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Rhabdoid Tumor: Imaging Findings of Rare Renal Tumor in Children

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Abstract **Case Report**

Rhabdoid tumors of the kidney are rare childhood renal neoplasms, extremely aggressive and typically carrying a grim prognosis, its common correlation with primary or metastatic central nervous system (CNS) lesions these tumors primarily impact infants, with an average age of onset at 11 months (ranging from birth to 9 years [1]. Initially thought to be a sarcomatoid subtype of Wilms' tumor, it is now recognized as a separate pathological entity [2]. At the beginning of the illness, typical clinical signs in clued hematurie, abdominal pain, and abdominal swelling. Imaging studies are the primary method used to diagnose renal masses in children.

Keywords: Rhabdoid tumors, kidney, nervous system, abdominal swelling.

INTRODUCTION

The rhabdoid tumor of the kidney is a very rare tumor entity. Only about 2% of pediatric renal tumors are rhabdoid tumors. 80 % of patients are under 2 years old [3]. It is considered the most aggressive malignant tumor of the kidney in children and must be distinguished from nephroblastoma. The determining prognostic factors for the therapeutic course of renal tumors in children are the degree of malignancy and metastasis. 90% of patients with nephroblastoma can be cured, in contrast, rhabdoid tumor has a very unfavorable prognosis. It is considered therapy-refractory and quickly leads to death due to rapid progression [4, 5].

CASE REPORT

A 6-month-old girl with significant abdominal distension (Figure 1) for a week accompanied by gastrointestinal issues (diarrhea, vomiting), in a febrile context. The clinical examination reveals an abdominal mass extending to the right iliac fossa; umbilical hernia with collateral circulation is present On ultrasound: a large mass involving the right kidney measuring approximately 8 x 6 cm, with tissue-like echotexture, hypoechoic, heterogeneous, containing areas of hyperechoic and anechoic zones, vascularized on color Doppler. The CT scan with intravenous contrast, and non-contrast intravenous shows a large right renal mass with a solid-cystic appearance, extensively necrotic, containing air bubbles, associated with a large amount of peritoneal effusion (Figure 2). With histopathologic diagnosis of rhabdoid tumor of the kidney."

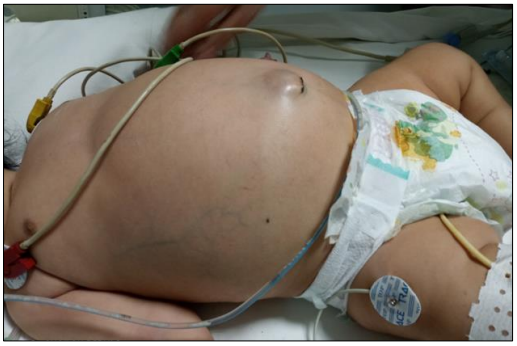

Figure 1: 6 month sold girl with significant abdominal distension

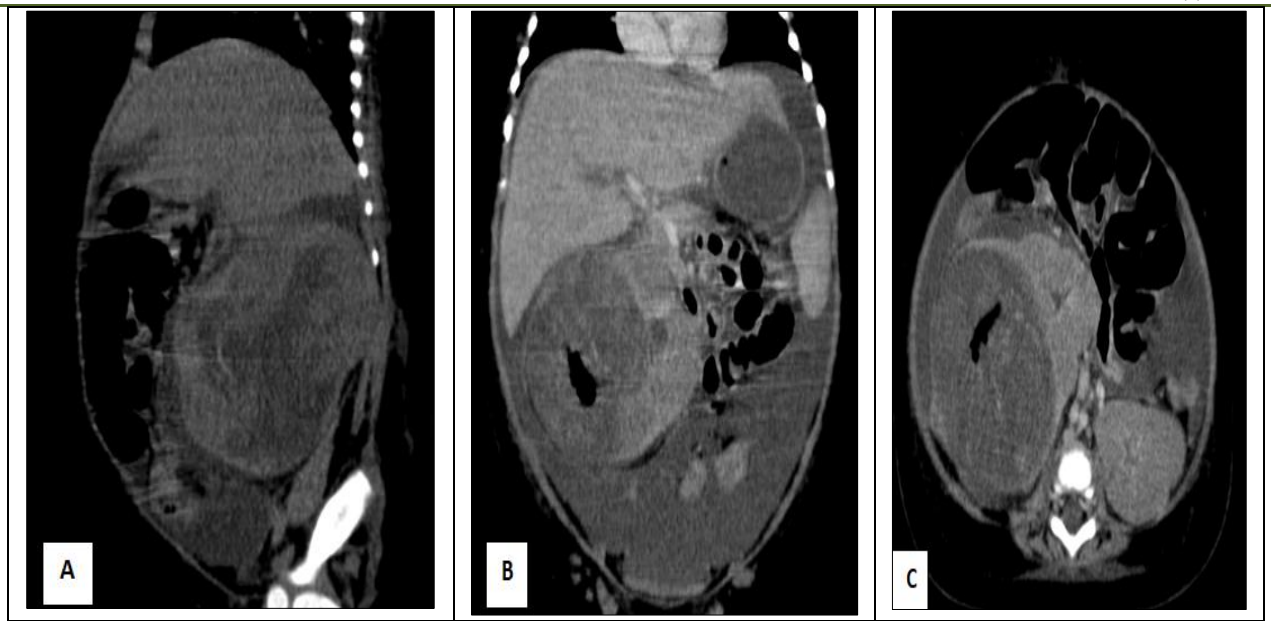

Figure 2: CT scan sagittal slice with non contrast intravenous (A) axial and coronal slices with intravenous contrast (B , C) , shows a large right renal mass with a solid-cystic appearance, extensively necrotic, containing air bubbles, associated with a large amount of peritoneal effusion

DISCUSSION

The rhabdoid tumor of the kidney is recognized as an uncommon malignant tumor in children, with an unknown histologic origin, which differs from Wilms' tumor [6, 7]. Despite its name, "rhabdoid," which suggests histological resemblance to rhabdomyosarcoma, the tumor cells do not exhibit myogenous characteristics [1]. Rhabdoid tumors might initially present with hematuria, but owing to their aggressive nature, symptoms may be indicative of metastatic spread. Hypercalcemia related to an inappropriate release of parathormone has also been reported [9].

The link between rhabdoid tumors and concurrent or subsequent primary intracranial masses or brain metastases has been identified as a distinguishing characteristic. The brain lesion typically occurs close to the midline and frequently involves the posterior fossa. Cases of primitive neuroectodermal tumors, ependymomas, as well as cerebellar and brainstem astrocytomas have all been reported in association with rhabdoid tumors. The majority of tumors are large, centrally located, and diagnosed at stage III or IV. The 18-month survival rate is only 20%, with no reported survivors at the 5-year mark [1].

The heterogeneous pattern has been documented in cases of rhabdoid tumor of the kidney. This characteristic remains consistently present among the imaging findings of RTK, whether observed through sonography, CT scans, or MRI [2-9]. This is associated with necrotic and/or hemorrhagic regions within the tumor. Upon diagnosis, the tumor presents as bulky (with a diameter exceeding 4 cm). Its rapid growth has been highlighted. The occurrence of RTK in the central region of the kidney has been observed in three out of the five cases reported in the literature; a mass arising from the upper or the lower pole has been rarely documented. Subcapsular fluid collection, irregular thickening of the renal capsule, calcifications of RTK, and bilateral lesions are possible [8]. The radiographic presentation of RTK might resemble that of a typical Wilms' tumor or another malignant kidney neoplasm like clear cell sarcoma. It could also mimic a benign mass such as mesoblastic nephroma (Boland's tumor), which often occurs during the first months of life [12]. However, cerebral MRI or CT and a thoracic CT scan are indicated after histological confirmation due to the frequent metastasis to the lungs.

The histological diagnosis of RTK is based on the detection of a particular type of cell which is characterized by a round, vesicular nucleus and a large central eosinophilic nucleolus. In addition, these cells may have eosinophilic cytoplasmic inclusions [10].

CONCLUSION

Rhabdoid tumor has the worst prognosis of all renal tumors. It is highly aggressive and metastasizes early, with most patients presenting with advanced disease.

CT findings of calcification, subcapsular hematoma, and a lobulated appearance in a large, centrally located, and heterogeneous renal mass in a child are indicative of a rhabdoid tumor of the kidney.

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