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Unexplained Crying Episodes Reveling a Cystic Nephromas

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Abstract

Cystic nephromas (CNs) are uncommon, benign renal neoplasms reported in infants/young children of both genders and in adult females, concerning the pediatric population few studies have been conducted. The diagnosis of cystic nephroma is based on clinical signs, imaging tests, and anatomo-pathological study. In children, CNs can appear as a palpable abdominal mass in most of cases, hematuria or recurrent urinary infections. They are characterized by multycystic architecture and the exclusive presence of mature nephrogenic elements. Treatment is surgical with a very good prognosis in most of cases. We are going to report a case of a 13-month-old girl child diagnosed with Cystic nephromas who presented to pediatric emergency with unexplained crying episodes in order to increase clinicians awareness about this rare tumor.

Keywords

Cystic Nephroma, Benign Renal Tumor, Child, Good Prognosis

1. Introduction

Cystic nephroma is a rare, benign renal tumor of uncertain etiology [1], described for the first time in 1892 by Edmunds with very few cases described by authors especially concerning young children. The combination of clinical, biochemical, and histological characteristics associated with radiological features is essential for establishing the diagnosis. The final diagnosis is based on the histopathologic examination; the prognosis is well. We report a case of a 13-month-old girl child who presented with unexplained crying episodes diagnosed with CNs.

2. Observation

A 12 months' year old girl with a history of recurrent pyelonephritis and intermittent fever, there was admitted to the pediatric emergency for unexplained crying episodes evolving two months ago. The initial examination showed a conscious alert little girl, with high blood pressure (106/73), distended abdomen was noted on inspection with retroperitoneal mass measuring approximately 4 cm firm, lumbar contact was positive, the rest of the clinical exam was normal and the biological test showed a mild anemia and a high level of uremia (56 mg/dl). Ultrasound showed an oval mass of the right renal pelvis with a double cystic and echogenic tissue component measuring 95 × 55 × 46 mm. The investigation has been completed with an abdominal and pelvic CT ray which showed a huge mass of the right renal pelvis multilocular responsible of the rarefaction of renal parenchyma. The child has been benefited from a nephro ureterectomy of the right kidney, the macroscopic exam showed a total destruction of the renal parenchyma which was replaced by a lesion made of several cystic formations with irregular walls with no vegetations, the urethra was clean, no infiltration has been described, microscopic examination showed benign tumor proliferation made of several juxtaposed cystic cavities of different sizes, bordered by a cubic or flattened epithelium. The cells have a nucleus without cytonuclear atypia. The stroma separating these cavities is sparsely cellular, containing only lymphocytes and fibroblasts. For our patient treatment was based on total right nephro ureterectomy with good evolution thereafter (**Figure 1**).

3. Discussion

Childhood cystic nephroma is a rare, benign, renal tumor made of multilocular cysts without blastemal or undifferentiated elements [1] [2]. Pediatric cystic nephromas account for 2% to 3% of primary renal tumors [3]. It appears most commonly in younger children under 4 years old, most of whom are boys [4] [5]. Familial cases have been linked to DICER1 germline mutations and familial pleuropulmonary blastoma (PPB), whose main phenotypic spectrum includes PPB, PCN, ovarian Sertoli-Leydig tumors and multinodular goiter and less commonly, pineoblastoma, pituitary blastoma, nasal chondromesenchymal hamartoma and medulloepithelioma [6] [7] [8] [9].

Imaging tests combined with clinical history and physical examination are important for diagnostic suspicion and patient's follow-up. Pediatric cystic nephroma usually present as an asymptomatic palpable abdominal mass noticed by parents, with increasing abdominal girth. It may also be revealed by hematuria or possibly urinary tract infection. On CT/MRI, appears as a cystic, multilocular mass, most of time with pseudo capsule defined as a thin rim of tissue demarcating the margin of the lesion from the adjacent renal parenchyma; may about the renal pelvis or show protrusion/herniation into the renal pelvis [10].

The anatomopathological study allows a certain diagnosis of cystic nephroma. Macroscopically, a well circumscribed mass of cysts is observed, with a thick

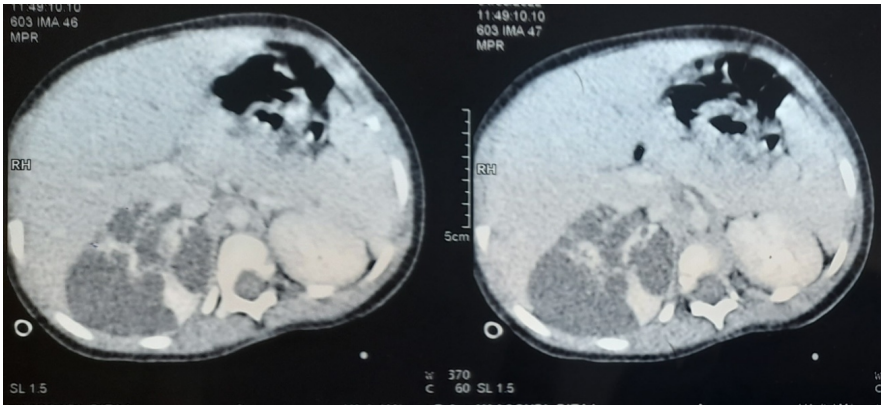

Figure 1. Cystic nephroma in our patient on the CT ray.

fibrous capsule, non-communicating fluid contents, separated by thin translucent septa ("honeycomb" aspect), where calcification, hemorrhage, and necrosis are unusual [11]. In microscopy, it has a flat epithelium, with eosinophilic cuboidal cells areas projected to the lumen as a "hobnail", containing mature tubules in its septa.

Differential diagnosis with other renal masses is challenging since such tumors present with different radiological characteristics and highly variable prognosis [12]. Although Joshi and Beckwith have described the criteria for the diagnosis of cystic nephroma, to date, there are no guidelines or consensus of this pathology [13]. In general, surgery based on nephrectomy that can be partial or total is choice number one because it allows a definitive diagnosis with anatomopathological analysis and it can be also a definitive therapy with an excellent prognosis. Partial nephrectomy is performed in small size tumors, usually by laparoscopic approach; however, when this is not possible, total nephrectomy is performed [14]. The differential diagnosis of other childhood neoplasms includes cystic partially differentiated nephroblastoma and solid tumors with cystic change such as nephroblastoma, clear cell sarcoma, congenital mesoblastic nephroma, and renal cell carcinoma. Occasionally, developmental disorder such as cystic renal dysplasia may also be mistaken as a cyst tumor.

4. Conclusion

Cystic nephroma is a rare benign tumor that affects children with an excellent prognosis. The association of clinical history, physical examination, imaging tests and histological characteristics is important to confirm the diagnostic.

Conflicts of Interest

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

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