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Adult Wilms' Tumor

- A Case Report -

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A case of Wilms' tumor in a 67 year-old female is presented. The tumor totally replaced the left kidney with extension to Gerota's fascia, the adrenal gland and ureter, with tumor thrombi in the left renal vein. Multiple bony metastases to the skull, shoulder, rib, ilium, acetabulum and femur were noted also. Microscopically, the tumor was composed predominantly of blastemal tissue with diffuse anaplasia with areas of tubular differentiation and chondroid elements. Clear cell nests were found in the proximal ureter with gradual maturation downward.

Key Words: Wilms' tumor, bony metastases, clear cell nests

Wilms' tumor is a renal tumor of childhood originating from the renal blastema. But it is rarely found in adults, and there have been more than 200 case reports in the literature (Culp and Hartman 1948; Jagasia and Thurman 1965; Chung *et al.* 1984) and in Korea (Lee *et al.* 1970; Seo *et al.* 1984; Yoon *et al.* 1984; Kim and Lee 1985; Park and Seo 1985).

Wilms' tumor in adults presents a diagnostic problem due to its rare incidence and difficulty in differential diagnosis from renal cell carcinoma or other sarcomatoid tumors (Kilton *et al.* 1980).

We report a case of adult Wilms' tumor showing multiple bony metastases and clear cell nests in the ureter.

CASE HISTORY

A 67 year-old female was admitted to the Department of Urology due to intermittent gross hematuria and urinary frequency for 20 days. Her body weight had decreased 5kg during the previous 6 months. Blood pressure was 120/80 mmHg. The patient looked acutely ill. The skin was warm and dry. The conjunctivae were not pale and the neck was supple. The abdomen was soft and flat except for a huge, tender mass in the left upper abdomen. Bowel sounds were increased but otherwise unremarkable. The hemoglobin was 11.5gm/dl, hematocrit 35.2%, and white blood cell count 5,600/mm^3 with a normal differential. The platelet count was 290×10/ℓ. Urinalysis revealed a specific gravity of 1.005, WBC 3-5/HPF, RBC 2-3/HPF, and neither protein nor glucose were detected. Blood glucose was 100mg/dl, amylase 67U/ℓ, sodium 135mEq/l, potassium 4.2mEq/l, chloride 100mEq/l, and CO_2 content 23mM/l.

A cystoscopy performed before admission revealed a nodular mass above the left ureteral orifice. Intravenous pyelography showed nonvisualization of the left kidney and an irregular filling defect on the superior aspect of the bladder (Fig. 1). An abdominal CT scan revealed that the left kidney had been replaced by a low density mass which grew into the renal pelvis (Fig. 2). There were tumor thrombus in the left renal vein and a 2cm sized, oval shaped lymph node in the left paraaortic area.

Under the impression of renal cell carcinoma stage IV, a left radical nephrectomy was done. The pathological diagnosis was Wilms' tumor. Twenty days after surgery, the patient was discharged with clinical improvement.

Eleven days later the patient was readmitted because of chest pain for 2 days. Physical examina-

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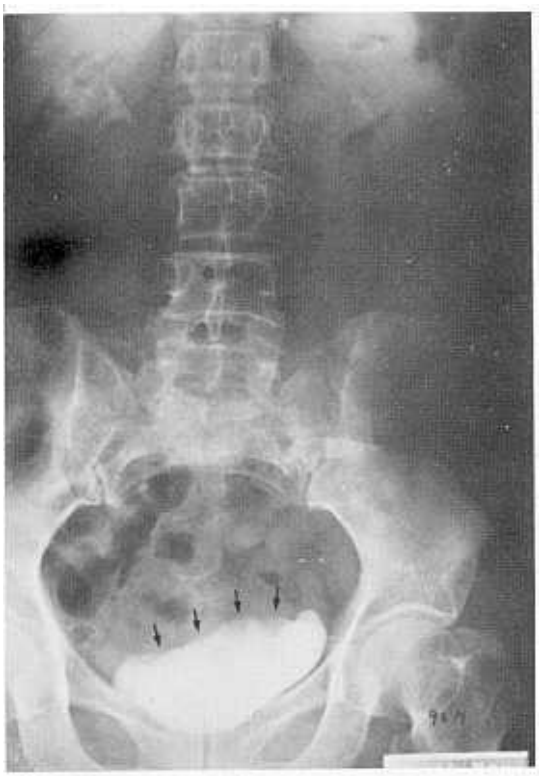

Fig. 1. Intravenous pyelography shows nonvisualization of the left kidney and an irregular filling defect on the superior aspect of the bladder (black arrows).

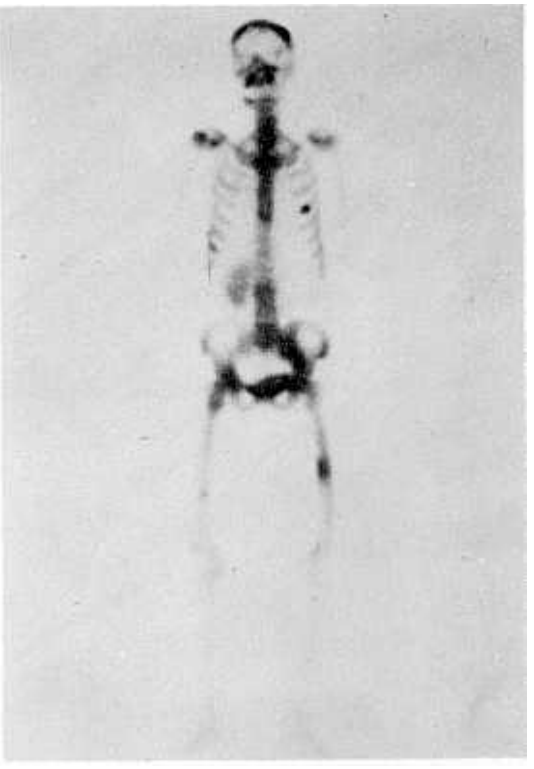

Fig. 3. A whole body bone scan show an increased uptake in the skull, right shoulder, left 5th rib, left iliac bone, right acetabulum and left femur.

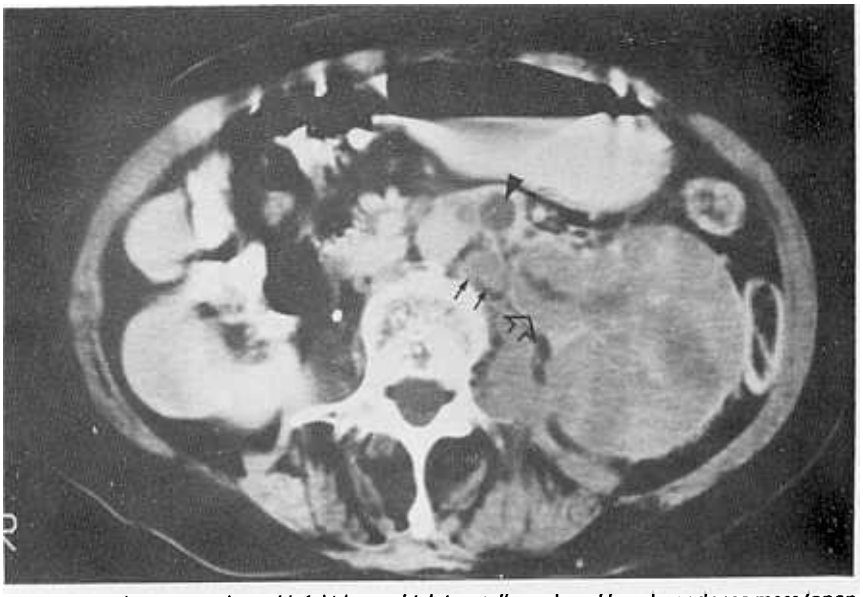

Fig. 2. Abdominal CT scan shows an enlarged left kidney which is totally replaced by a hypodense mass (open arrow). A tumor thrombus (arrowhead) is in the left renal vein and an enlarged lymph node is noted in the left paraaortic area (black arrows.)

tion was unremarkable except for the surgical wound on the left mid abdomen. CBC and urinalysis were within normal limits. A whole body bone scan showed an increased uptake in the skull, right shoulder, left 5th rib, left iliac bone, right acetabulum and left femur (Fig. 3). Radiotherapy was recommended but refused. The patient progressively suffered from respiratory difficulties and a decrease in consciousness, and on the 6th hospital day rales were heard in both lung fields. The patient expired on the 22nd hospital day. Permission for autopsy was denied.

PATHOLOGIC FINDINGS

The radical nephrectomy specimen weighed 1480 gm, and was composed of the left kidney with 7cm of the ureter attached and the adrenal gland. The external surface of the kidney was relatively well preserved and covered by Gerota's fascia, but showed multifocal invasion. It measured 18×17×15cm. On section, the tumor had totally replaced the kidney, and the normally appearing renal parenchyma did not remain (Fig. 4). The pelvic cavity was obliterated by the tumor. A thickening of up to 1.5cm in the proximal portion of the ureter was noted and its lumen was narrowed. The tumor was solid, partially lobulated by fibrous bands, and pinkish gray with extensive necrosis. The renal vein was dilated by tumor emboli. About 20 paraffin blocks were obtained from the kidney and total mapping of the ureter was performed.

Microscopically, the tumor was largely composed of blastemal tissue with diffuse anaplasia, that is, sheets of small round cells with scanty cytoplasm. Primitive tubules and benign chondroid islands were scattered, but skeletal muscle differentiation could not be found by routine hematoxylin-eosin, phosphotungstic acid hematoxylin, and myoglobin PAP staining (Dako Inc) (Fig. 5 & 6). A few tubules and sclerotic glomerull were remained between the tumor, but the dysplastic foci was not found. Electron microscopy was done with paraffin blocks, which showed cell junctions in the epithelia differentiation area (Fig. 8). All of these findings were diagnostic of Wilms' tumor.

Interestingly, clear cell nests were found in the muscle coats and serosa of the distal ureter intermixed with compressed small tumor cell clusters, resulting in a narrowing of the lumen (Fig. 7). The clear cells had oval to round, relatively uniform nuclei with occasional nuclear enlargement and hyperchromatism and were surrounded by reticulin fibers, mimicking transitional cells or clear cell sarcoma. The ureteral mucosa was almost detached, but evidence of in situ transitional cell carcinoma was not found. Epithelial membrane antigen PAP staining (Dako Inc) showed strong positivity in the clear nests and weak positivi-

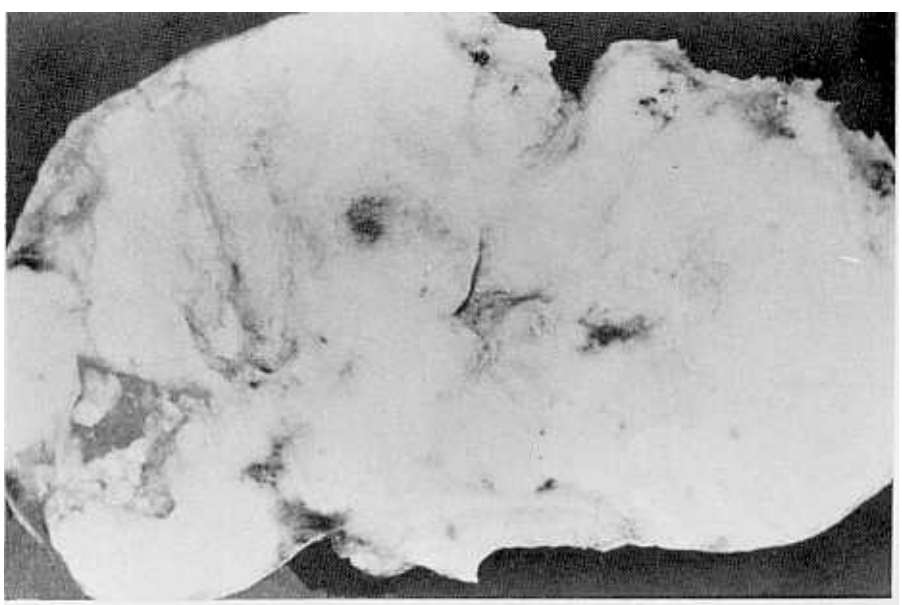

Fig. 4. Cut section of the left kidney reveals a gray white tumor mass which replaces the renal parenchyma entirely.

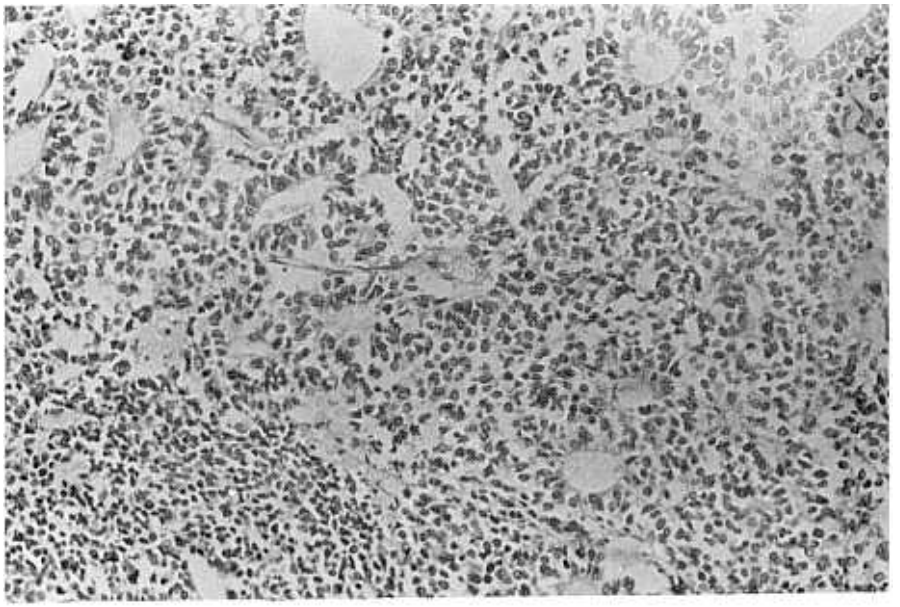

Fig. 5. The tumor cells show distinct tubular differentiation (hematoxylin & eosin, ×150).

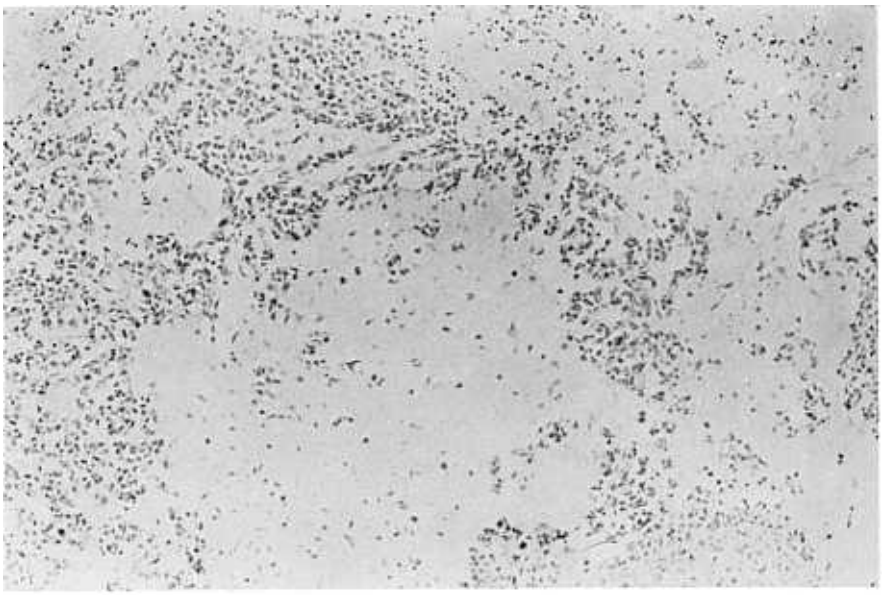

Fig. 6. An area of cartilagenous differentiation is present (hematoxylin & eosin, ×100).

ty in the primitive tubules in the kidney. Periodic acid-Schiff, oil red 0, toluidine blue stains and myoglobin were all negative in the clear cell nests of this case.

Mapping of the ureter showed a tendency to gradual nuclear maturation and clearing of the cytoplasm from the proximal to distal ureter.

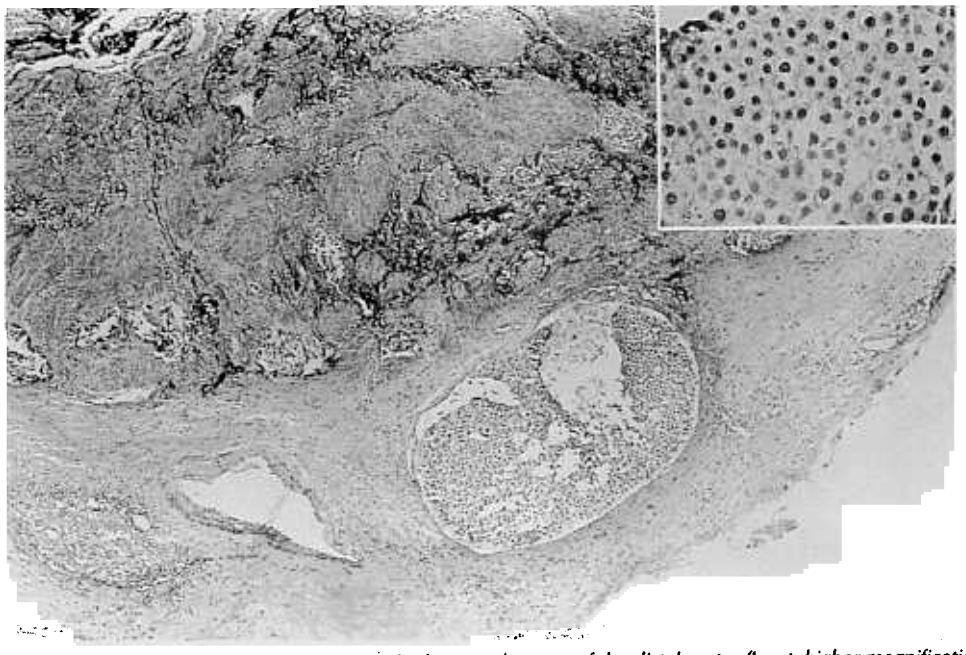

Fig. 7. Clear cell nests are noted mainly in the muscular layer and serosa of the distal ureter (Inset: higher magnification, ×250) (hematoxylin & eosin, ×40).

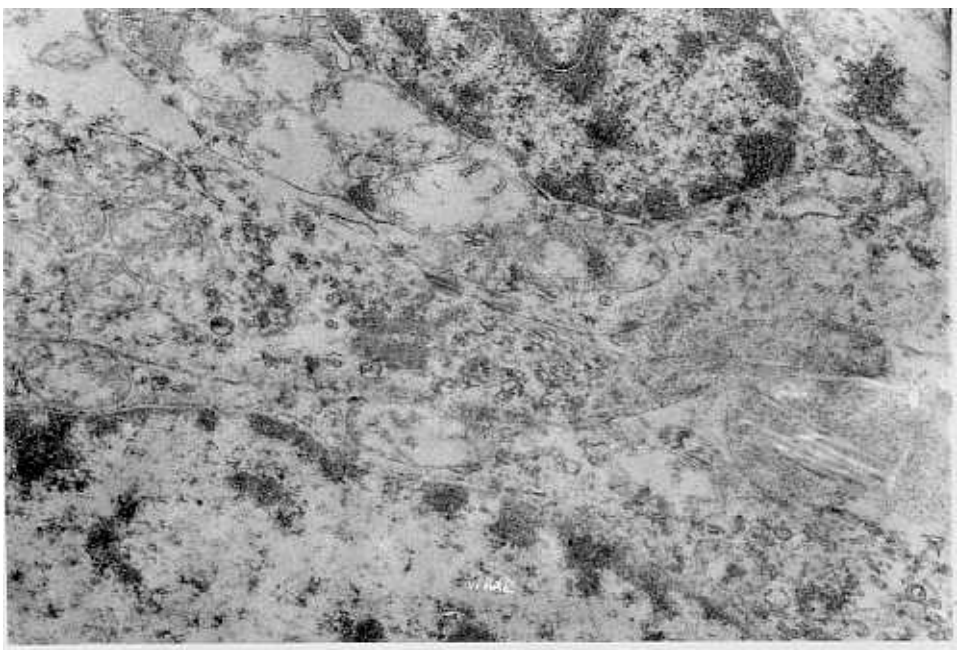

Fig. 8. Electron micrograph of the tumor cells shows many rough endoplasmic reticulum, mitochondria and cell junctions (uranyl acetate & lead citrate, ×8,000).

DISCUSSION

Since 56 cases of adult Wilms' tumor were reported by Esersky *et al.* in 1947, over 200 cases have been reported in the literature (Culp and Hartman 1948; Chung *et al.* 1984; Yoon *et al.* 1984; Kim and Lee 1985; Park and Seo 1985)

However there is some argument that not all of the reported cases are truly Wilms' tumor, especially in the monophasic spindle or sarcomatous forms (Oslen and Bischoff, 1970). Kilton *et al.* (1980) listed the following as criteria for adult Wilms' tumor: primary renal tumor, primitive blastematous spindle or round cell components, formation of an abortive or embryonal tubular or glomeruloid structure, no area of hypernephroma, and patient's age over 15 years.

Our presented case fulfilled the criteria of adult Wilms' tumor, but there are some peculiar aspects. Firstly, the patient presented multiple bony metastases, which are infrequent in the usual Wilms' tumor. Secondly, the tumor directly invaded the ureter throughout its course to the ureteral orifice, without significant disfiguration of the renal contour, which is more likely to occur in transitional cell carcinoma. Thirdly, besides the classical light microscopic features of Wilms' tumor, the clear cell nests were found only in the ureter with gradual maturation.

Clear cell sarcoma of the kidney is characterized by clear, relatively bland looking cells containing a prominent vascular network (Bechwith and Palmer, 1978). Clinically, it presents a high frequency of bone metastasis and poor prognosis (Marsden *et al.* 1978). However, this tumor is known to develop exclusively in children and rarely to have foci of tubular differentiation, which is not in accordance with our case. In addition, the morphologic features in our case were not typical of clear cell sarcoma.

Transitional cell carcinoma of the renal pelvis may be so undifferentiated and aggressive that it can infiltrate the renal parenchyma and ureter. However, there was no in situ transitional cell carcinoma and the pattern of invasion was not of mucosal spread, but through muscle coats.

Then, the question is, what is the nature of the clear cells? We do not exactly know. We suspect that these cells are differentiating cells from the renal blastema and may be of transitional cell differentiation, with regard to gradual maturation in the ureter, normally the transitional cell harboring area.

The prognosis of Wilms' tumor is well known in children. Beckwith and Palmer (1978) and Sandstedt *et al.* (1987) described a tumor with cellular anaplasia, clear cell sarcoma and malignant rhabdoid tumor with an unfavorable prognosis. Lawler *et al.* (1975), Chatten (1976), and Kheir *et al.* (1978) stated that epithelial differentiation offered a better prognosis. In adults, treatment is not well established and the prognosis is uncertain. Generally it is treated in the same manner as in children, and multimodality treatment is given including surgery, chemotherapy and irradiation (Babaian *et al.* 1980). with regard to the prognosis, there is some controversy probably according to case selection and treatment modality (Altug *et al.* 1964; Kilton *et al.* 1980). However, our patient had a grave prognosis and died of tumor metastasis.

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