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Adult Wilms tumor: Case report

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ABSTRACT

Wilms tumor (WT) occurs infrequently in adults. Even rarer is adult WT with extension by direct intravascular spread into the right side of the heart. The present report describes a WT with intracaval and intracardiac extension in a 38-year-young man. In addition, thrombus extension above the infrahepatic IVC represents a major technical topic for surgeons because of the possible occurrence of uncontrollable hemorrhages and tumor fragmentation. We report the results of a surgical approach to caval thrombosis including the isolation of the IVC from the liver as routinely performed during liver harvesting. The morphologic and immune-histochemical findings confirmed the diagnosis.

1. Background

Wilms tumors (WT) are the most common cancers in children that start in the kidneys. Conversely, it is very rare in adults, with an incidence of about 0.2 per million per year in the USA and Europe. There are only a few reports of always small series of patients treated with different protocols. Until recently, a standardized treatment for adults with Wilms' tumor is missing, and exact data on prognosis and late effects are not available. Outcome for adults is inferior compared with children, although better results are reported when treated within pediatric trials.

In this report, a case of WT in adult patient is discussed.

2. Case report

A Caucasian 38-years young man presented to our Department with abominal pain, macroscopic hematuria, edema lower limbs and anemia (8.5 mg/dL) showed by biochemestry investigations. An abdominal ultrasound revealed a hypo-echoic area measuring 10 cm in the right kidney with extension into the right renal vein and inferior vena cava. Afterwards, a Computerized Tomography (CT) scan total body was performed, confirming a mass in the right kidney with extension into the right renal vein and inferior vena cava, and further extension into the right atrium.

In addition, the presence of several nodules was highlighted into both lung parenchyma. A cardio ultrasound confirmed the extension into the right atrium, showing the absence of involvement of the tricuspide valve. Retrograde urography confirmed that the right renal pelvis was compressed and distorted, but was unobstructed. The right ureter also was unobstructed. Renal function tests were normal. Urine analysis revealed no microscopic hematuria.

Based on the clinical findings and investigations, a preliminary diagnosis of renal carcinoma of the right kidney was made.

In May 2013,the following surgical procedure has been used:

Sub-costal bilateral incision.

- Right colonic flexure mobilization.
- Kocher maneuver with isolation of the infrahepatic IVC and of the renal vein confluence.
- Isolation of the hepatic pedicle's elements that are surrounded by a surgical tape.
- Section of the falciform, triangular and hepato-gastric ligaments.
- Detachment of diaphragmatic peritoneum from the liver as far as the identification of suprahepatic veins and suprahepatic IVC that is surrounded by a surgical tape
- Dissection of the IVC from the posterior margin of the liver through a careful section of all the accessory hepatic veins.
- Complete overturning toward up and left side of the liver that is now hanged up to the three main suprahepatic veins.
- Isolation of the proximal tract of the IVC up to the iliac confluence.
- Radical inter-aortocaval (left side tumor) or caval and retro-caval (right side tumor) lymphadenectomy and section of the renal artery.

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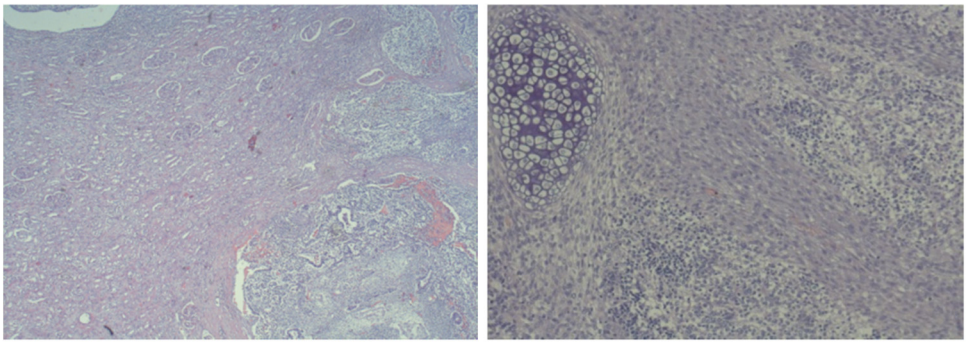

Fig. 1. (a) Triphasic cellular pattern because of the presence of undifferentiated blastemal cells (H/E, 4×). (b) Triphasic pattern: a neoplastic admixture of epithelial (neuroepithelial rosette like structures), blastemal and stromal (immature muscular and cartlageneous) components (H/E, 10×).

- Suprahepatic IVC clamping.
- Clamping of the controlateral renal vein, of the proximal IVC and of the hepatic pedicle
- Large cavotomy and renal vein venotomy with enbloc removal of the thrombus, the kidney and of a buttonhole of caval wall close to the renal vein confluence.
- Suture of the IVC and declamping of the vessels.
- Retro-caval lymphadenectomy.

The resected right kidney measured 21 cm × 13 cm × 9 cm (including perirenal adipose tissue), with ureter of 10 cm of length. Near the renal pelvis there was an unicentric rounded mass measuring 12 cm × 4 cm × 2.5 cm that showed, on cut surface, a gray/tan appearance with soft consistency areas, due to the presence of necrosis and hemorrhage. The organ capsule appeared grossly infiltrated by the tumor and there was an extension into the renal vein, with a neoplastic thrombus that measured 22 cm × 2.1 cm × 2.3 cm.

Histologically, the lesion showed diffuse necrotic-hemorrhagic and cystic changes, with a triphasic cellular pattern because of the presence of undifferentiated blastemal cells and cells differentiating toward epithelial and stromal lineages (Fig. 1a and b). The blastemal cells were small, closely packed, and mitotically active rounded or oval cells with scant cytoplasm, and overlapping nuclei containing evenly distributed, slightly coarse chromatin, and small nucleoli.

The epithelial line was made of rosette-like structures and tubular structures. The stromal component was made of spindle cells with elongated nuclei and storiform pattern of growth (immature muscular tissue) and of immature adipose and cartilaginous tissues.

The neoplastic immunophenotype was also triphasic:

- EMA +, Cytokeratin MNF116 clone + (Fig. 2) for the epithelium component;
- Vimentin +, Desmin + for the stromal component (Vimentin focally positive in blastemal cells);
- NSE +, N-CAM +, s100 ± (neuroepithelium and blastema).

The neoplasm infiltrated the renal capsule and perirenal adipose tissue. The renal vein showed an ectatic lumen with thrombotic organized material and several neoplastic foci within.

The adrenal gland was free of neoplastic invasion.

Three of seven intercavoaortic lymph nodes showed metastatic neoplastic repetition.

Thus, the final diagnosis was Wilms Tumor in the adult, teratoide type (*sec. Rosai & Ackerman's, Surgical Pathology, 2011*), Stade III sec. SIOP (*Med Pediatr Oncol 2002;38:79–82*) (Fig. 3).

A FISH analysis was made for evaluate the 12p chromosome status, with ratio 12p/centromere12 = 1. The signals disposition and the ratio value were not compatible with the presence of short arm 12 isochromosome.

The FISH analysis results, combined with WT1 immunoreactivity, came out in favor of Nephroblastoma without anaplasia.

The patient was discharged after 8 days with a good kidney function (creatinine 1 mg/dL, urea 5 mmol/L, potassium 4.1 mg/dL, hemoglobin 11.7 g/dL) and no complications were observed.

Actually, the patient has been received six courses of adjuvant chemotherapy postoperatively, including one courses with Dactinomycin, Adriamycin, and Mesna, two courses with Paclitaxel and Carboplatin, two courses with Paclitaxel, Ifosfamide, and Mesna, and one course with Vinorelbine and Dactinomycin. No side effects were observed.

At the follow up of 6 months, the patient is alive and progression free disease. CT scan total body and US Doppler imaging showed the presence of an adequate iliac and caval flow.

3. Discussions

The preoperative diagnosis of adult WT is extremely difficult because there are no specific radiographic findings that can

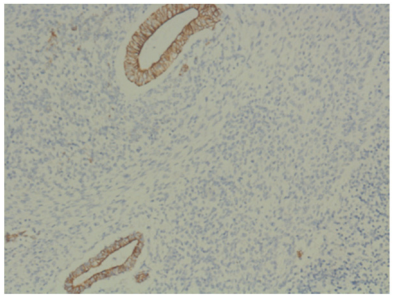

Fig. 2. Pattern of Cytokeratin MNF116 (10×).

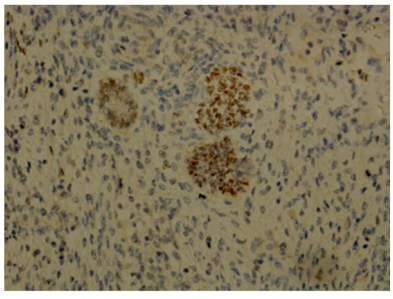

Fig. 3. Wilmor tumors (10×).