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Case Report

Using imaging to diagnose renal tumors beyond nephroblastoma ☆,☆☆

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ABSTRACT

Wilm's tumor (WT) accounted for the vast majority of renal tumors in children (92%). However, there are many atypical or rare forms of kidney cancer, and it is certainly useful to have a diagnostic orientation in imaging to differentiate between the different diagnoses, guiding that way the therapeutic management.

We report the cases of 3 patients who were initially diagnosed with nephroblastoma on the basis of radiological data (via PACS search), underwent pre-operative chemotherapy and then nephrectomy, and whose anatomopathological evidence came back in favor of benign renal tumors.

Nephroblastoma is the most common renal tumor in children, but other benign tumors should not be neglected, radiological signs that may point to this entity should be taken into account, so that neoadjuvant chemotherapy with its enfeebling side effects can be avoided.

Introduction

The study of pediatric renal tumors has had a significant impact on the field of oncology. Wilm's tumor (WT) represented the large majority of renal tumors in children (92%). However, with the latest WHO 2016 classification, pathologists have identified more than 40 histological types of renal tumor [1].

These tumors have different clinical presentations and evolutionary risks, with highly heterogeneous and sometimes highly misleading histological aspects, and specific cytogenetic and molecular profiles.

Abbreviations: CT, computerized tomography; RMI, remote medical imaging; MSD, stromal metanephric tumor; MAF, metanephric adenofibroma; MA, metanephric adenoma; CMN, congenital mesoblastic nephroma.

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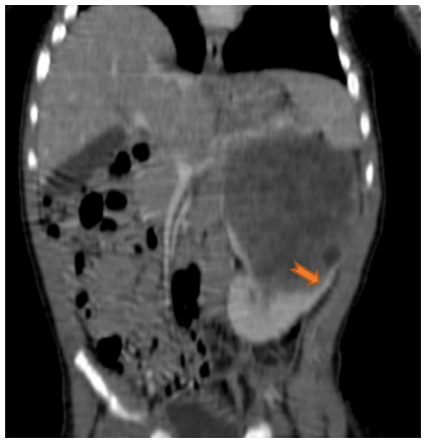

Fig. 1 – Coronal slice CT scan through the abdominal region, showing a shoulder sign (arrow) in favor of an upper and medio-polar left renal tissue mass.

Many of them perfectly mimic Wilms' tumor clinically and radiologically, and some cases have been published in the literature where nephroblastoma was suggested and the biopsy evoked another diagnosis, such as neuroblastoma or congenital mesoblastic nephroma [2,3].

As a result, the histopathological diagnosis of renal tumors determines the patient's therapeutic management.

The aim of this study is to determine the value of imaging in correcting the diagnosis and characterizing each histological type.

Case presentation

Case 1 : Stromal metanephric tumor

We report the case of an 8-month-old child from a consanguineous marriage, pregnancy carried to term with good adaptation to extrauterine life, good psychomotor development, vaccinated according to the national immunization programme, whose clinical history dates back 15 days with the appearance of abdominal pain with constipation, and deterioration in general condition.

On clinical examination, the patient was in good general condition, with an estimated weight of 8 kg, however, the abdominal examination revealed a left lumbar mass.

The patient had initially abdominal ultrasonography, which revealed a mass with 2 component, echogenic tissue which is the predominant composant with central and peripheral anechogenic cystic zones. This mass is forming a shoulder sign with adjacent healthy renal parenchyma.

The cathecolamines were negative in the biological test and the CT scan revealed a polar superior and medio-renal left

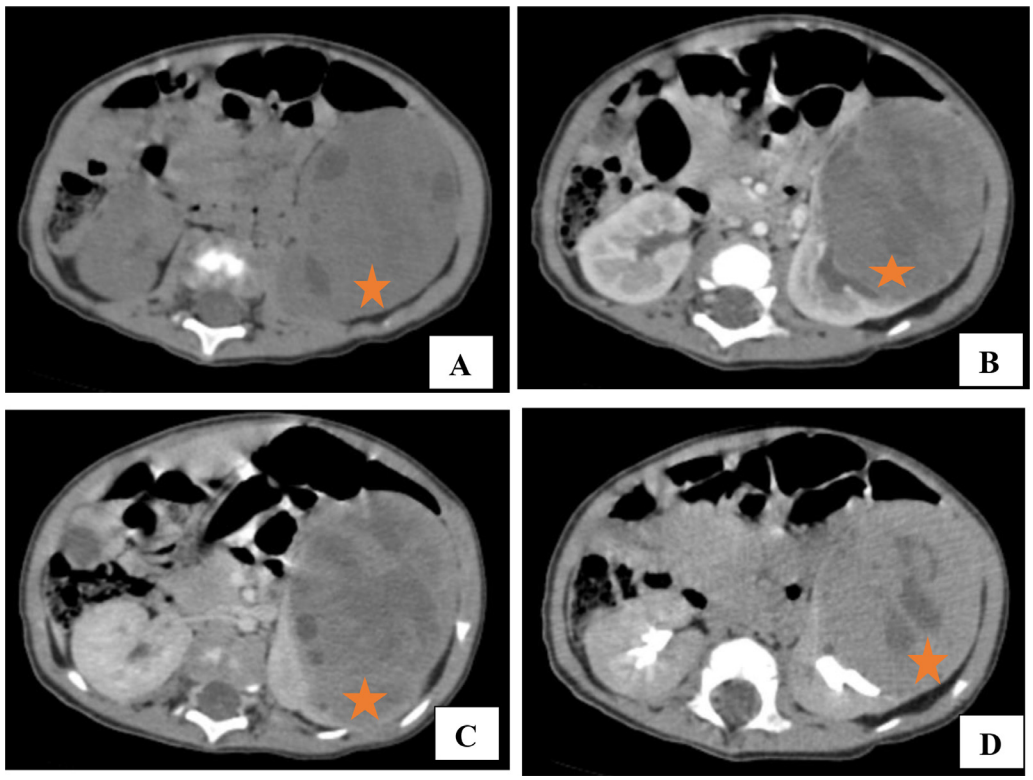

Fig. 2 – Axial-slice CT scan through the abdominal region in spontaneous contrast (A), at arterial (B), venous (C) and late times (D) ; showing progressive enhancement of the upper and medio-polar left renal mass (star), indicating its fibrous nature.

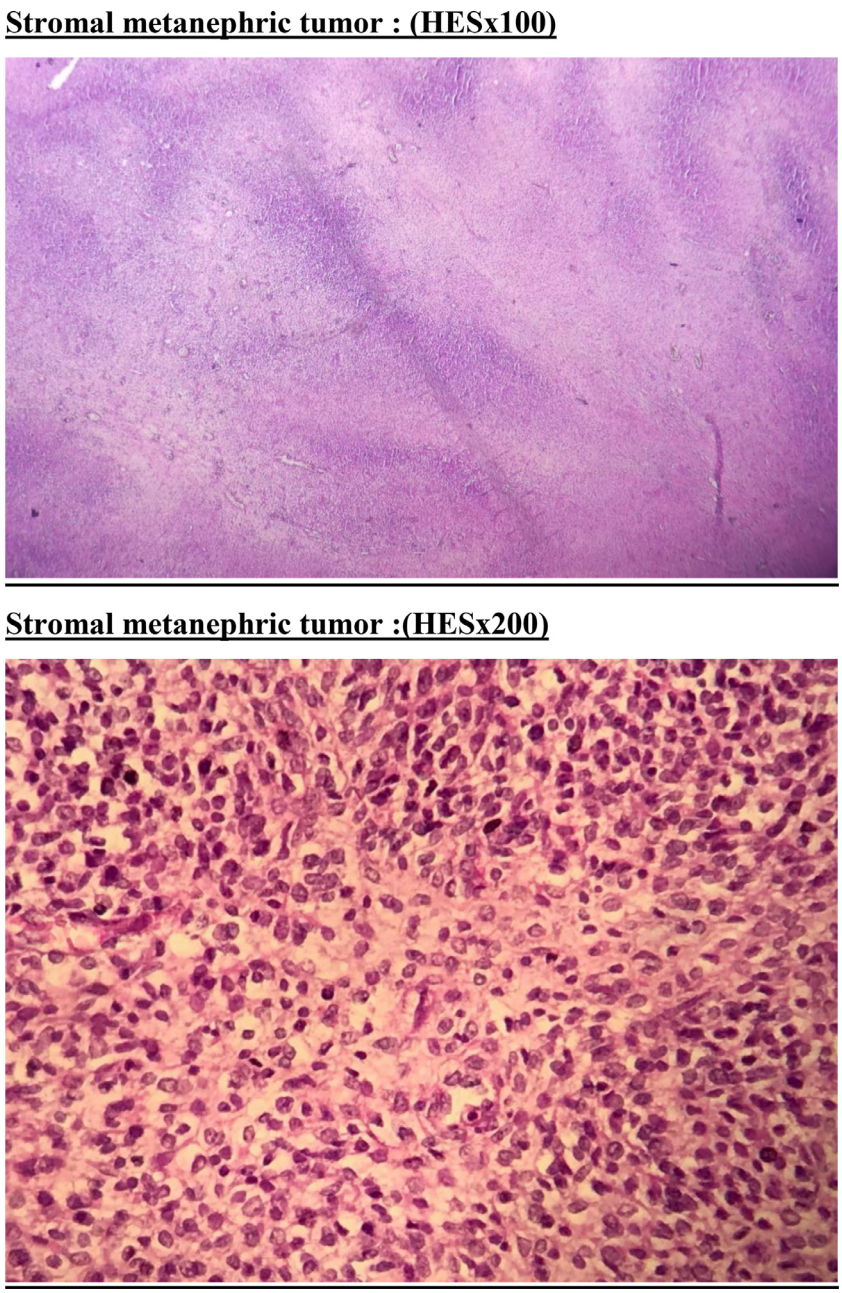

Fig. 3 – Proliferation is sometimes made up of dense, very basophilic foci, composed of rounded cells, and sometimes of eosinophilic foci, less cellular.

tumoral process, with poly-lobed contours, solidocystic, with a majority solid component isodense in spontaneous contrast, heterogeneously enhanced containing hypodense areas unchanged by contrast injection with peripheral and central cystic component (Figs. 1 and 2).

The patient received 2 months of pre-operative chemotherapy, then he had total nephrectomy, and the pathological study showed a tumor whose histological and immunohistochemical aspects were in favor of a benign metanephric stromal tumor (Fig. 3).

Case 2: Cystic nephroma

We report a second case about a 16-month-old female infant, from a non-consanguineous marriage, pregnancy carried to term with good adaptation to extrauterine life, good psychomotor development.

The symptomatology began 3 months ago with the appearance of an abdominal mass on the right flank, progressively evolving in a context of apyrexia, without any hemorrhagy syndrome or hematuria.

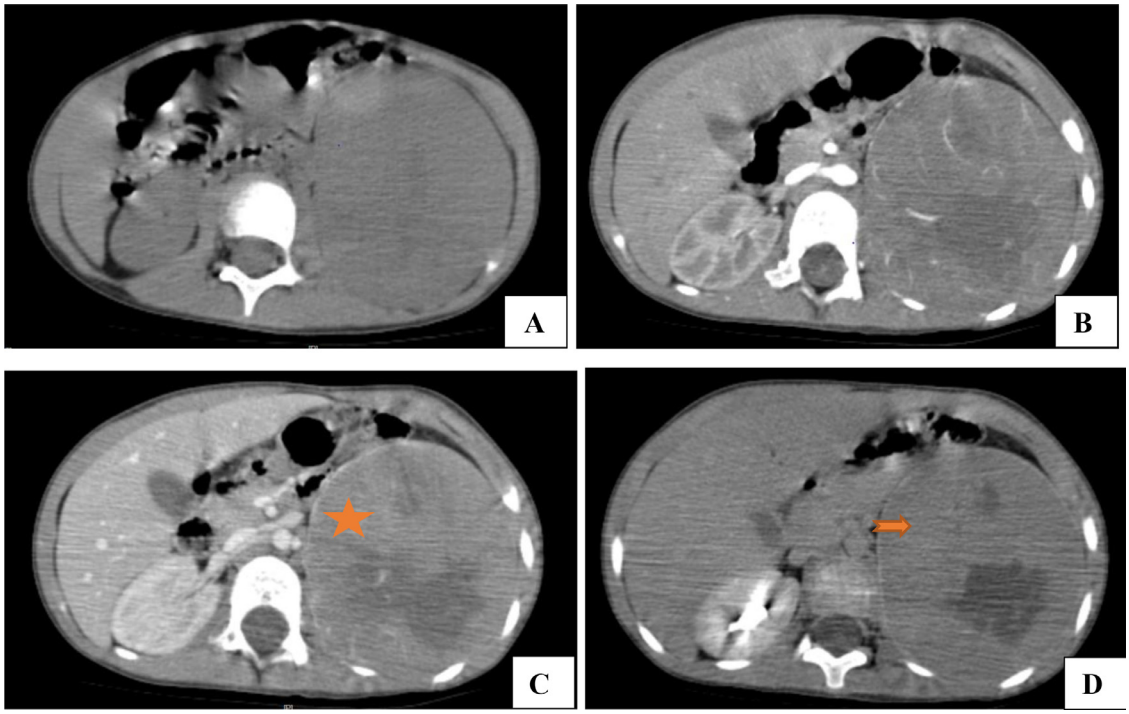

Fig. 4 – Axial section CT scan of the abdominal region in spontaneous contrast (A), at arterial (B), venous (C) and late times (D) ; showing early enhancement of the left renal mass (star), with washout at late excretory time (arrow) of nephroblastoma tumor tissue differentiating it from late enhancement of fibrous tissue in metanephric stromal tumor.

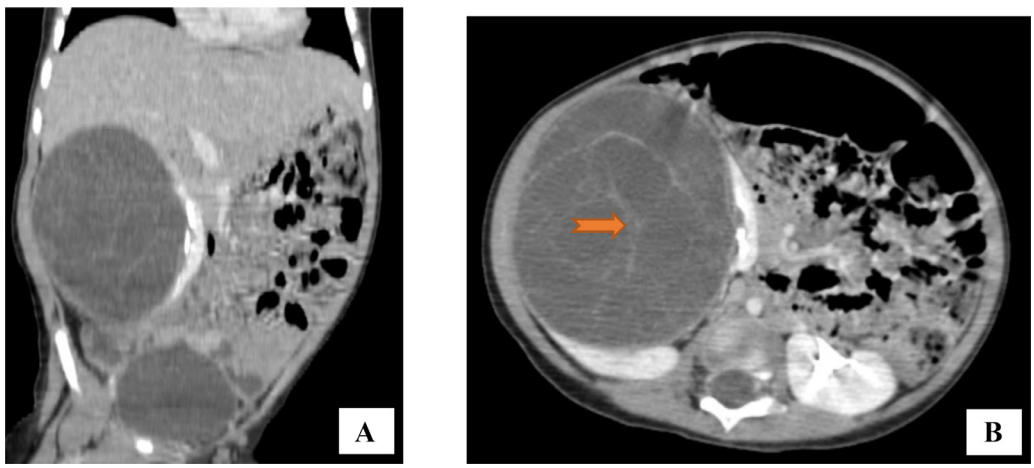

Fig. 5 – Coronal (A) and Axial (B) slice CT scan of the abdominal region showed a multilocular cystic right renal mass, enclosing thick contrast-enhancing septa (arrow).

Abdominal examination revealed an abdominal mass on the right flank extending to the right iliac fossa, about 14 cm long, oval in shape, firm in consistency, attached to the deep plane with lumbar contact.

The patient underwent a CT scan, which showed a right mediorenal nephroblastoma with no signs of locoregional extension (Fig. 5), and the biological tests reveals a negative catecholamines.

The patient also received preoperative chemotherapy, then benefited from a total nephrectomy, and the anatomopathological study showed a tumor whose histological and immunohistochemical aspects were in favor of cystic nephroma.

Case 3: congenital mesoblastic nephroma

The third case is about a 9 months old infant, from a non-consanguineous marriage, pregnancy carried to term with good adaptation to extrauterine life, good psychomotor development, vaccinated according to the national immunization programme.

The symptomatology began 1 month ago with the appearance of an abdominal mass on the right flank, progressively evolving.

Abdominal examination revealed an abdominal mass on the right flank extending to the right iliac fossa, approximately 10 cm long, oval, fixed to the deep plane with lumbar contact.

The CT scan revealed a well-defined right renal mass with regular contours and heterogeneous enhancement (Fig. 8), wich histological and immunohistochemical appearance was in favor of a congenital mesoblastic nephroma in its classical form (Fig. 9).

Discussion

Renal tumors in children include malignant tumors such as Wilm's tumor (or nephroblastoma, (90%), clear cell sarcoma (3%) and rhabdoid tumor (2%), as well as benign mesoblastic nephroma (2%).

The latest WHO histopathological classification of renal tumors classifies the different tumor types according to their histological type, the aim of this classification is to highlight the macroscopic characteristics and to correlate them with the radiological aspects.

This classification identifies [4]:

- **Renal cell tumors**
- **Metanephric tumors**
- **Nephroblastic tumors**
- **Mesenchymal tumors**
- **Mixed mesenchymal and epithelial tumors**
- **Neuroendocrine tumors**
- **Hematopoietic and lymphoid tumors**
- **Germ cell tumors**
- **Metastatic tumors**

In the category of benign tumors, this classification includes in:

- Metanephric tumors: Metanephric adenoma, Metanephric adenofibroma, and Metanephric stromal tumor.
- Mixed epithelial and mesenchymal: Cystic nephroma.
- Mesenchymal tumors: Congenital mesoblastic nephroma and ossifying renal tumor of children.

Metanephric stromal tumors, congenital mesoblastic nephroma, and cystic nephroma are rare tumors of the kidney.

They are often asymptomatic and are discovered by chance, mainly in newborns or during infancy, at the same time as clear cell sarcoma of the kidney, rhomboid tumor of the kidney, ossifying tumor of the kidney of childhood and Wilm's tumor, with which they can easily be misdiagnosed.

No attempt has yet been made to clearly document the imaging characteristics of each of these different tumors.

Metanephric tumors are rare benign tumors forming a lesion spectrum with a purely epithelial tumor (metanephric adenoma), a purely stromal tumor (metanephric stromal tumor) and a biphasic epithelial-stromal tumor (metanephric adenofibroma).

Adenofibromas and stromal tumors are capsule-free and frequently involve renal glomeruli and tubules in the periphery. They may extend into the renal sinus and perirenal fat.

Metanephric adenofibromas are mixed tumors with a highly variable proportion of epithelial and stromal components metanephric adenoma and metanephric stromal tumor.

The diagnosis of metanephric stromal tumor requires extensive sampling of the tumor to rule out a possible focal epithelial contingent. The stromal component is qualitatively comparable in these 2 entities [5].

It is described us a non-enhancing unencapsulated unilateral renal tumor mass lesion, in a young child, with heterogeneous enhancement after injection on CT scan.

In our case, we observed that on injected CT scans, the of metanephric stromal tumor showed gradual enhancement, more showen in the late stages (Fig. 2), which testifies to the fibrous nature of the lesion, related to its stromal component.

Mixed epithelial and stromal tumor is a benign tumor including cystic nephroma, combining a mesenchymal and an epithelial component. Malignant transformation is rare, only eleven cases of malignant Mixed epithelial and stromal tumor are reported in the literature.

On imaging, this tumor has the appearance of an intraparenchymal or intra-sinusal cystic mass, usually unilateral, forming a multilocular cyst of grade 3 in Bosniak's classification (Fig. 5).

Histological study shows fine septa with some residual renal parenchyma. In the macroscopic features, the cystic nephroma is entirely cystic without a solid nodule [6].

It combines 2 components: an epithelial component and a stromal component. The epithelial component consists of tubes of variable size, sometimes dilated, lined with cubic or cylindrical cells, with eosinophilic cytoplasm. The stromal contingent is made up of spindle-shaped cells of variable cell density (Fig. 6).

Malignant transformation occurs in the mesenchymal component such as; synovialosarcoma, rhabdomyosarcoma, chondrosarcoma, or undifferentiated sarcoma [7].

Congenital mesoblastic nephroma (CMN) was first described by Bolande et al. in 1967 [8]. Macroscopically, the tumor is firm, yellow and poorly limited. Necrosis and cystic areas may be present. Microscopically, the tumor is composed of fibroblasts and myofibroblasts, intertwined [9].

There are 3 types of mesoblastic nephroma:

- The typical form, which corresponds to the previous description;
- The cellular form, characterized by high cell density and mitotic activity;
- Mixed forms.

Cystic areas and necrosis most often correspond to cellular forms.

The classical type is the most benign and is usually treated by resection alone and has a good prognosis [10].

However, few studies have described the computed tomography (CT) and magnetic resonance imaging (MRI) manifestations of CMN [11,12].

Certain specific signs can be used to differentiate CMN from other renal tumors. For example, on enhanced CT, the

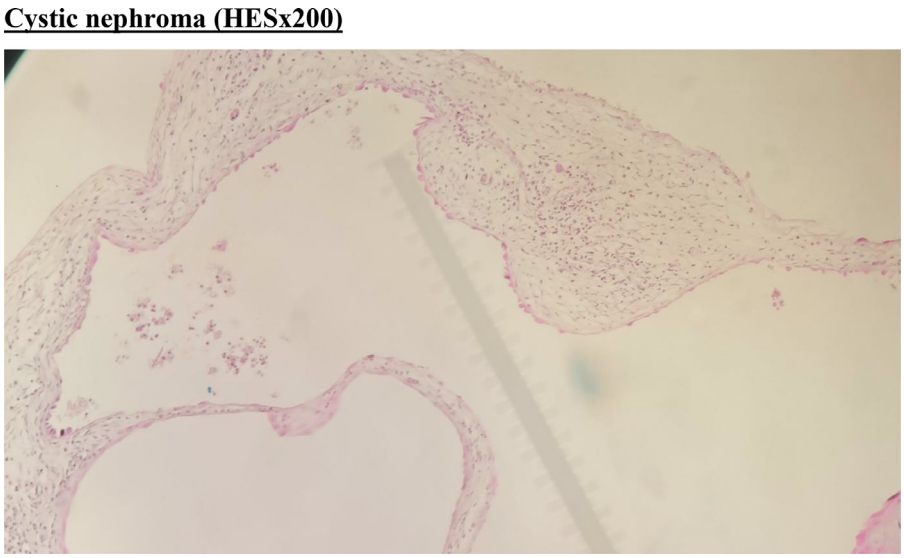

Fig. 6 – Multiple cystic formations bordered by simple flattened epithelium with a hobnail cells appearance.

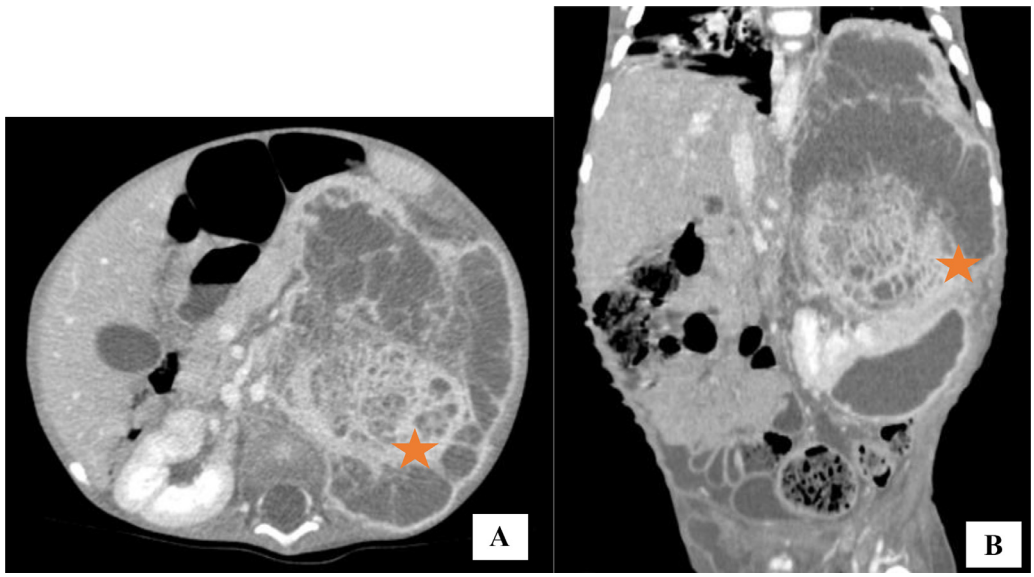

Fig. 7 – Axial (A) and coronal (B) CT sections scan of the abdominal region showing a left tumoral process, with two component, a predominantly cystic component and a solid component heterogeneously enhanced (arrow) shown in the cystic nephroblastoma differentiating it from cystic nephroma.

"double layer sign" at the edge of CMN and the "intratumor pelvic sign" of a non-encapsulated tumor are of high value for early diagnosis and differential diagnosis (Fig. 8), especially for the classic hystological subtype [6].

This sign is explained by a light to moderate non-uniform enhancement, relatively weaker than that of the renal cortex, in the corticomedullary phase of the CT scan, while the solid components of the tumors showed areas of liquefactive necrosis. In the nephrographic phase, these solid components were further enhanced, but the intensity remained lower than that of the renal parenchyma, probably due to renal dysfunction [13].

For the cellular histological subtype, the enhancement pattern is generally heterogeneous, and the existence of necrotic and hemorrhagic areas makes it difficult to differentiate from Wilm's tumor.

According to the literature, MRI manifestations of CMN vary greatly. CMN often displays hypointense signals on the T1WI sequence, relatively hyperintense, isointense, or lower-intensity signals than the peripheral renal parenchyma on fluid-sensitive sequences, such as T2WI, and heterogeneous enhancement on enhanced scan [14].

The main differential diagnosis for these tumors is Wilms' tumor. Age is an important discriminant parameter. Even if

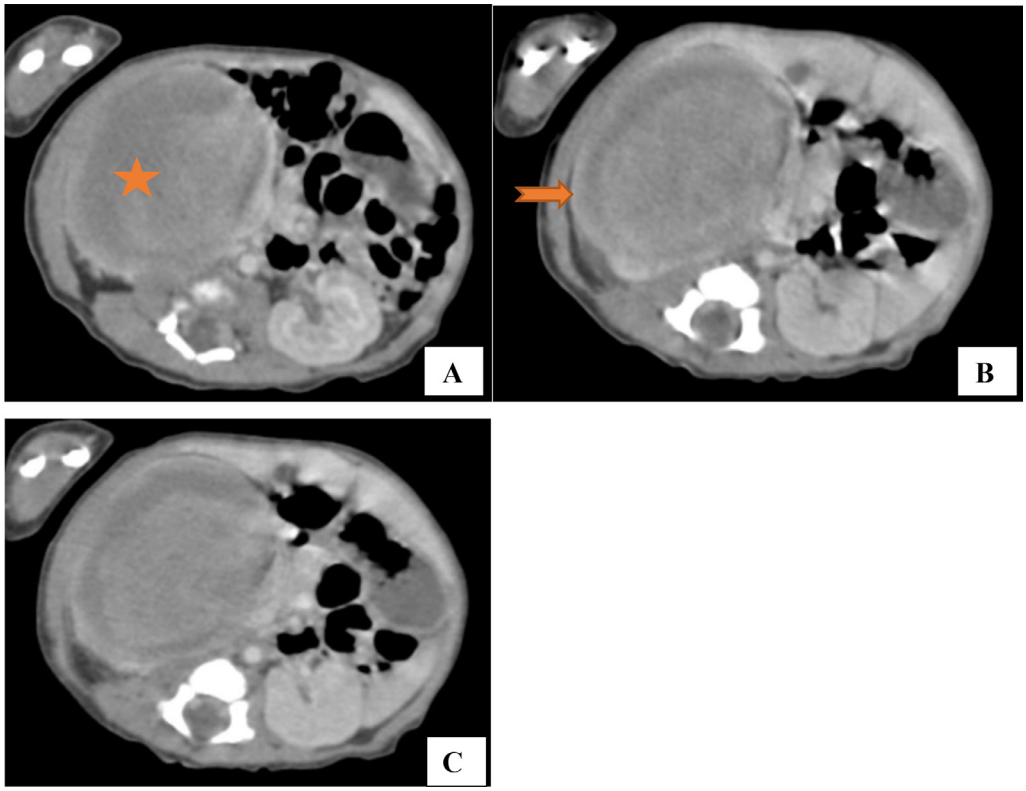

Fig. 8 – Enhanced axial CT of the abdomen, double-layer sign (arrow) **can be observed in the nephrographic phase, with the "intratumor pelvic sign"** (star).

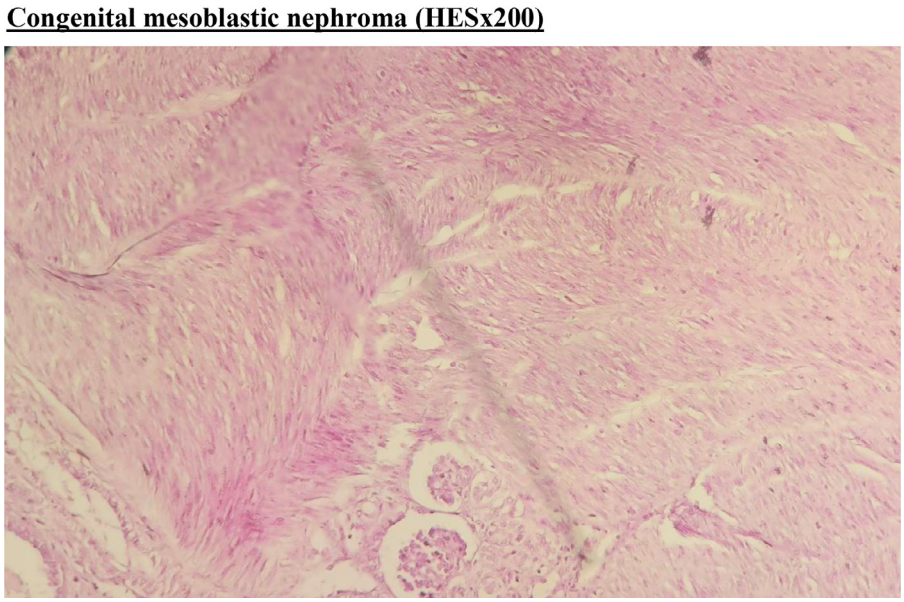

Fig. 9 – Renal parenchyma with a fusocellular proliferation of long intersecting bundles.

Wilms' tumor is the most common renal neoplasm in children, only 2% of cases occur before the age of 3 months [15].

In contrast to CMN and MST, Wilm's tumor has a pseudocapsule, and can invade the renal veins, inferior vena cava and even the right atrium, developing distant metastases (Figs. 4 and 7). These behaviors are rarely observed in metanephric tumors.

Clear cell sarcoma of the kidney is an aggressive solid cystic tumor that usually occurs between the ages of 2 and 3 years [16], is extremely rare before the age of 6 months and

very frequently presents with bone metastases. On imaging, it presents as round or oval masses in the renal medulla, of low or mixed density and usually greater than 10 cm in diameter; abundant cystic and necrotic components; scattered, linear, and punctate calcifications surrounding necrotic tissue; rich tumor blood supply and multiple fine tumor blood vessels; and high enhancement in solid areas, with cloudy, tabby, striped or scattered patterns [17].

Conclusion

Combining our experience of the imaging findings in our cases and the literature we have reviewed, we believe that the imaging manifestations of some benign tumors have certain morphological and dynamic characteristics.

The absence of a capsule or signs of invasion of neighboring vascular structures, particularly the renal veins and inferior vena cava, are some of the morphological features that should be taken into account when examining these tumors.

On the other hand, dynamic criteria such as the "double layer sign" surrounding the tumor are great diagnostic value in classical congenital mesoblastic nephroma, and the progressive enhancement seen in the late stages confirms the fibrous nature of the lesion, particularly in metanephric stromal tumors.

The incidence of benign renal tumors is minimal, but their importance lies in their extremely benign nature. The clinician must therefore be aware of their existence and take them into account in the differential diagnosis of childhood renal masses, so that neoadjuvant chemotherapy with its enfeebling side effects or a nephrectomy can be avoided.

Patient consent

Informed written consent was obtained from all patients for publication of the Case Report and all imaging studies.

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