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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-old 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.

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## 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-year-old man presented to our Department with abdominal pain, macroscopic hematuria, edema lower limbs and anemia (8.5 mg/dL) showed by biochemistry 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 tricuspid 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.

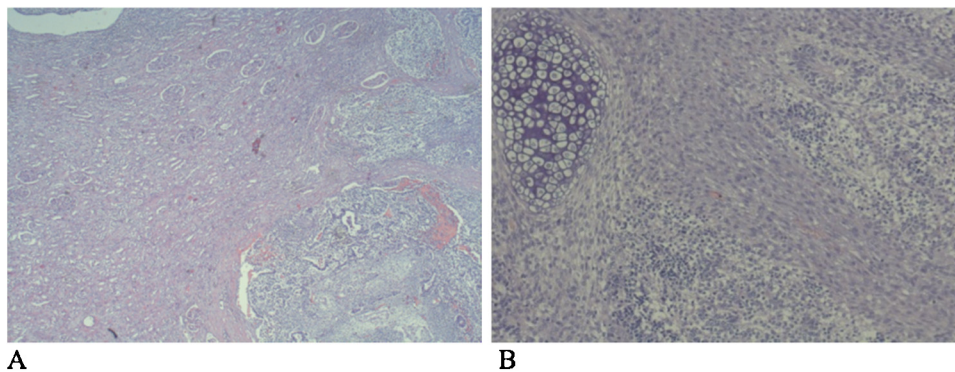
Based on the clinical findings and investigations, a preliminary diagnosis of renal carcinoma of the right kidney was made.

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 cartilageous) 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.

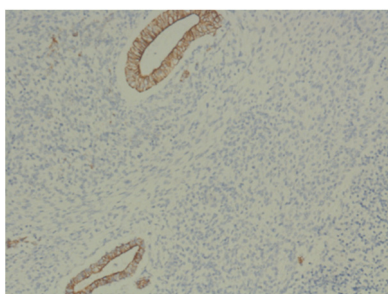
The resected right kidney measured 21 cm × 13 cm × 9 cm (including perirenal adipose tissue), with ureter of 10 cm of length. Near the renal pelvis there was an unicentric rounded mass measuring 12 cm × 4 cm × 2.5 cm that showed, on cut surface, a gray/tan appearance with soft consistency areas, due 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);



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

- 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 intercavaortic lymph nodes showed metastatic neoplastic repetition.

Thus, the final diagnosis was Wilms Tumor in the adult, teratoid 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.

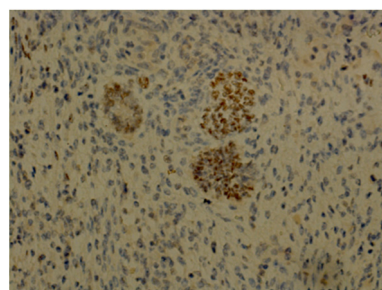
The patient was discharged after 8 days with a good kidney function (creatinine 1 mg/dL, urea 5 mmol/L, potassium 4.1 mg/dL, hemoglobin 11.7 g/dL) and no complications were observed.

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. 3.** Wilms tumors (10×).

distinguish it from the more common adult malignant renal neoplasms [1].

In the current case, the classic triphasic pattern was clear, with no areas suggestive of renal cell carcinoma. Thus, the final diagnosis was adult WT.

Caval thrombosis is a relatively common event occurring in about 10% of patients affected by renal tumor [2]. Echocardiography and color flow Doppler ultrasound are helpful in detecting inferior vena caval and right atrial tumor if the WT has spread through the inferior vena cava into the right atrium. The present case was diagnosed as inferior vena cava and right atrium involvement by the CT examination and echocardiography although tumor type could not define before surgery. US color Doppler was performed to study the iliac-caval axis.

Extension does not take place by vascular invasion but via tumor progression inside the lumen of the renal vein and subsequently of the IVC. Caval thrombosis is not considered a contraindication to radical surgical treatment; on the contrary surgical removal of the tumor and the thrombus, improves the prognosis [3], even in the presence of distant metastases.

Five-year survival rates of patients with renal tumor and caval thrombosis undergoing radical surgery range from 25 to 64% [4]. The validity of this treatment in presence of disease extension to perinephric tissues and regional nodes remains unclear.

The present case reports the results of a fully abdominal surgical technique used for the treatment of patients affected by Level III caval thrombosis. Retro-hepatic IVC is a complex anatomical region. As a consequence, the surgical removal of a thrombus extended to this tract is a high-risk procedure requiring an effective complete surgical control of the vessel. In Ljungberg's [5] experience, the surgical strategy for these cases depends on the length of the thrombus and whether or not the thrombus has infiltrated the wall of the vena cava.

If the thrombus can be easily removed, complete resection is the treatment of choice, however, in cases of atrial thrombus, and more particularly if the thrombus involves the intima, these authors suggest that the thrombus not be touched, and the tumor should be treated by preoperative and postoperative chemotherapy.

Our approach is directly imported from liver transplant surgery. It includes the complete overturning of the liver and the isolation of the entire tract of the infra-, retro- and suprahepatic IVC as commonly used for the removal of both donor and recipient liver. Following that, minimal blood loss (500 mL were calculated in this case) and the complete opening of the IVC allow to reduce the risk of thrombus fragmentation during the removal.

In a series of 26 patients affected by retro-hepatic caval thrombosis undergoing thoraco-abdominal access, Nesbitt et al. [6] reported a significantly lower rate of mortality (overall 2.7%) and the reduction of intraoperative blood loss. Surgical strategy reported by Nesbitt included in all cases a careful mobilization of the liver and a large cavotomy as described in the present study.

As reported [7], in our experience on 15 patients with renal cancer and caval thrombosis, overall perioperative mortality was absent and 14 patients (93.3%) were alive at a mean 53.9 months of follow up.

In this report, the patient is lived and progression free disease at 6 months of follow up, showing a good kidney function and an adequate iliac and caval flows.

The prognosis of the disease was reported to be dismal, with an event-free survival of 20–30% in the 1980s. The first series showing an improved survival was presented by Arrigo et al. for the National Wilms' Tumor Study in 1990 [8].

They reported on 27 adult patients in whom an event-free survival of 67% was achieved. Relapses occurring later than 2 years after diagnosis were not reported in this study. This is in concordance with the pediatric population.

The combined use of thoraco-abdominal access and cardio-circulatory arrest for the treatment of retro-hepatic and thoracic lesions has also been suggested by other authors [9]. However, an increase in invasivity and economical costs can be assumed.

In conclusion, due to the rarity of WT in adults, no firm treatment guidelines have been established to date. Adult WT is treated according to recent pediatric protocols. In earlier reports it was agreed that all stages of adult WT warrant aggressive multimodal therapy [surgery, chemotherapy, and radiotherapy] because of its poor prognosis.

Furthermore, our case confirms that careful detachment of the IVC from the liver, as commonly performed in transplant liver surgery, can be a safe and effective surgical procedure for patients affected by Level III caval thrombosis to improve the outcome.

#### Conflict of interest

The authors have declared that no conflict of interest exists.

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The authors have declared that any sources of funding for this report.

#### Ethical approval

No ethical approval.

#### Author contributions

Dr. Morabito was involved in writing the paper and concept. Dr. Guglielmo was involved in data collection. Dr. Melandro was involved in follow up and data collection. Prof. Mazzei was performed as a supervisor. Dr. Alesini was involved in pathology analysis. Prof. Bosco was performed as a pathology supervisor and involved in final analysis of slides. Prof. Berloco was performed as a final supervisor and involved in study design.

#### Key learning points

- The key is the type of surgical procedure to treat the tumor. This approach can be used to determine a radical resolution of tumor mass with a good prognosis. In fact the follow up confirm that this approach is safe and in association with drug therapy can improve the survival.

#### References

- [1]. Kaur N, Gupta A, Attam A, et al. Adult Wilms Tumor: management considerations. *Int Urol Nephrol* 2005;**37**:17–20.
- [2]. Langenburg SE, Blackburne LH, Sperling JW, et al. Management of renal tumors involving the inferior vena cava. *J Vasc Surg* 1994;**20**:385–8.
- [3]. Tongaonkar HB, Dandekar NP, Dalal AV, et al. Renal cell carcinoma extending to the renal vein and inferior vena cava: results of surgical treatment and prognostic factors. *J Surg Oncol* 1995;**59**:94–100.
- [4]. Kuczyk MA, Bokemeyer C, Kohn G, et al. Prognostic relevance of intracaval neoplastic extension for patients with renal cell cancer. *Br J Urol* 1997;**80**:18–24.

- [5].Ljungberg B, Stenling R, Osterdahl B, et al. Vein invasion in renal cell carcinoma: impact on metastatic behavior and survival. *J Urol* 1995;**154**: 1681–4.
- [6].Nesbitt JC, Soltero ER, Dinney CP, Walsh GL, Schrupp DS, Swanson DA, et al. Surgical management of renal cell carcinoma with inferior vena cava tumor thrombus. *Ann Thorac Surg* 1997;**63**:1592–600.
- [7].Gallucci M, Borzomati D, Flammia G, et al. Liver harvesting surgical technique for the treatment of retro-hepatic caval thrombosis concomitant to renal cell carcinoma: perioperative and long-term results in 15 patients without mortality. *Eur Urol* 2004;**45**:194–202.
- [8].Mitry E, Ciccolallo L, Coleman MP, et al. Incidence of and survival from WILMS TUMOR in adults in Europe: data from the EUROCARE study. *Eur J Cancer* 2006;**42**:2363–8.
- [9].Ohwada S, Satoh Y, Nakamura S, Tanahasi Y, Otani Y, Lino Y, et al. Left-sided approach to renal cell carcinoma tumor thrombus extending into suprahepatic inferior vena cava by resection of the left caudate lobe. *Angiology* 1997;**48**:629–35.

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