—Case Reports—

Congenital Mesoblastic Nephroma Presenting with Hematuria in a Neonate

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Congenital mesoblastic nephroma (CMN) is a rare tumor of infancy. CMNs can be histologically divided into classic, cellular, and mixed subtypes. Cellular CMNs are difficult to differentiate from Wilms tumors. Herein, a neonate with cellular CMN accompanied by macroscopic hematuria, is described. The clinical, pathological, and imaging features of the disease are discussed.
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Key words: congenital mesoblastic nephroma (CMN), kidney, pediatric neoplasm

Introduction

Congenital mesoblastic nephroma (CMN) is a relatively rare pediatric renal tumor, accounting for 3%–10% of all pediatric renal tumors[1]. CMN has three histological subtypes: classic, cellular, and mixed subtypes. Herein, a case of cellular CMN in a five-month-old male neonate is presented.

Case report

A five-month-old male neonate suffered from macroscopic hematuria for four days. Ultrasonography showed a 6.0 cm×6.3 cm×4.4 cm, well-defined solid mass in the left kidney with partial cystic degeneration and bulky blood vessels (**Fig. 1a–b**).

An unenhanced CT scan showed a heterogeneous solid mass bulging out of the renal parenchyma (**Fig. 2a**). The mass showed obvious heterogeneous enhancement on the corticomedullary phase (with necrosis in the center and many vessels on the edge), continued enhancement on the nephrographic phase, and slight wash-out on the delayed phase (**Fig. 2b–d**). There was no suggestion of any renal vein defect or lymphadenopathy. On the basis of the radiographic findings, the mass was considered to be a Wilms tumor, and the neonate underwent a radical left-sided nephrectomy. Grossly, the mass was heterogeneous with necrotic and hemorrhagic tissues. Histological analysis demonstrated a highly cellular tumor, mainly consisting of spindle cells arranged in bundles. There were mildly pleomorphic tumor cells, with relatively low mitotic activity, suggesting that it was not aggressive. Smooth Muscle Actin (SMA) was focally expressed in the tumor cells, which were positive for Vimetin. On the basis of these findings, the patient was diagnosed with cellular congenital mesoblastic nephroma (**Fig. 3**). At a one-year follow-up examination, the patient was free of disease.

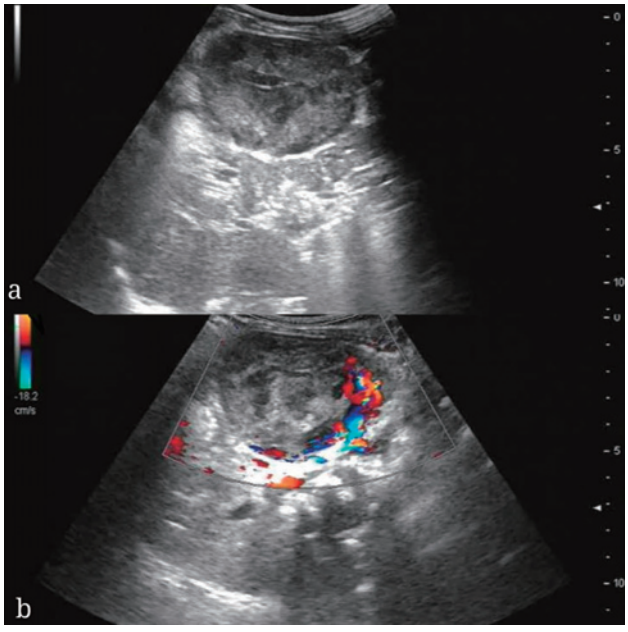

Fig. 1 (**a**, **b**) Ultrasonography detection showed a well-defined solid mass in the left kidney with partial cystic degeneration and bulky blood vessels.

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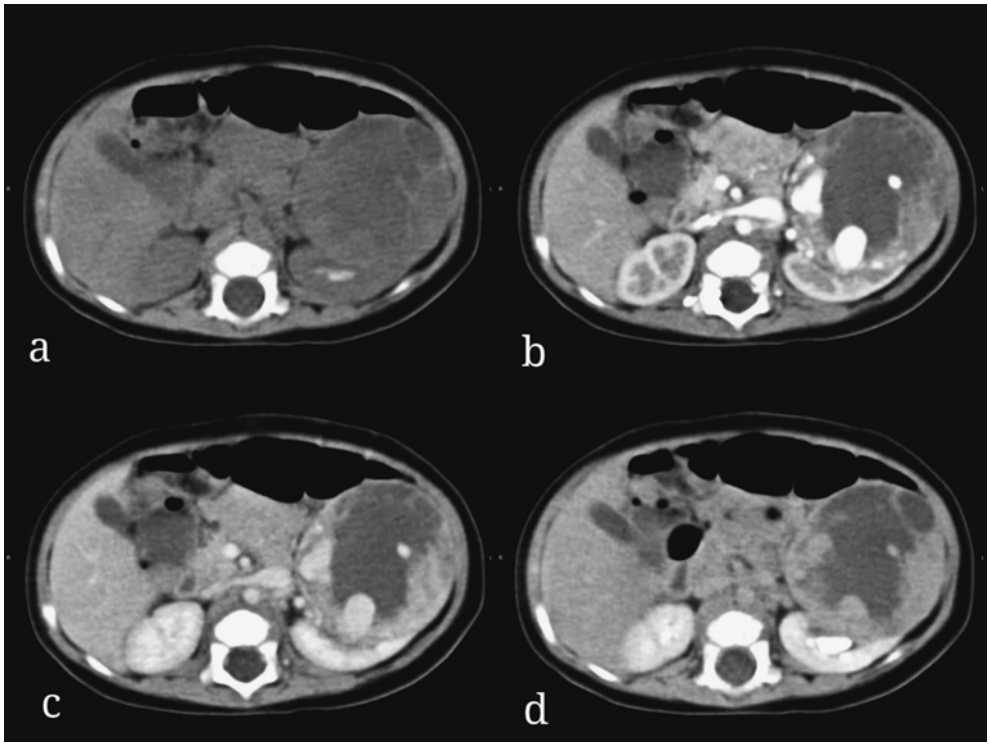

Fig. 2 An unenhanced CT scan showed a heterogeneous solid mass bulging out of the renal parenchyma (Fig. 2**a**). The mass showed obvious heterogeneous enhancement on the corticomedullary phase (with necrosis in the center and many vessels in the edge), continued enhancement on the nephrographic phase, and slight wash-out on the delayed phase (Fig. 2**b**, **c and d**).

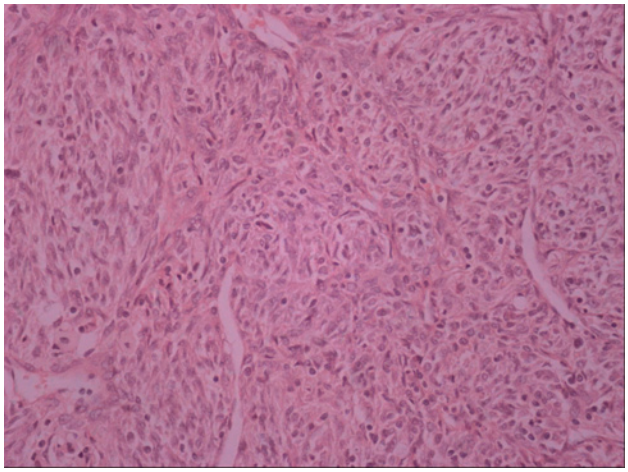

Fig. 3 Hematoxylin and eosin (H&E) staining (×400)

Discussion

CMN is a rare pediatric renal tumor. However, it is the most frequently diagnosed kidney neoplasm in newborn and young infants, accounting for 3%–10% of pediatric renal tumors and 18% of renal tumors within the first 7 months of life[2]. CMN was first reported by Bolande *et al.*[3] in 1967 as a benign renal stromal neoplasm of infancy. Later on, evidence of more invasive clinical behavior (i.e., recurrence, as well as metastasis in the lung and liver tissues) was described[4]. According to the cellularity and mitosis, CMN was divided into classic, cellular, and mixed subtypes[5].

Grossly, classic CMNs are solid tumors with a whorled, coarsely trabeculated cut surface. On histological examination, uniform, elongated, spindle-shaped cells arranged in bundles with entrapped glomeruli and tubules with a normal appearance were seen in classic CMNs. On the other hand, cellular CMNs are soft, fleshy masses, often with multicystic areas of gross hemorrhage or clear fluid accumulation. On histological examination, cellular CMNs are formed from spindle cells, haphazardly arranged in a sheet-like mode[6]. These tumors display nuclear atypia, a high mitotic rate, and an increased nuclear/cytoplasm ratio. Moreover, cellular CMNs are characterized by more invasive behavior, with intratumoral hemorrhage, necrosis, and cystic change, which may be related to the macroscopic hematuria that was seen in our case. The cellular lesions are prone to invade into the perinephric fat and connective tissues, rather than the renal pelvis and vascular pedicle. The risk of recurrence after excision for cellular CMN is always higher than in classic CMN.

These two main CMN subtypes differ in their imaging

features, depending on their pathological type and components. Classic CMNs are predominantly large, uniform solid masses, with minimal, largely peripheral enhancement. On the other hand, cellular CMNs are mainly cystic tumors with necrosis, cystic changes, hemorrhages, and heterogeneous enhancements, which are difficult to differentiate from Wilms tumors. Misdiagnosis of cellular CMNs might be associated with Wilms tumors, clear cell sarcomas, and kidney rhabdoid tumors. Wilms tumors are the most common kidney tumor of infancy, with their peak incidence at 3–4 years old, and less than 2% of cases occur in patients younger than 3 months. Appearance of vascular infiltration and pulmonary metastasis is much more suggestive of Wilms tumors[7]. Clear cell sarcomas and rhabdoid tumors are much more rarely seen, especially in infants.

Surgical nephrectomy has been considered as an adequate treatment for all CMN subtypes. Cellular CMNs usually recur and are therefore often treated with surgery and adjuvant chemotherapy. Overall prognosis for CMN is generally excellent. However, disease recurrence and metastasis are more common than originally thought, especially for cellular CMNs. Possible risk factors include incomplete resection, patient age, and cellular type[2,8]. For cellular CMNs, cases of translocation-positive tumors have significantly more satisfactory relapse-free survival[7]. Recurrence generally occurs in the first year, particularly for the cellular CMNs. Regular postoperative follow-up should be performed, especially within the first year after excision.

Whenever a well-defined renal mass is encountered, regardless of the appearance, and especially within the first year of life, the possibility of CMN should be considered.

Conflict of Interest: None declared.

References

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