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Fetal rhabdomyomatous nephroblastoma -a variant of Wilms tumor: A case report and review of the literature

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ABSTRACT

Fetal rhabdomyomatous nephroblastoma is a weird variant of Wilms tumor. It's a chemo-resistant tumor, characterized for invading the collecting system, and having greater rates of bilateral involvement. The tumor chiefly consists of fetal striated muscle with particularly distinct striations and central nuclei, and isolated regions of typical trimorphic nephroblastoma. The following article brings in a new case of this rare pathology, associated with a review of the literature

Key Words: Fetal rhabdomyomatous nephroblastoma; chemotherapy; Wilms tumor.

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Introduction

Fetal rhabdomyomatous nephroblastoma (FRN) is a weird variant of Wilms tumor (WT), first described by Eberth in 1872 as "myxoma sarcomatodes renum". The tumor

particularly distinct striations and central nuclei, and isolated regions of typical trimorphic nephroblastoma. It is a chemo-resistant tumor, characterized by invading the collecting system, and having greater rates of bilateral involvement [1-9]. The following article brings the principal clinic, therapeutic and evolutive characteristics in a new case of this rare pathology and a review of the literature.

Case report

23-month-old girl with a palpable abdominal mass of a month of evolution. On physical examination, the patient was in good general conditions, adequate weight and height for age. A large firm mass was palpated in the right l circulation and hepatomegaly, extending to the left hypochondrium.

Abdominal ultrasonography showed a solid mass of 125x128x110 mm replacing the right kidney. Contrast enhanced thoraco-abdominal computed tomography (CT) showed a right kidney lesion of 135x130x128 mm, suggestive of WT, with anterior displacement of the right

lobule of the liver and retroperitoneal structures; collapsed inferior vena cava and unspecific subpleural nodules in superior lobules [**Fig. 1**].

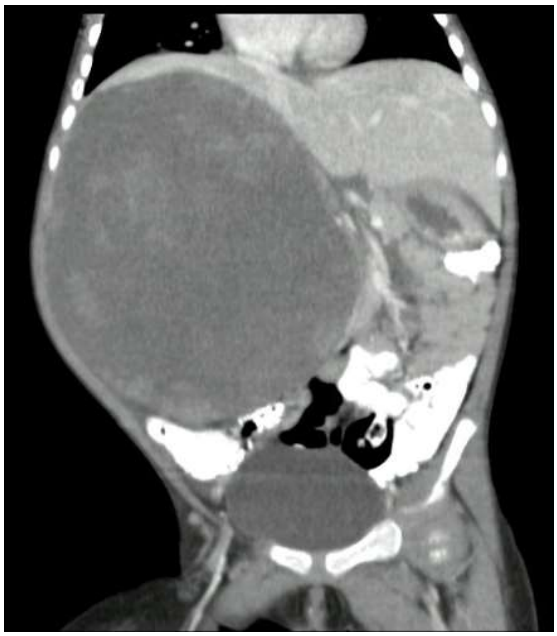


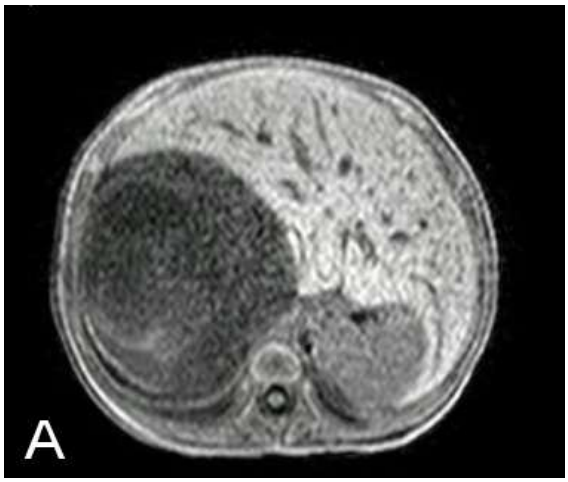
Fig. 1. Contrast enhanced CT, showing a right renal mass of 135x130x128 mm, suggestive of Wilms tumor.

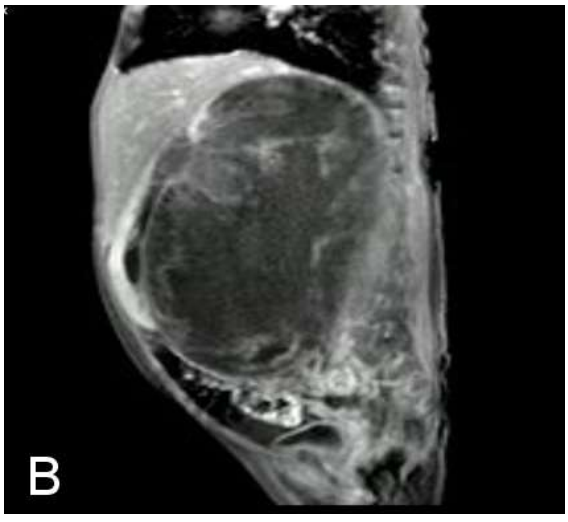
Elevated blood pressure was found, with diagnosis of state 2 hypertension, without vital organs compromised. Amlodipine, clonidine and captopril were indicated.

An abdominal mass suggestive of WT as a first option was considered, but a neuroblastoma was also considered because of the early appearance. Taking into account the possible metastatic disease in the lung, the size of the lesion, the involvement of near structures, and the high risk of intraoperative tumoral rupture, neoadjuvant chemotherapy was initiated, with the following protocol:

Week 0: Actinomycin 45ug/kg,
Week 1: Vincrystin 0.05mg/kg,
Week 2: Vincrystin 0.05mg/kg,
Week 3: Vincrystin 0.05mg/kg,
plus Doxorrubycin 1.5 mg/kg,
Week 4: Vincrystin 0.05mg/kg,
Week 5: Vincrystin 0.05mg/kg.

She presented fever during the third week of treatment, associated with abdominal pain, without observing decrease in the size of the mass in physical examination or improvement of blood pressure rates. 24 hours' urine catecholamine's were taken to rule out neuroblastoma (vanillymandelic and homovanillic acid), with normal results. Control magnetic nuclear resonance showed a kidney dependent abdominal mass occupying the right hemi abdomen, of greater size compared to previous images [**Fig. 2**].





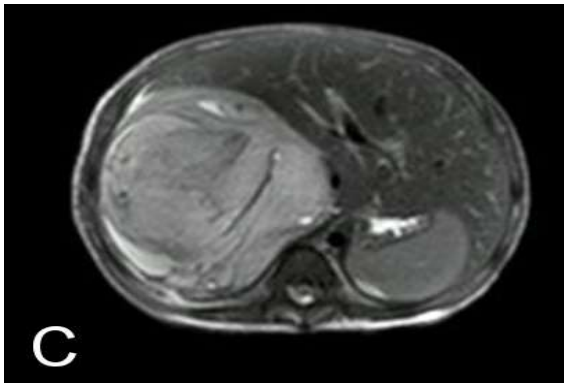


Fig. 2A-C. Magnetic nuclear resonance, showing increase of the tumoral mass, with bleeding and necrosis areas **(A)** Axial T1. **(B)** Sagittal **(C)** Axial T2.

The chemotherapy protocol continued for 5 more weeks. A right radical nephrectomy was performed, finding an approximately 20 cm tumor, adhered to the vena cava and right kidney artery, with solid consistence and cystic areas, plus enlarged interaortocaval lymphatic nodes up to 1.5 cm. Due to 650 cc of bleeding, she required transfusion of 400 cc of packed red cells and 160 cc of plasma.

Post-operative evolution was satisfactory, without complications, and improvement of blood pressure values. Adjuvant chemotherapy continued (week 6), meanwhile the pathology report was ready.

The final pathology report showed an irregular, light brown mass that weighted 1.249 grams, and measured 17x13x11 cm, with solid and necrotic areas. A renal remnant of 3x3x2 cm was found. It was classified as a FRN, with the following histologic findings; Tumor mass, predominantly stromal, with occasional mitosis, trapped renal tubules, skeletal muscle heterologous differentiation and fusocellular mesenchymal differentiation.

Immunohistochemistry showed positivity in tumoral cells with Vimentine and CD34 in the stromal component; myogenin in the heterologous differentiation component, and WTi in the tubular epithelial component, as EMA (epithelial membrane antigen). The cellular proliferation index, Ki67, was 10-20% [Fig. 3].

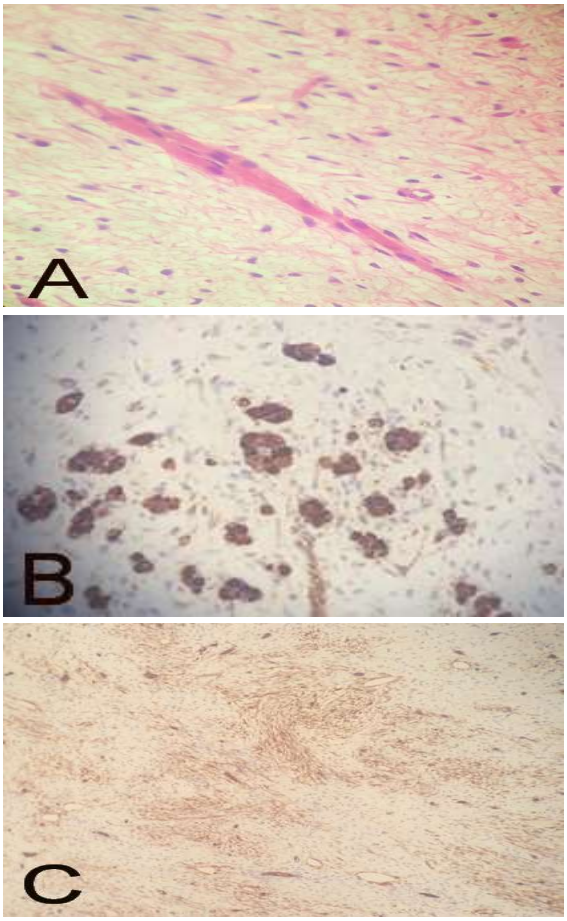


Fig. 3. Histology and immunohistochemistry. **(A)** Striated muscle heterologous differentiation. **(B)** Positive WTi in the epithelial tubular component. **(C)** Positive CD34 in the stromal component.

PET scan was done 2 months after surgery, without finding lesions that suggested the presence of tumoral disease. Expectant management was decided.

Discussion

FRN is a monophasic mesenchymal variant of WT [1], with an estimated prevalence of 1.5-4.5%. It's bilateral in 30% of cases. Local relapses and the presence of metastatic disease

imply a worse prognosis. This tumor doesn't respond well to chemotherapy protocols (because of its mesenchymal component: fetal rhabdomyomatous tissue) [2]. Wigger [1] first used the term fetal rabdomyomatous nephroblastoma in 1976.

It is microscopically characterized by striated muscle with different striations and central nuclei. The histology of this muscle reminds that to fetal striated muscle, and it's accompanied by undifferentiated tissue areas, neoplastic epithelium and mesenchymal components, such as adipose and myxoid tissue, and cartilage islands.

It is microscopically and macroscopically similar to congenital mesoblastic nephroma, and the only difference is quantitative, because in this one there is a predomination of smooth muscle, while in FRN the presence of striated muscle fibers predominates [3]. Nonetheless, congenital mesoblastic nephroma has unique characteristics, as it is considered the most common solid tumor of the newborn, usually identified in the first 3 months of life.

It's origin is given by proliferation of early nephrogenic mesenchymal tissue with monomorphic histology, mesenchymal cells proliferations, and embrionary metaplasm soaked in normal renal tissue. It's usually benign, with complete healing after a nephrectomy with wide margins [4].

It's known that FRN is usually of greater size than WT, but its behavior in a complete resection scenario is less aggressive. There is an important difference in the age of presentation, according to Wigger, FRN predominates in 1-year-old [1], and WT presents in patients of 2-3 years old. There aren't reported cases in patients older than 4 years old.

Beckwit et al. [5] described that the clinical behavior of nephroblastoma is defined by the aggressiveness of the tumor. A blastomatous predominant WT is associated with a high aggressiveness pattern (76% of cases in 3-4 stage), but due to its good answer to chemotherapy, free disease survival rates are high. FRN is usually diagnosed in early stages (80%), but it is chemo resistant, so patients present low survival rates if the tumor is not totally resected.

Pollono [6] described a 14 patient's cohort diagnosed with FRN, with a mean age of 27 months, and a bilateral presentation of 22%. After cytoreduction and adjuvant therapy, only 6 patients were free of disease, and 8 had died. The same findings were reported by Saba [7], and Maes et al [8], whom noted poor survival rates in patients with FRN taken into incomplete resection of the tumor.

Anderson proposed the hypothesis that a poor response to chemotherapy in patients with bilateral WT was associated with post chemotherapy presence of rhabdomyomatous histology [9]. He demonstrated a significant association between post chemotherapy rhabdomyomatous differentiation and poor radiologic answer.

Tumor size has been employed as a prognosis indicator, due to the fact that in classic nephroblastoma, it is inversely related with survival rates [10,11]. Some authors have described that volumes greater than 551 cc, survival rates are close to zero. Maes found a mean volume of 965 cc (17.3 to 2520 cc) in patients with FRN [8]. However, the mean survival rate in these patients was of 4 years.

There is no way to differentiate FRN from classic WT in initial studies. Nonetheless, it is important to emphasize that initial study of an abdominal mass is done with ultrasonography. When solid lesions suggestive of malignant disease are found, a contrast-enhanced CT must be done, because of its greater sensibility

for establishing vascular, local and lymphatic extension [12]. Magnetic resonance is the imaging study of choice to determine vascular involvement [12].

Conclusions

FRN is a variant of WT, with better prognosis when local control is adequate. Diagnosis is histological. It responds poorly to chemotherapy, so an aggressive resection is important to achieve greater survival rates.

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