



How to Distinguish Non-Metastatic Rhabdoid Tumor Form Wilms Tumor in Young Infants Preoperatively: A Case Report and Literature Review

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Abstract

Background: Rhabdoid tumor of the kidney is an uncommon and aggressive tumor characterized by poor outcomes. Given the rarity of this tumor, the diagnosis is still a challenge and the treatment is not well codified.

Case Report: A 4-month-old girl presented with abdominal mass. Clinical and radiologic explorations suggested nephroblastoma. The diagnosis of rhabdoid tumor was made post operatively based on anatomopathological findings.

Keywords: Renal; Rhabdoid; Wilms; Child

Introduction

Rhabdoid Tumor of the Kidney (RTK) is an aggressive tumor, which occurs in infancy, and early childhood accounts for 2% of renal tumors in childhood [1]. It was initially described as sarcomatous type of wilms tumor because of the histologic resemblance of its cells to rhabdomyoblasts. Rhabdoid tumor is recognized now as a distinct entity. Clinical presentation and radiological explorations do not often differentiate it from a Wilms Tumor (WT). Whereas the overall survival rate for WT exceeds 85%, the survival rate for RTK is only 23% [2]. We present a case of RTK considered initially as nephroblastoma and describe the clinical and radiologic findings and management.

Case Presentation

Our patient was preterm born at 34 weeks of gestation *via* caesarian section to a healthy 34 years old mother and was conceived through *in vitro* fertilization; the pregnancy was well followed-up and normally unrolled. The prenatal trimestral ultrasound was normal. No congenital anomalies or malignancies were noticed in the family history. The birth weight was 2 kg 300 gm. The patient was admitted at the age of 4 months in our department for vomiting and fever. The parents said that they noticed an abdominal distension few weeks earlier. The physical examination showed a right lumbar mass. The abdominal ultrasound followed by the CT scan showed a large heterogeneous retroperitoneal mass with scattered calcifications measuring 10 cm. The tumor was predominantly solid and focally cystic with the presence of areas of hemorrhage and necrosis (Figure 1). It was at expense of the right kidney and did not invade vascular structures or nearby organs. Biologic explorations showed anemia, hypercalcemia and normal amounts of catecholamines. The diagnosis of nephroblastoma was highly suspected. The patient didn't undertake pre-operative chemotherapy and had surgical treatment consisting on enlarged nephroureterectomy as recommended by the SIOP. During surgery, the tumor was encapsulated occupying the entire kidney and was intimately adherent to the right renal vein, the inferior vena cava. A total macroscopic resection of the tumor was realized. The patient died after the surgery due to a hemodynamic instability. The macroscopic examination of the specimen showed a large encapsulated tumor at the right kidney and measuring 12 cm × 6 cm × 5 cm. The microscopic examination found sheets of cells with an eosinophilic cytoplasm and a large nucleus showing a central nucleolus. Numerous mitoses were seen and areas of hemorrhage and necrosis were present. The immunochemistry indicated vimentin and EMA positivity and parts of cells expressed keratin and cyclin D1. INI1, AML and Desmin in the tumor

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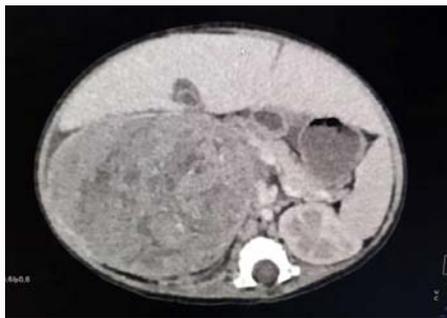


Figure 1: CT scan showing the heterogeneous renal tumor with areas of necrosis and scattered calcifications.

cells were negative. The diagnosis of RTK was based on these findings.

Discussion

The RTK is considered one of the most lethal malignancies in pediatric oncology with an overall survival rate of less than 25%. It occurs predominantly during early childhood. Eighty percent of cases are diagnosed at an age less than two years. The median age reported by SIOP was 13 months and by NWTSG was 10.6 months [2]. However the mean age at diagnosis in wilms tumor is 36 month [2,3]. The primary site of rhabdoid tumor is renal at a rate of 45%, the central nervous system at a rate of 18% and other tissue at a rate of 35% [4]. Epidemiology features suggest that *in vitro* fertilization, preterm birth and low birth weight are associated with a higher risk of occurrence of the rhabdoid tumor [5,6]. Our patient had all these features but in front of the age and radiologic findings the diagnosis of wilms was evoked. The discovery of an abdominal mass during physical examination is the most constant clinical sign, but this cannot guide the diagnosis. Some clinical features can suggest the diagnosis of RTK from other tumors: fever, hematuria, a young age and high tumor stage [7]. Rhabdoid tumors can be seen in the newborn and are in this case, possibly revealed by subcutaneous tumor nodules [8]. Symptoms may also be preferable to metastasis. Common metastatic site include the lung, the brain and bone. The biological assessment can reveal abnormalities such as anemia and hypercalcemia [7,9]. Hypercalcemia is rare in case of wilms tumor however it occurs in approximately 20% of rhabdoid tumors [3]. It is attributed to the ectopic production of parathyroid hormone related protein by the tumor. Our patient presented persistent hypercalcemia needing a preoperative correction during 2 weeks consisting on hydration and bisphosphonate therapy. This hypercalcemia was considered as a paraneoplastic syndrome. In our case the diagnosis of mesoblastic nephroma was initially evoked accounting for the age of patients. However subsequent radiologic features eliminated this diagnosis. In radiological explorations, rhabdoid tumors appear similar to nephroblastoma but certain features can orient the diagnosis: subcapsular fluid collection with areas of hemorrhage and necrosis is more correlated with a rhabdoid tumor (71% vs. 12% of wilms tumors) [10,11]. A multilobulated architecture especially when surrounded by low attenuation material, a sign of hemorrhage and necrosis, is more present in rhabdoid tumor [11-13] and calcifications are encountered in 66% of rhabdoid tumors vs. 6% to 15% of wilms tumors [11,13,14]. Calcifications and necrosis were present in our case but considering the fact that there were no specific signs and given the rarity of rhabdoid tumors, it was considered as Wilms tumor.

Seventy percent of patients had advanced stage at the time of

diagnosis [2-4] and 24% had metastasis [3]. In the case of our patient, the tumor would have been classified as stage I.

In the cytopathologic study, tumors are typically solid with extensive hemorrhage and necrosis; the cells are characterized by eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. In immunochemistry, it is characterized by positivity to vimentin and negativity to INI1 [15].

The prognosis is related to the age at the time of diagnosis [2,3,16]. Diagnosis at an early stage and the use of radiotherapy are also associated with better outcomes [17]. The preoperative chemotherapy does not seem to improve the survival rate of patients with RTK [2,3,18] even if it seems to decrease the tumor volume [3]. Given the rarity of this tumor there is no standard treatment option for RTK [18,19].

Conclusion

To conclude epidemiologic features such as low birth weight, age less than 2 years, history of *in vitro* fertilization, biologic elements such as hypercalcemia, anemia and calcifications in CT Scan may orientate to the diagnosis of rhabdoid tumor, and should be included in a diagnostic score that would guide the therapeutic attitude.

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