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CASE REPORT

Peri-renal Macrocystic Lymphatic Malformation in a Neonate: A Case Report

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ABSTRACT

Perirenal lymphatic malformation (LM) is a rare benign lesion that often simulates as perinephric collection or parapelvic cyst. A neonate, antenatally diagnosed with a renal cystic mass, was investigated to have a perirenal LM on postnatal ultrasonography and MRI. The LM was excised successfully. Histopathologic examination of the specimen confirmed the diagnosis. Perirenal LM should be evoked for a renal cystic mass diagnosed prenatally. Knowledge of this entity can prevent an unnecessary nephrectomy.

Key words: Kidney; Lymphatic malformation; Cystic renal mass; Antenatal diagnosis

INTRODUCTION

LM is a benign lesion, seen in children, and mainly located in the neck and axillary regions.[1] It is classified as macrocystic, microcystic and mixed based on the size of the cysts. It rarely affects the kidneys. Its presence around kidney may pose a diagnostic and management challenge. We report a case of prenatally diagnosed renal cyst, which turned out as perirenal cystic LM.

CASE REPORT

A 36-year-old mother was referred at 34-week gestation for evaluation of fetal renal cyst. The prenatal ultrasound showed a unilateral cystic renal mass. After 5 weeks, a male baby was delivered (3kg). He was asymptomatic with Apgar score of 8 and 9 at 1 and 5min, respectively. The newborn had an overall good condition. The physical examination revealed a right flank mass. Biochemical investigations revealed normal renal and liver functions. Urine analysis was normal. The sonographic examination showed a retroperitoneal septated cyst measuring 8×5cm and pressing the right kidney and the liver. There was no blood flow on color Doppler ultrasonography. MRI showed a huge fluid-filled mass in the left pararenal area with enhanced internal septa and thin walls. The renal parenchyma was compressed. The perinephric collection appeared homogeneously hypointense on T1 weighted images and hyperintense on T2 weighted images. The MRI opinion was of perirenal cystic LM (Fig.1).

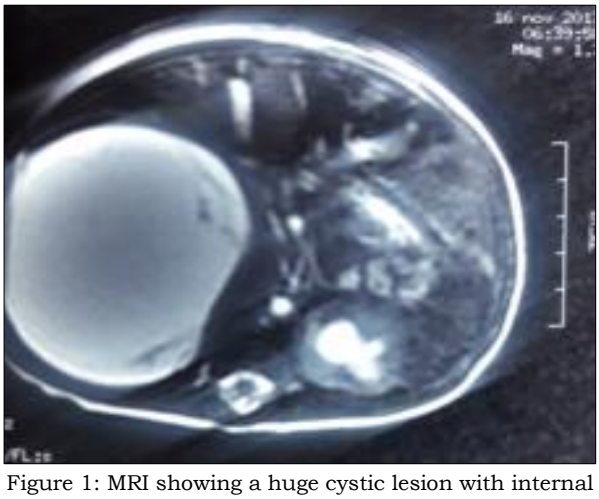

Figure 1: MRI showing a huge cystic lesion with internal septa located in the perirenal space with compressed renal parenchyma in front of the mass.

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After initial workup, surgery was performed using lumbotomy. The mass was located in the perirenal space. It was composed of voluminous cystic lesion filled with clear fluid (Fig.2 A). The renal parenchyma was compressed and laminated in front of the mass. The cyst was separated carefully from the renal parenchyma and excised totally (Fig.2B). The postoperative course was free of complications. The histopathologic examination found many cysts formed by endothelial cells with focal inflammation. No signs of malignancy were seen. The cells lining the cystic space stained positive for factor-VIII-related antigen and CD34 and negative for keratin. These findings confirmed the diagnosis of a cystic LM. No recurrence was noted on follow-up of 8 months.

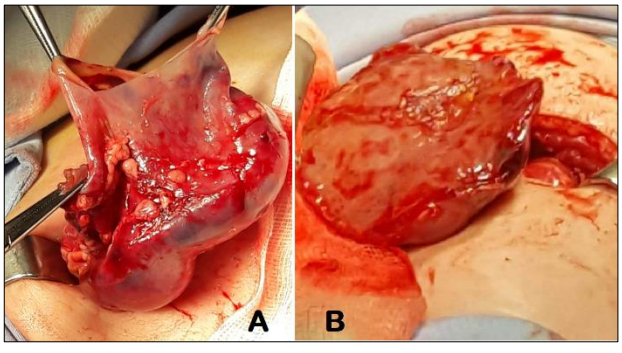

Figure 2: A) The cystic LM is being excised. B) After excision, the compressed renal parenchyma is quite obvious.

DISCUSSION

Perirenal cystic LM is a rare entity.[2,3] Much of the information about this disorder is based on isolated case reports. The entity is thought to be the result of the obstruction of the perirenal lymphatics. Thus, the lymphatic tissue around the kidney fails to establish a normal communication with the rest of the lymphatic system. The result is a dilatation of lymphatic ducts around the kidneys and the formation of unilocular or multilocular cystic mass. The other type of renal LMs are parapelvic or intrarenal and are instead thought to be caused by the obstruction of the renal pedicle lymphatics.[2,3]

Clinical manifestations are quite polymorphic. Most cases reported in the literature were asymptomatic and found incidentally. However, some cases presented with abdominal mass, abdominal pain, hypertension, hematuria, and proteinuria.[4] Antenatal diagnosis, as seen in our case, has not been reported previously. Perirenal LM are usually bilateral. A review of literature showed that only 4 out of 22 reported cases were unilateral.[5] Our case adds one more case to the unilateral category.

Perirenal LM appears as fluid density collections with or without septation enveloping the kidney.[5] Ultrasonography usually shows echoic perinephric collections with clear transmission and well-defined distal walls.[6] MRI showed typical unilateral perirenal cysts hypointense on T1 and hyperintense on T2 weighted images. The clinical history and normal renal parameters and C-reactive protein and imaging findings were suggestive of perirenal LM.

The treatment is mainly complete surgical excision as done in our patient. However, sclerotherapy with OK-432 or bleomycin is also gaining popularity as a primary treatment. Surgery is often required to avoid complications and for definitive diagnosis. Yet in order to reduce the risk of recurrence the resection should be as complete as possible. Laparoscopic excision of renal cyst lymphangioma, has also been described in the literature.[7] In our case, although the lymphangioma was huge and compressing the kidney, open surgical excision succeeded and the salvation of the kidney was possible.

Conclusion:

Perirenal LM should be evoked for a renal cystic mass diagnosed prenatally. The polymorphism of clinical and radiological aspects could make the diagnosis difficult in the preoperative period. Imaging findings, in this case, had an important role in suggesting the diagnosis. Histopathologic examination with immunochemical markers may confirm the diagnosis. Knowledge of this entity can prevent an unnecessary nephrectomy.

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an under-standing that every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

Author Contributions: All the authors contributed fully in concept, literature review, and drafting of the manuscript and approved the final version of this manuscript.

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