Case Report

Adult Wilms tumour: A rare entity

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Abstract

WILMS TUMOR (nephroblastoma) in adults is rare. Only 3% of Wilms tumors are reported in adults. Most adult patients are diagnosed unexpectedly following nephrectomy for presumed renal cell carcinoma. We present a 40 yr old female patient who came with complaints of lump and pain in left lumbar and hypochondrium region. After examination and workup, provisionally diagnosed as renal cell carcinoma of left kidney. Radical nephrectomy was done and histopathology revealed Wilms tumor.

Keywords: Wilms tumor, Radical nephrectomy

1. Introduction

Wilms tumour is the second most common intraabdominal cancer of childhood and the fifth most common paediatric malignancy overall (approximately 6% of all paediatric cancers),¹ named after the 19th century German surgeon Carl Max Wilhelm Wilms.

Only 3% of Wilms tumours are reported in adults.² Most adult patients are diagnosed unexpectedly following nephrectomy for presumed renal cell carcinoma.³ In 5-10% of patients, both kidneys are affected at the same time (synchronous bilateral Wilms tumour) or one after the other (metachronous bilateral Wilms tumour). The overall survival to the tune of 83% has been recently reported with the use of primary nephrectomy followed by adjuvant combination chemotherapy.⁴

Adult Wilms tumor is diagnosed based on the criteria given by Kilton, Mathews, and Cohen.⁵ These include: 1) The tumor 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 tumor diagnostic of renal cell carcinoma; 5) pictorial confirmation of histology and 6) patient's age >15 years.

The most common presentation is an asymptomatic abdominal mass. Other presentations are abdominal pain, hematuria, urinary tract infection, hypertension, gross hematuria and fever. Advanced disease may rarely present with respiratory symptoms due to lung metastases.¹

2. Case report

A 40 YR OLD female came to surgery opd with complaints of pain in left lower abdomen since 10-12 yrs and lump
in left lower abdomen since 8yrs. Pain was diffuse and non radiating. Lump of initial size about 15×10 cms increased in size over 8 yrs to present size associated with pain. There was history of loss of appetite, disturbed sleep, and weight loss. There was no history of hematuria, urinary incontinence, per vaginal discharge, backache, prolong cough, fever, previous surgery and no similar complaints present prior to 12 yrs. Bowel and bladder habit was normal. There was no menstrual disturbance.

On examination, patient was afebrile. Vitals were stable. Pallor was present. There was no icterus, cyanosis, clubbing and lymphadenopathy. Abdomen examination revealed lump of size 20×15 cms in left lumbar region extending to left hypochondriac and not crossing the midline. Skin over swelling was normal. Lump was moving with respiration. Lower margin was well defined. Upper border fused with costal margin. There was no local rise of temperature. Tenderness was absent. It was firm in consistency and not freely mobile. It was bimanually palpable, ballotable with dullness in renal angle. Skin over the swelling was pinch able. Other systemic examination was normal. Provisional diagnosis of renal cell carcinoma of left kidney was made.

Blood investigations along with kidney function test were within normal limit. Ultrasound examination revealed large heterogenous, solid mass lesion with internal vascularity and calcification in the left kidney and is most likely of neoplastic origin.

CT scan revealed large heterogeneously enhancing, well defined soft tissue attenuation lesion with lobulated margins involving the left hypochondrium and lumbar region arising from the left kidney (interpolar and lower polar region) with calcification within showing necrotic area. These imaging finding are in favor of neoplastic etiology of left kidney (Renal cell carcinoma). No renal vein/collecting system invasion is seen with enlarged homogenously pre and pre and para-aortic lymph nodes. (Fig-1)

Patient was planned for radical nephrectomy. Left sub costal incision taken. Intraoperatively there was evidence of huge lump in left hypochondrium and left lumbar region, left renal vein identified ligated divided followed by left renal artery. There was evidence of enlarged, multiple paraaortic lymph nodes. Ligation and division of left ureter done at the junction of proximal 2/3rd and distal 1/3 rd lump remove along with the gerota fascia and proximal 2/3 of ureter. (Fig-2)
Histopathological examination revealed Sections from different areas of the partly encapsulated renal mass show a tumour composed predominantly of blastemal and epithelial elements arranged in sheets, cords and lobules separated by fibrous septa. The blastemal areas are extremely cellular and composed of small round to oval primitive cells with scanty cytoplasm. Occasional mitotic figures are seen. The epithelial component is characterized by formation of numerous tubules with a lumen and lined by a single layer of columnar cells with elongated nuclei. Areas of fibrocollagenized tissue with calcification are seen. Large areas of necrosis are present. Surrounding compressed renal parenchyma is seen externally in some areas. A small fragment of adrenal tissue is also seen. Sections taken from the surgical margin of the ureter show normal histology. Impression is Wilms tumour of left Kidney. (Fig-3)

Postoperative course of the patient was normal and was discharged on 10th post-op day. Patient was advised chemotherapy but lost for follow up.
3. Discussion

In adults, Wilms tumor is larger and ill-defined, with areas of necrosis and hemorrhage. About half of the patients have stage 3 or 4 disease. An update from the NWTS group about treatment outcomes in adults with favorable histology Wilms tumor (FHWT) described 45 patients treated in the modern era and overall survival rate was 82%. Reinhard et al in 2004 concluded that adults can be cured in a high percentage by a multimodal treatment according to pediatric protocols. Literature also suggest adult Wilms tumor has a worse prognosis than in the pediatric population.

National Wilms Tumor Study (NWTS) and other studies have recommended multimodal therapy for the disease with surgery, chemotherapy (actinomycin D, vincristine and doxorubicin) for 15 months and tumor bed irradiation in the case of stage 3 disease. Less aggressive therapy with two drugs is advised in stage 1 and 2 disease.

Our case presented with complaints of lump and pain in left lumbar and hypochondrium region. After examination and workup provisional diagnosis of renal cell carcinoma of left kidney was made. Radical nephrectomy was done and histopathology revealed Wilms tumor. Though Wilms tumor in adult is rare but possibility of it should be considered in a patient presenting with renal lump.

References