

CT features of adult Wilms' tumor : a case report

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Wilms' tumor is the most common malignant renal neoplasm of childhood, but rarely occurs in adults. We report a case of adult Wilms' tumor in a 69-year-old man who showed signs of a palpable mass and weight loss. A CT scan showed a large, well-circumscribed mass involving the left kidney. The mass was predominantly cystic with foci of solid nodules. The cystic portion represented a large area of necrosis on the specimen. Histologically, the tumor was composed of primitive blastematos spindle cells with formation of abortive glomerular and tubular structures, compatible with Wilms' tumor.

Key words : *Wilms' tumor, Nephroblastoma, CT.*

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Wilms' tumor เป็นมะเร็งปฐมภูมิของไตที่พบบ่อยที่สุดในวัยเด็ก แต่พบได้น้อยมากในผู้ใหญ่ คณะผู้วิจัยรายงาน *Wilms' tumor* ในผู้ป่วยอายุ 69 ปี ซึ่งมาพบแพทย์ด้วยอาการมีก้อนในท้องร่วมกับ น้ำหนักลด จากการตรวจด้วยเอกซเรย์คอมพิวเตอร์พบก้อนขนาดใหญ่ที่ไตข้างซ้าย โดยก้อนมีลักษณะ ขอบเขตชัดเจน ส่วนใหญ่ของก้อนเป็นถุงน้ำและมีบางส่วนเป็นก้อนเนื้อ ลักษณะพยาธิทางกายวิภาค พบว่าส่วนที่เห็นเป็นถุงน้ำนั้น เป็นส่วนมะเร็งที่เกิด *necrosis* ลักษณะพยาธิวิทยาพบว่าก้อนนั้นประกอบด้วย *primitive blastematus spindle cells* ร่วมกับ *abortive glomerular* และ *tubular structures* ซึ่งเข้าได้กับ *Wilms' tumor*

Wilms' tumor, or nephroblastoma, is the most common malignant renal neoplasm of childhood. In adults, the occurrence of this neoplasm is rare, occurring in about 0.5% of all renal neoplasms.⁽¹⁾ The diagnostic criteria for Wilms' tumor in adults, based upon Kilton et al. definition,⁽²⁾ are as following: 1) age over 15 years, 2) presence of primary renal neoplasm, 3) histologic evidence of fetal renal tissue, i.e., the presence of abortive or embryonic glomerulotubular structures within an immature spindle cell stroma, 4) absence of histologic evidence of renal cell carcinoma.

An adult Wilms' tumor, diagnosed according to these criteria is reported.

Case Report

A 69-year-old man was presented with a nontender palpable mass and signs of weight loss for 4 months. He did not notice hematuria or other urinary tract symptoms. The physical examination

revealed a huge, firm abdominal mass within the left upper abdomen. The urinalysis was normal. The intravenous urography showed a large, intrarenal mass causing distortion of the lower pole calyces of the left kidney. An ultrasonogram revealed mixed echogenicity of this mass. A CT scan of the abdomen showed a large well-circumscribed mass, predominantly cystic, mixed with foci of solid components. Following intravenous injection of the contrast medium, there was minimal irregular peripheral enhancement. Pseudocapsule, the compressed normal renal parenchyma, was apparent (Fig 1,2). There was no evidence of tumor thrombus in the renal vein nor IVC. Preoperative diagnosis was left renal tumor, most likely a cystic form of renal cell carcinoma. Left nephrectomy was performed.

A 22 x 16 x 12 cm., mixed solid and cystic mass was noted involving the left renal parenchyma. The tumor tissue was dark red with diffuse intratumoral

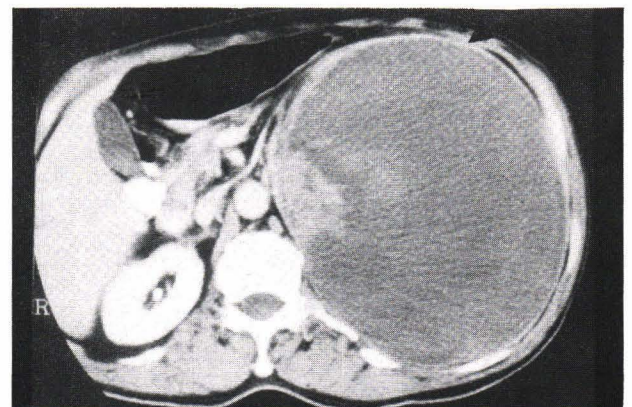
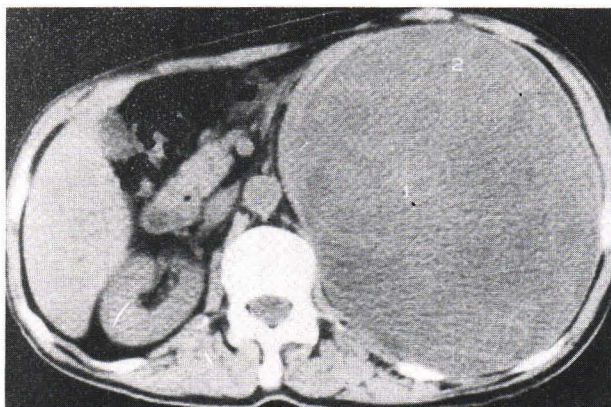


Figure 1. A & B : CT scan, pre and post intravenous contrast study, showing a large well-circumscribed mass predominantly cystic, mixed with foci of solid component, which is of minimal irregular enhancement. Pseudocapsule, composed of compressed normal renal tissue, is also enhanced. (arrow)

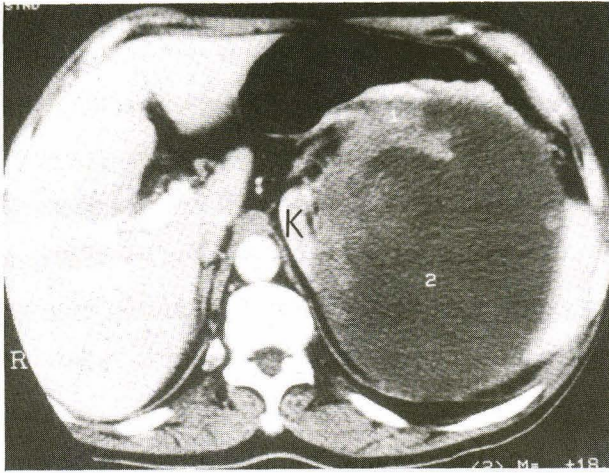


Figure 2. CT scan, post intravenous contrast study, showing normal renal parenchyma (k) at the periphery of the mass, giving the beaking, "claw-sign" appearance; indicating that the mass is of intrarenal in origin.

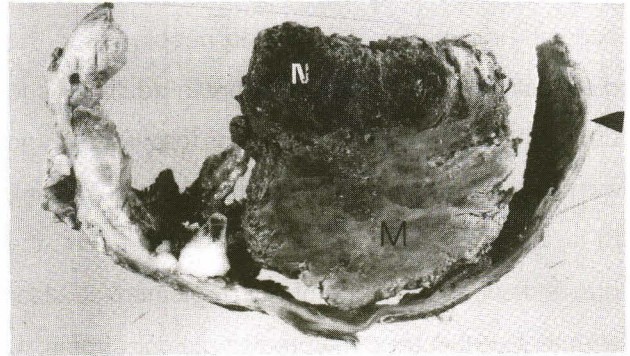


Figure 3. Gross pathology showing well-circumscribed mixed solid and cystic mass (M) with area of hemorrhage and necrosis (N). Note a pseudo-capsule surrounding the mass (arrow).

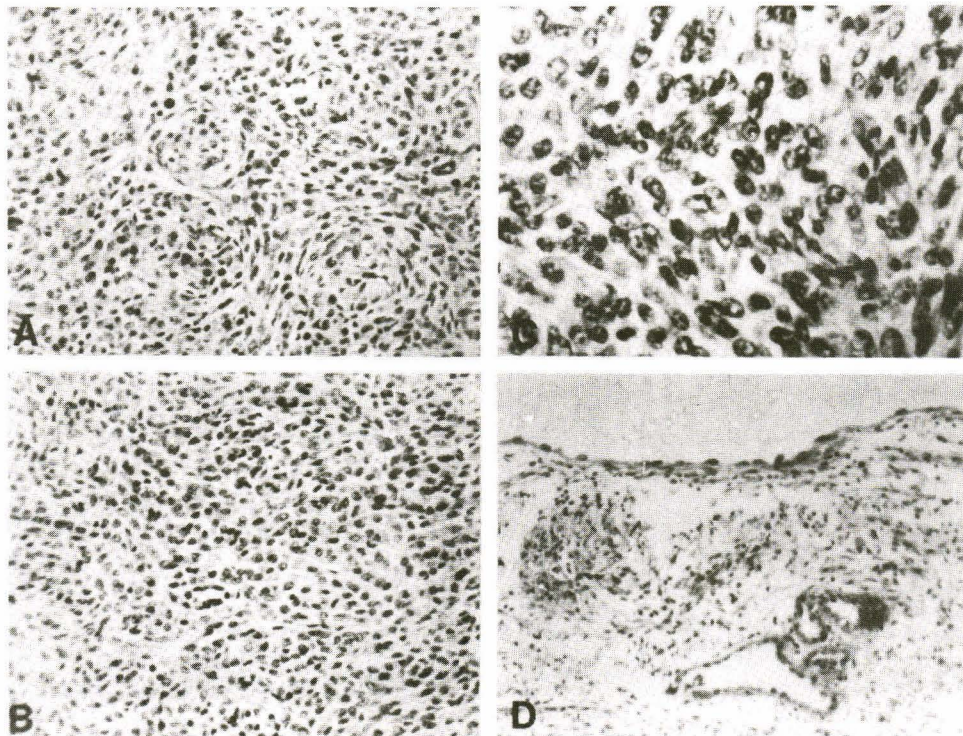


Figure 4. (a,b,c,d) : Microscopic features of the tumor show
 (a,b) pleomorphic primitive spindle cells with abortive glomerular and tubular structures (H & E x 200)
 (c) mitotic figures (H & E x 400) (d) cuboidal cell lining of the cystic duct (H & E x 100)

hemorrhage. The periphery of the mass was gray-white and rubbery in texture (Fig.3). Microscopically, the tumor was composed of primitive blastematos spindle cells with frequent mitosis. Occasionally, the tumor cells were arranged in abortive glomerular and tubular structures. (Fig. 4)

Based upon the historic findings, adult Wilms' tumor was diagnosed. Bone scans and chest radiographs for metastases showed negative. The patient was discharged and a program of combined irradiation and chemotherapy treatment was arranged.

Discussion

Wilms' tumor, a primary renal neoplasm containing primitive blastema and embryonic glomerulotubular structure, occurs most commonly in young children.⁽³⁾ About 240 cases of adult Wilms' tumor have been reported in world literature.⁽⁴⁾ However, the actuality of the incidence in adults is difficult to determine because of some histologic confusion with rhabdoid or clear cell sarcoma.^(4,6) Kilton et al. provided a clear definition of adult Wilms' tumor and emphasized the existence of fetal renal tissue.⁽²⁾ The adult Wilms' tumor, reported here, fulfills the criteria set by Kilton.

The mean age of adult Wilms' tumor is 30 - 40 years.^(2,6) However cases within older age groups are not uncommon, as highlighted by our 69 - year-old patient. The oldest reported case was 84 years.⁽⁷⁾ Similar to renal cell carcinoma, patients with adult Wilms' tumors usually show signs of abnormal mass, abdominal pain or hematuria.^(2,6) It should be noted that a case of adult Wilms' tumor is usually large and palpable in contrast to a small size or renal cell carcinoma at the time of presentation.⁽⁶⁾

Pathologically, adult Wilms' tumor is usually large with expansile growth patterns resulting in a well-defined margin, often with a prominent pseudocapsule composed of compressed normal renal tissue.^(6,8) Focal hemorrhage and necrosis are common. Occasionally cystic changes within the tumor are quite striking.^(1,9) Our presented case also shows this typical pathological pattern. Less commonly, the tumor may have infiltrative growth patterns with an ill-defined and poor margin.⁽⁹⁾ Microscopically, there is no difference between adult and childhood Wilms' tumor. The important histologic evidence is the presence of fetal renal tissue which is shown by abortive or embryonal glomerulotubular structures within an immature spindle cells stroma, as also presented in our case.

An imaging finding method, particularly a CT scan, provides good imaging modality to show the pathological features of adult Wilms' tumors. In an attempt to correlate with the pathological findings, two CT features of adult Wilms' tumor have been described, the large, expansile mass and the infiltrative mass.⁽⁹⁾ The first pattern is more common and the mass is usually predominantly cystic with some foci of solid components.⁽¹⁰⁾ The solid part shows variable enhancement after intravenous injection of the contrast medium.^(6,9) Pseudocapsule, the normal renal parenchyma which is compressed by the tumor, will enhance, giving a so-called crescent or rim sign.^(6,9) This common CT feature concurs with CT findings in our patient. The diffuse infiltrative pattern is less common. CT will demonstrate an ill-defined mass which completely replaces the normal kidney. This CT feature is similar to that found in renal lymphoma.⁽⁹⁾ Calcification, resulting from hemorrhage or necrosis in the lesion, is rare in adult Wilms' tumor, although

they are more likely to be seen than when they occur at a more typical age.⁽¹¹⁾ The calcification is almost always subtle, never extensive or dense.⁽¹²⁾

Similar to renal cell carcinoma, adult Wilms' tumor may invade into the renal vein and vena cava. Therefore, these structures must be evaluated carefully. In our case, CT clearly shows the absence of the angioinvasion by the tumor. Distant metastasis seems to be more common in adult Wilms' tumor than in children.⁽¹³⁾ Lungs are the most common site of involvement.⁽⁵⁾ Other reported metastatic sites include; skin, liver, bone, bladder, sigmoid colon, orbit, brain, spinal cord and contralateral kidney.⁽¹⁴⁾ Therefore the preoperative metastatic evaluation is recommended.^(5,14)

Staging of adult Wilms' tumor is performed according to the National Wilms' tumor study. Compared to childhood stage for stage, adult Wilms' tumor shows poorer prognosis, therefore aggressive multimodal treatment including radical nephrectomy, postoperative irradiation and chemotherapy are recommended.^(2,5) However, because of the rarity of this tumor at the present time the protocol for treatment of adult Wilms' tumor remains conflicting.⁽¹³⁻¹⁵⁾

In conclusion, although rare, adult Wilms' tumor should be included in the differential diagnosis in an adult patient whose CT scan reveals a large, well-circumscribed renal mass showing predominantly cystic with small foci of solid components. Other differential considerations are renal cell carcinoma and multilocular cystic nephroma.^(6,10,11)

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