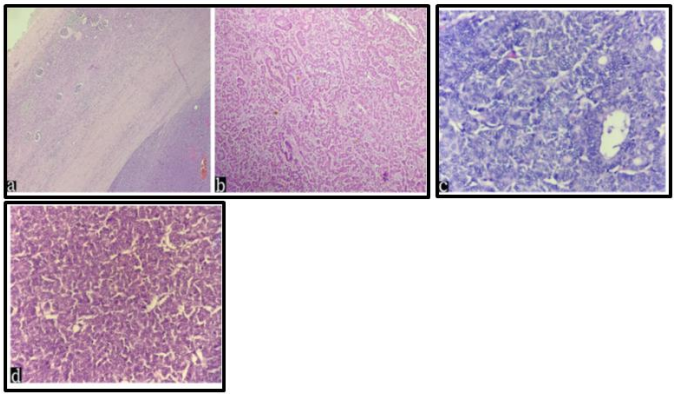

Figure 3 Photomicrographs (from left to right) (a)Wilm's tumor with well defined borders pushing renal parenchyma within (H&E, 40x) (b) epithelial component showing cells having more abundant cytoplasm & forming primitive tubules (H&E, 100x) (c) Tubular pattern showing pseudorosettes (H&E, 400x) (d) Blastemal component showing sheets of small round blue cells (H&E, 400x)

On IHC, tumor cells were positive for WT1, CK (epithelial component), CD56 (blastemal component) and negative for CD10, vimentin, synaptophysin and chromogranin.(Figure4)

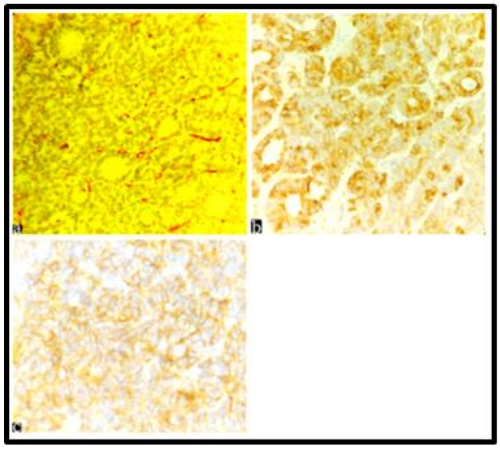

Fig 4 (a) WT1 immunostain demonstrating diffuse nuclear positivity expressed by tumor cells (b) Cytokeratin expressed in the cytoplasm of the epithelial/tubular cells (c) CD56 positivity (membranous staining) in the blastemal cells.

Based on histomorphological features and immunohistochemical profile, a final diagnosis of adult Wilms' tumor was made.

DISCUSSION

Wilms tumor is the most common kidney tumor in children, whereas renal cell carcinoma is most common in adults.[5] An incidence of 10 cases per million persons per year in the pediatric population has been reported, in contrast to less than 0.2 cases per million persons per year in the adult population.[4] Adult Wilms' tumour is diagnosed based on the criteria given by Kilton, Mathews and Cohen. These include:

- The tumour under consideration should be a primary renal neoplasm.
- Presence of primitive blastemic spindle or round cellcomponent.
- Formation of abortive or embryonal tubules or glomerular structures.
- No area of tumour diagnostic of renal cell carcinoma.
- Pictorial confirmation of histology and
- Patient's age >15 years.[1]

Histopathological examination show no difference between adult and pediatric Wilm's tumor.[4] Loss-of-function mutations of a number of tumor suppressor genes, including the WT1 gene located on chromosome 11p13, p53, familial WT1 and 2 (FWT1 and FWT2) genes, and at the 11p15.5 locus, are detected in patients with Wilms tumor.[5] Diagnostic delays and potential biological differences may lead to a more advanced disease at the first diagnosis in the adult population.[4]

Treatment comprise of multimodal therapy with surgery, chemotherapy (dactinomycin plus vincristine plus doxorubicin) for 15 months and tumour- bed irradiation with IMRT technique. Prognosis for adult patients with unfavorable histology and Stage IV disease (hematogenous metastases) is poor despite aggressive multimodal therapy.[1]

Alapont *et al*[2] reported the clinical manifestations, complementary explorations, treatment, and results from 3 males aged 16, 21, and 22 years with Wilms tumor. Computed tomography commonly suggested the diagnosis. They concluded that despite its aggressive treatment, such as radical

surgery, chemo- and radiotherapy, the prognosis in adults is worse than in children.

In a case report by Mahmoud *et al*[5], a 24-year-old woman was incidentally found to have a 2-cm left kidney mass. Fine-needle aspiration performed revealed metanephric adenofibroma. She gave a history of recurrent urinary tract infections. A routine ultrasound during her pregnancy showed a slight progression in the left kidney mass. A follow-up contrast computed tomography scan of the abdomen and pelvis revealed a 6.4 × 4.8 cm left upper pole kidney mass. She underwent left laparoscopic radical nephrectomy and adrenalectomy with para-aortic lymph node resection. Sections examined show tumor comprising of epithelial, blastemal and scant stromal elements. On IHC, tumor cells were positive for WT1, CK7, PAX8 and CD56. On the basis of histopathological and immunohistochemical findings, a diagnosis of Adult Wilms tumor was made.

Nerbi *et al*[3] published a case report of adult Wilms' tumor in a 22-year-old man, which was diagnosed unexpectedly the following nephrectomy for presumed renal cell carcinoma.

In a study performed by Sagar[1] in 2019, a 70year old female presented 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.

Renal cell carcinoma is the most common tumor in the adults, and in some cases, histological differentiation of nephroblastoma and RCC is difficult. As Wilms' tumor is a rare entity in adults, its occurrence is hardly suspected by both the Urologists as well as the Pathologists. The difficulties incorrect diagnosis may lead to inappropriate or delayed treatment and may thus contribute to a poorer prognosis in these
patients.[3]

CONCLUSION

Adult Wilms' tumour is a rare entity with mainly case reports-only literature. Being so rare, the diagnosis is generally made on postoperative pathology. The importance of registering such patients to international data bases will assist in the research and development of future management guidelines for this tumor.

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