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A Rare Renal Pediatric Tumor: Metanephric Stromal Tumor

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Abstract

A left abdominal mass revealed a metanephric stromal tumor in an 8-month-old infant. But before that, the ultrasound and CT scan of the mass suggested a nephroblastoma. This motivated neoadjuvant chemotherapy before total left nephrectomy. The sections of the Surgical specimen showed a whitish tumor of intramedullary location. On microscopic examination, the tumor corresponds to undifferentiated tumor proliferation. This imprecise diagnosis motivated an additional immunohistochemical study, which showed a CD34 positivity. The comparison of the histological aspects with the immunohistochemical aspects made it possible to confirm the diagnosis of a metanephric stromal tumor. The radiological check-up, carried out one year later, revealed no signs of recurrence. In this case, we would like to recall that, in addition to the impossibility of distinguishing radiologically, metanephric stromal tumor from clear cell renal sarcoma, imaging, not very helpful to the diagnostic approach of the first entity, could expose the patient to unnecessary neoadjuvant chemotherapy.

Keywords

Abdominal Mass, Infant, Metanephric Stromal Tumor, Nephroblastoma, Imagery Not Very Helpful, Unnecessary Neoadjuvant Chemotherapy

1. Introduction

Metanephric stromal tumor (MST) is one of the metanephric tumors [1]-[3]. These

tumors are distinguished by the overlap of their characteristic histopathological spectra and the presence of a BRAF mutation in most cases. MST is even more distinctive in his predilection for young children [1] [2]. It is a very rare benign renal tumor mainly observed in the pediatric population [1]-[10]. It represents a pure stromal variant of the metanephric adenofibroma and its first publication by Beckwith on a series of 31 cases dates back to 2000 [2] [3] [8]. Its diagnosis is difficult because of its rarity, the diversity and the complexity of its morphological presentation [4].

The interest of this entity resides in several orders of facts: first, the complete excision of the tumor is generally curative, then, its distinction from clear cell sarcoma will spare the child from adjuvant chemotherapy [6] [7] [9] [10].

In this paper, we report a new case of MST in an 8-month-old infant, and we describe the epidemiological, clinical, macroscopic and microscopic characteristics of this rare entity.

2. Case Presentation

Our patient is an 8-month-old male infant, born to an inbred marriage. He was admitted for abdominal pain with constipation. The onset of symptoms dates back to the 15th day of life.

The clinical examination found a left abdominal mass with lumbar contact. The rest of the clinical examination was normal. Ultrasound and CT scan of a nephroblastoma of the left kidney.

The assessment included the dosage of urinary catecholamines, which was negative, and the chest X-ray, which did not show pulmonary metastasis.

The patient received neoadjuvant chemotherapy followed by left nephrectomy.

Macroscopic examination of the specimen showed a mediorenal mass measuring 9 × 6.2 × 2 cm, well-circumscribed, whitish-colored and fleshy with gelatinous focus.

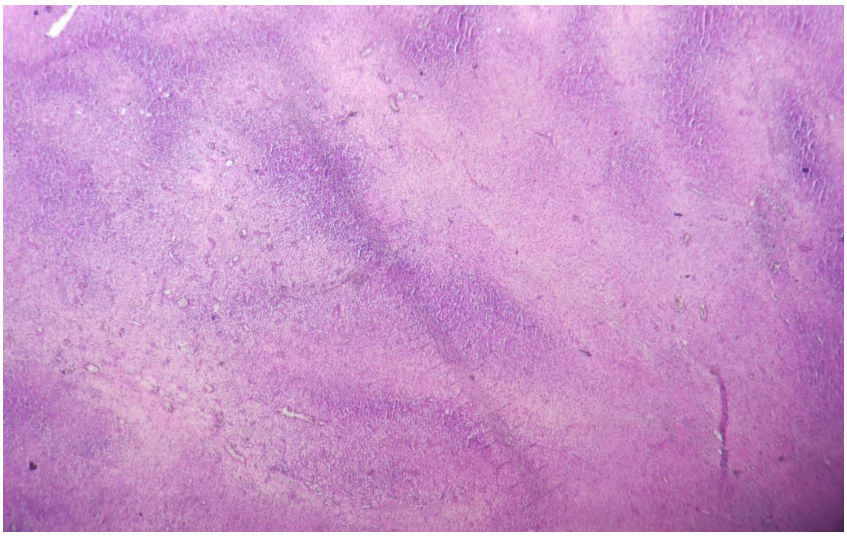

Figure 1. Tumeur stromale metanephric (HES ×100).

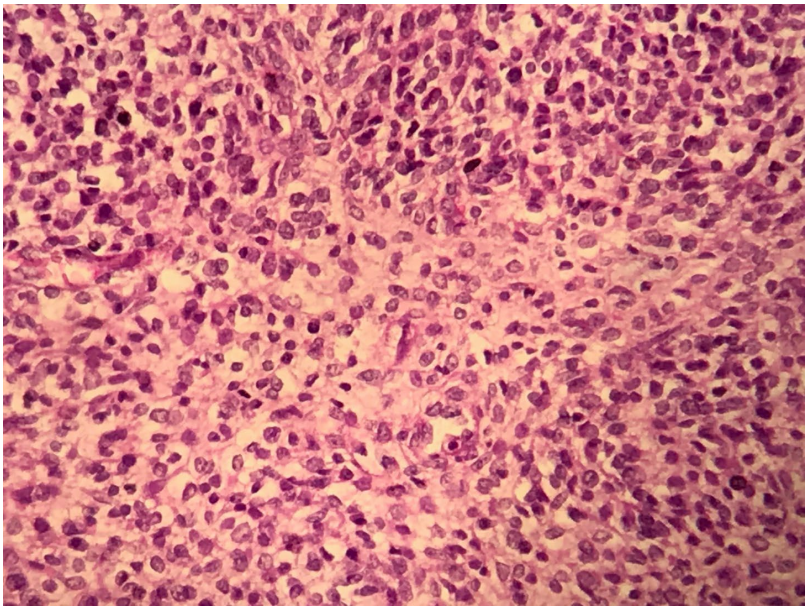

Figure 2. Tumeur stromale metanephric (CD34 ×200).

On microscopic examination, the tumor was non-encapsulated and corresponded to undifferentiated tumor proliferation (**Figure 1**).

This imprecise diagnosis motivated an additional immunohistochemical study, which showed a CD34 positivity (**Figure 2**). The comparison of the histological aspects with the immunohistochemical aspects made it possible to confirm the diagnosis of a metanephric stromal tumor. The radiological check-up, carried out one year later, revealed no signs of recurrence. In this case, we would like to recall that in addition to the impossibility of distinguishing radiologically, metanephric stromal tumor from clear cell renal sarcoma, imaging, not very helpful, to the diagnostic approach of the first entity, could expose the patient to unnecessary neoadjuvant chemotherapy.

3. Comment

MST is one of the new kidney tumors that was identified in 2000 by Argani and Beckwith [10]-[20]. It is a rare benign renal neoplasm, occurring mainly in small children, with an average age ranging from 13 to 24 months [8] [10]-[13] [19]. It has sometimes been described in adults [2] [11] [14] [19]. Of all the cases reported, there was no case of bilaterality [8] [12].

Its origin has not yet been clearly elucidated. However, some investigators have hypothesized that MST may arise from intralobular nephrogenic rests that have matured with loss of blastemal elements. In addition, the literature notes that the MST constitutes, with the metanephric adenoma (MA), a group of differentiated lesions which would be related to Wilms' tumor [8] [10] [12] [19].

Commonly, the tumor presents as an abdominal mass [12] [13] [15] [19] [20]. It can also be revealed by urinary symptoms or by hematological disorders (anemia) and hypertension. Some are found incidentally [11]-[13] [16] [19]. In our case, the

abdominal mass, located on the left, was associated with abdominal pain and constipation, with deterioration of health condition.

On imagery, the lesion can be solid, cystic or mixed [21]. This heterogeneity of appearance, associated with the rarity of this tumor, constitutes a source of difficulty in the interpretation of ultrasound [8] [9]. This often makes imagery less contributive [14] [16], as in our case, where ultrasound and CT scan suggested nephroblastoma. This presumed diagnosis motivated neoadjuvant treatment before performing a total left nephrectomy.

At macroscopy, the lesion is typically tan, lobulated, partially cystic and fibrous with a mean diameter of 5 cm. It is often centered in the medulla and is usually unifocal, but about one-sixth of cases are multifocal. It is unencapsulated and has a scalloped border that, on close inspection, subtly infiltrates the adjacent normal parenchyma [10] [16]. The present case confirms the medullary localization. The tumor is whitish in color, fleshy in appearance, and centered by a gelatinous focus.

Microscopically, examination at low magnification shows geographic differences in the degree of cellularity, producing a vaguely nodular variation in tumor cellularity. Tumor cells are spindle and stellate with thin hyperchromatic nuclei and indistinct cytoplasmic extensions [13] [19].

The tumor tends to surround native renal tubules and blood vessels, forming concentric "onion skin" rings or collarettes around these structures in a myxoid background [19]. Angiodysplasia of trapped arterioles may be evident in the form of epithelioid transformation of medial smooth muscle cells and myxoid change. Likewise, in the trapped glomeruli, hyperplasia of the juxtaglomerular cells can be observed. This phenomenon may be responsible for hypertension with hyperreninism. Heterologous stromal differentiation may exist in the form of a glial or a cartilagenous tissue [9] [14] [15] [17] [19].

In the diagnostic process of TSM, immunohistochemistry is an important contributor. Indeed, tumor cells of MST show positive immunostaining for vimentin. They demonstrate varying degrees of CD34 positivity: in fact, the more cellular, less myxoid spindle cell areas found at the periphery of the collarettes show strong CD34 positivity. Otherwise, tumor cells of MST show no immunostaining for cytokeratins, S100 protein, or Desmin. Areas with glial differentiation show positive immunostaining for S100 protein and glial fibrillary acidic protein [5] [9] [13] [15] [16] [19].

MST should be distinguished from the following two entities: First, the classic type of congenital mesoblastic nephroma (CMN). Indeed, the two have in common the localization in the renal medulla and the presence of bland spindle cells. However, MST is histologically distinguished by its scalloped border and its subtle infiltration, CMN being deeply infiltrating; its much less uniform cellularity, giving it at low power, a vaguely nodular appearance, unlike the marked linear variations observed in the cellularity of CMN; and the presence of perivascular cellular or hypodense onion-skin collars, angiodysplasia, juxtaglomerular cells hyperplasia and heterologous differentiation to glial tissue, characteristics absent in CMN. The two

entities may present a cartilaginous differentiation, but this is rare in the CMN. Immunohistochemically, MST stands out from CMN (Desmin positive) by its negativity to Desmin. Then, there was a spindled clear cell sarcoma (SCCS) of the kidney. MST is distinguished histologically by the absence of regular, branched capillary vascular pattern, which characterizes SCCS; the presence of angiodysplasia and heterologous stromal differentiation, which are not observed in the SCCS**.** Immunohistochemically, MST stands out from SCCS (CD34-) by its positivity to CD34 [5]-[10] [14] [15] [18] [19]. In practice, distinguishing TSM from SCCS will avoid unnecessary adjuvant treatment for a patient [9] [10] [16].

Until now, molecular genetic analysis of the STD has not revealed a characteristic anomaly. So currently, the diagnosis of MST is essentially based on the morphological and immunohistochemical examination [5]. However, since 2011, one case of complex chromosome rearrangement involving chromosome 17q has been reported [5] [10] [13]. Moreover, in 2016, to address the possibility that BRAF V600E mutations may underlie the pathogenesis of the MST, Argani's team identified 7 cases of pure MST, 1 case of stroma-predominant metanephric adenofibroma (MAF) and 1 case of MST that arose in a patient with neurofibromatosis type 1. The study showed BRAF V600E mutations in 6 of 7 cases of pure MST. The only case of MST that arose in a patient with type 1 neurofibromatosis was negative. Argani's team also found the identical BRAF V600E mutation in the stroma and epithelial components of MAF [21].

Tumor excision is an adequate therapy. Indeed, like all the cases of MST reported, the evolution of this one was benign, without metastases [8]-[10] [13]-[15]. However, recurrence or extra-renal angiodysplasia is possible [13].

4. Conclusions

We have just reported a case of MST of the left kidney in an 8-month-old infant. This case was revealed by a left abdominal mass, which was examined on imaging and suggested nephroblastoma of the left kidney. Standard histological analysis, supplemented by immunohistochemistry, made it possible to make the diagnosis.

It should be remembered that any abdominal mass, especially in small children, must motivate the systematic and meticulous search for the epithelial component and the blastematous component; and that, thinking about MST, would provide the advantage of avoiding unnecessary adjuvant therapy.

If necessary, immunohistochemical study by CD34 and Desmin will help to distinguish MST from other tumors, mainly congenital mesoblastic nephroma (benign tumor) and clear cell renal sarcoma (malignant tumor).

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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