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Multilocular Cystic Nephroma in a Child: A Rare Case

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Abstract Multilocular cystic nephroma (MLCN) is classically described as a benign slow-growing renal tumor. The aim of this study was to evaluate MLCN in a child in the West of Iran. The ultrasound examination showed a multicystic mass measuring 6.6x6.5 cm with multiple internal septae in the middle and lower left kidney in a 19-month-old boy. Another sonography revealed multicystic mass 7.4x6.6cm in left renal pole with a diagnosis of multicystic nephroma. The lab data were within normal limits, except for hemoglobin level, that was 11.6 g/dl. The gross examination revealed kidney tissue measuring 10x8x7 cm including multiloculated cyst measuring up to 8 cm and the microscopic examination revealed multiple cysts lined by flat epithelium with spindle cell stroma containing abortive tubular elements and rare foci of small round cells. In conclusion, MLCN occures more in male children and female adults. The results showed that low hemoglobin level had association with some cases of MLCN. Inflammation may be seen in MLCN, but presence of any amount of blastemal cells is diagnosed as cystic partially differentiated nephroblastoma by most experts.

Keywords: *Multilocular cystic nephroma, children, case report*

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1. Introduction

Cystic nephroma, also called multilocular cystic nephroma (MLCN), is a rare, non-genetic, benign, renal cystic lesion and presents as a unilateral, multicystic renal mass without solid nodules [1]. MLCN is classically described as a benign slow-growing renal tumor, and there are more than 200 cases reported in the literature [2]. The MLCN lesions typically have a bimodal age, with peak incidence in male children under 24 months and another one in women over 40 old [3]. Adult-onset cystic nephroma is histogenetically and morphologically different from pediatric cystic nephroma [4] and may include polycystic kidney, nephroblastomas, Wilms' tumor, hydronephrotic kidney, mesoblastic nephroma and cystic renal cell carcinoma [3]. The aim of this study was to assess MLCN in a child in the West of Iran.

2. Case Report

A 19-month-old boy referred to the radiologist on 31st July, 2016 for abdominal ultrasound evaluation with the chief complaint of nausea and vomiting. The ultrasound examination showed a multicystic mass measuring 6.6x6.5 cm with multiple internal septae in the middle and lower left kidney. The left renal normal contour was changed. The right and left renal dimensions were 7.6x3.6 cm and 6.2x2.4 cm, respectively. Another sonography on 8th August revealed multicystic mass 7.4x6.6 cm in left renal pole with a diagnosis of multicystic nephroma. Then, the computed tomography (CT) abdomen was done and showed a multilocular cystic lesion measuring 7.0x6.3 cm in left renal pole containing multiple thick septae. Weak enhancement after contrast injection proposed nephroblastic nephroma or Wilms'tumor with no peripheral extension as the radiologic diagnosis.

The patient referred to the urologist and admitted for surgical excision of mass on 27th August. The lab data were within normal limits, except hemoglobin level was 11.6 g/dl. Left radical nephrectomy was done on 28th August and the specimen was sent to the pathologist. The gross examination revealed kidney tissue measuring 10x8x7 cm and ureter measuring 6x0.4 cm. The cut sections revealed multiloculated cyst measuring up to 8 cm containing clear fluid with a smooth outer surface (Figure 1). The microscopic examination revealed multiple cysts lined by flat epithelium with spindle cell stroma containing abortive tubular elements and rare foci of small round cells (Figure 2 & Figure 3). The histologic diagnosis on 14th September showed MLCN with less than 5% of small round cell component and free surgical margins. Differentiation between chronic inflammatory cells (lymphocytes) or small foci of blastemal elements was not possible. Immunohistochemistry was not used for differentiation in the small non-expansile foci. Before receiving the final pathology report, the patient had received two doses of Actinomycin. He discharged in 30th September with good condition and was alive on 19th October, 2016.

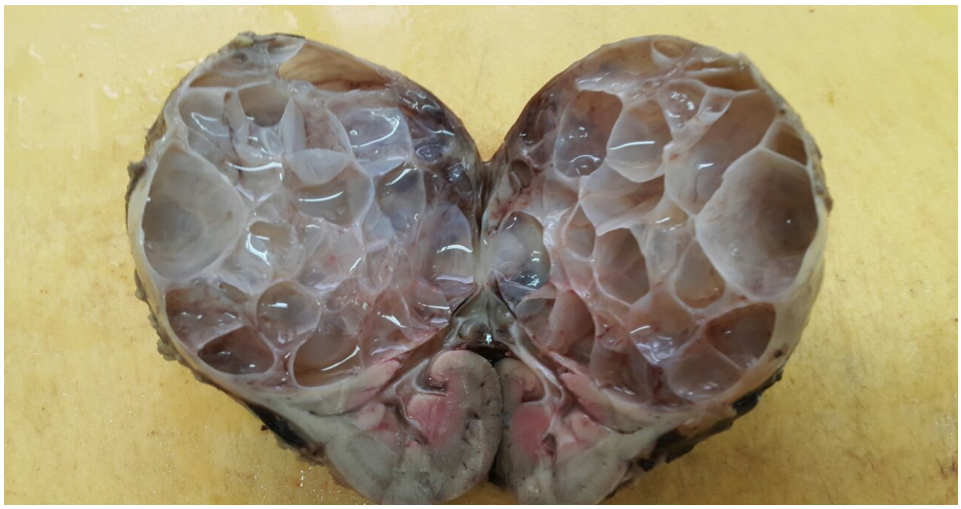

Figure 1. Gross specimen, multiloculated cyst with smooth outer surface

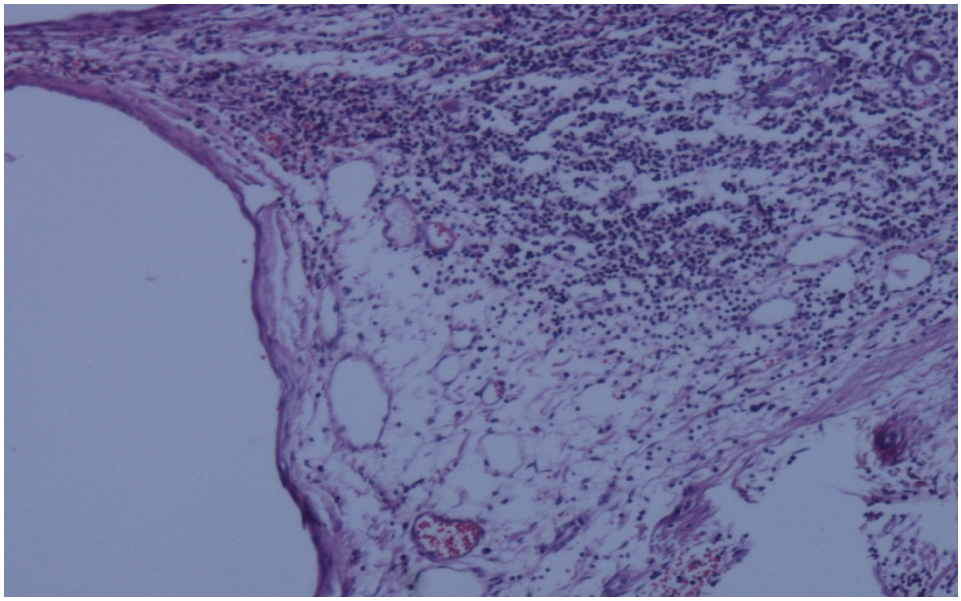

Figure 2. Histologic appearance: cyst with flat epithelial lining, stromal component and small round cells (Hematoxylin & Eosin staining,×100 magnification)

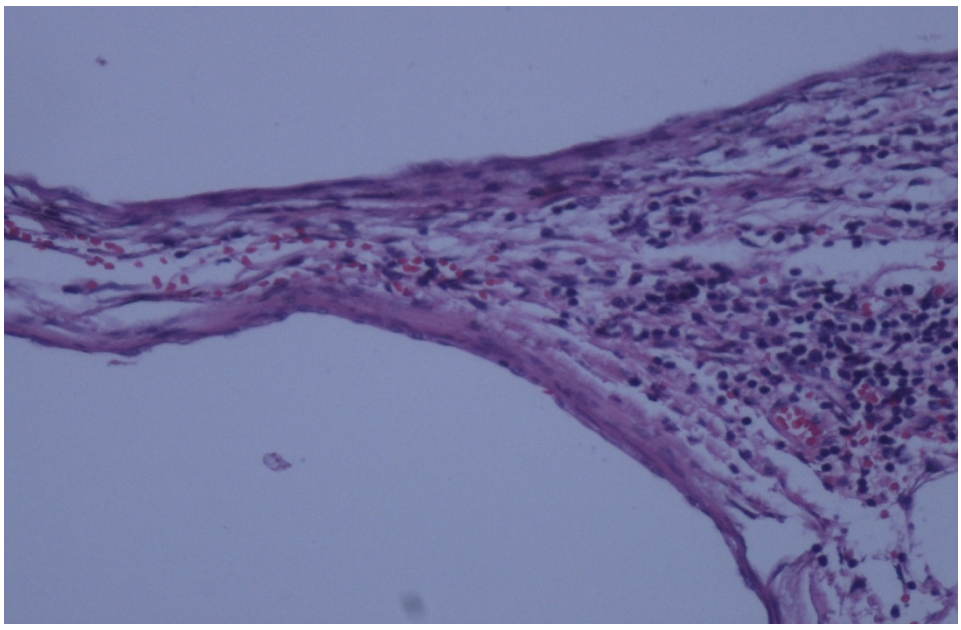

Figure 3. Histologic appearance (Hematoxylin & Eosin staining,×200 magnification)

3. Discussion

This case evaluated MLCN in a 19-month-old boy that had the chief complaint of nausea and vomiting that hemoglobin level was 11.6 g/dL. Wilkinson et al. [5] reported six cases with MLCN that the mean age of all patients was 52.3 years (range, 35-65 years) and male/female ratio was 1/5 (16.7% male). A systematic review in 2015 [2] showed that out of 179 cases with MLCN, 113 female patients (63%) and 66 male patients (37%). The median age was 32 years. Madewell et al. [6] evaluated 58 patients that age range was 3 months to 69 years old. Also, out of 25 male patients, 22 (88%) had age<24 months and out of 33 female patients, 21 (63.6%) had age>30 years. Children younger than 2 years old and adults in middle age (40–69 years old) are more commonly affected. Some case series describe a higher incidence in male children (male-to-female ratio, 3:1) [7]. Most cases in the literature were asymptomatic and discovered incidentally or presented with abdominal mass in children and non-specific urinary tract symptoms in the adults including hematuria [3]. Our case was presented with nausea and vomiting, which was unusual. González-Serrano et al. [8] reported a 73-year-old female that laboratory findings from routine blood tests and urinalysis were normal, whereas in a 18-month boy [9] hemoglobin level was 10.5 g/dL. Also, Doğan et al. [10] assessed a 16-month girl that hemoglobin level was 9.3 g/dl. Our presented case also had a rather low hemoglobin level of 11.6 g/dL. Madewell JE and the colleagues reported 58 patients that MLCNs were typically well circumscribed by a thick, fibrous capsule, and there was compression of adjacent renal parenchyma and the masses varied in size from 3x4cm to 17.5x33.5cm (mean: 8.7x9.7cm) [6]. About our case, the gross examination revealed kidney tissue measuring 10x8x7 cm containing multiloculated cyst up to 8 cm. The microscopic features consisted of cysts lined by cuboidal, hobnail or flattened epithelium [1,3,6,11] with eosinophilic cytoplasm [11], bland nuclear chromatin and were separated by distinct fibroblastic stroma [1,6]. It also had mature renal tubule in the fibrous interval [3]. The microscopic examination of our case revealed multiple cysts lined by flat epithelium with spindle cell stroma containing abortive tubular elements and rare foci of small round cells. These small round cells resembled lymphocytes, but with rare larger ones. Immunohistochemistry was not done for definite identification of the origin. They were less than 5% of the lesion. Inflammation is reported in cystic nephroma [1] that if this small cell component be considered as blastema, cystic partially differentiated nephroblastoma (CPDN) can be the diagnosis [10]. MLCN and stage 1 CPDN treatments are often surgery alone [12]. The number of these cases are too small for statistical analysis in the literature [13], but both of them showed 100% overall survival in the study of Luithle et al. [13]

4. Conclusions

MLCN occures more in male children and female adults. The results showed that low hemoglobin level had association with some cases of MLCN. Inflammation may be seen in MLCN, but presence of any amount of blastemal cells is diagnosed as cystic partially differentiated nephroblastoma by most experts. Clinicians should consider the diagnosis of multilocular cystic nephroma in the differential diagnosis of any cystic renal mass in children especially under 2 years old. Surgery must be done for definite histologic diagnosis.

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