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Case Report

Bilateral peri-renal macrocystic lymphatic malformation in pediatric age-unusual presentation

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ABSTRACT

Lymphatic malformations are benign lesions, found in children, most common locations are neck and axillary regions. We report here a case of bilateral Peri-renal macrocystic lymphatic malformation in a 12-year-old female, masquerading as bilateral renal cysts. A 12-year-old female presented with complaints of pain in right flank for one week associated with fullness in right flank. On evaluation a provisional diagnosis of right perirenal lymphangioma was made and ultrasound guided pigtail drainage done. The pigtail output was 200-300ml daily for a week and it was removed and child was discharged. However, on follow up, CT was done and bilateral peri-renal lymphatic malformation(right>left) was diagnosed. Patient was then planned for exploratory laparotomy which revealed bilateral perirenal cyst, circumscribing both the kidneys completely. Excision of bilateral cysts was done and sent for histopathological evaluation. Cyst Fluid analysis revealed normal creatinine, LDH, total cholesterol, total protein, triglycerides, urea levels. Post-operative period was uneventful. Histopathological examination revealed cystic lesions formed by endothelial cells with focal inflammation, no signs of malignancy were seen. The child was followed up post-operatively and there was no evidence of recurrence. Perirenal lymphatic malformation is a rare entity. The entity is thought to be the result of the obstruction of the perirenal lymphatics. We are presenting this case for its rarity and unusual presentation. Macro cystic Lymphatic malformations are common diagnosis but around the kidneys, there are only very few cases reported in literature. Prognosis of cystic lymphatic malformation is excellent.

Keywords: Renal lymphangiectasis, Hygroma renale, Bilateral renal cysts, Paediatric lymphatic malformation of bilateral kidneys

INTRODUCTION

Lymphatic malformation is a benign lesion, found in children, most common locations are neck and axilla.[1] It is classified into macrocystic, micro cystic and mixed types, based on the size of cysts. It rarely affects kidneys.

Peri-renal lymphatic cysts, also termed hygroma renale or cystic lymphangiectasis, are rare. They must be differentiated from solitary, simple or multilocular cysts and cysts of pyelogenic or post-traumatic origin.[1] Its presence around kidney may pose a diagnostic and management challenge. Peri-renal lymphatic malformation is a rare disorder characterized by developmental malformation of the perirenal lymphatic system.[2] We report here a case of bilateral peri-renal macro cystic lymphatic malformation in a 12-year-old female, masquerading as bilateral renal cysts.

CASE REPORT

A 12-year-old female presented with complaints of pain in right flank for one week associated with fullness in right flank. There were no complaints of vomiting, fever or burning micturition. Biochemical investigations revealed normal renal and liver function parameters.

Urinalysis was normal. Ultrasonography revealed a cystic mass arising from right kidney. Further CECT abdomen and pelvis was done and a provisional diagnosis of right perirenal lymphangioma was made. Ultrasound guided pigtail drainage was done. Output was around of 200-300 ml from pigtail daily for one week, we were hesitant to try any sclerosant injection into cyst without being sure of the diagnosis, as perirenal lymphatic malformation is a rare diagnosis. Hence, we removed pigtail after a week and discharged the child and followed her up after 2 weeks. However, on follow up, this time ultrasound revealed bilateral cystic mass arising from kidneys. Further CT-scan was done which revealed similar findings as last time but this time even on left side, homogenously hypo dense collection, with 5.5 and 2.7 cm maximum thickness in right and left kidneys respectively, suggestive of bilateral perirenal lymphangioma (right>left), scans also suggested nephromegaly (right>left)-before pigtail drainage and during current scan too (Figure 1 and 2).

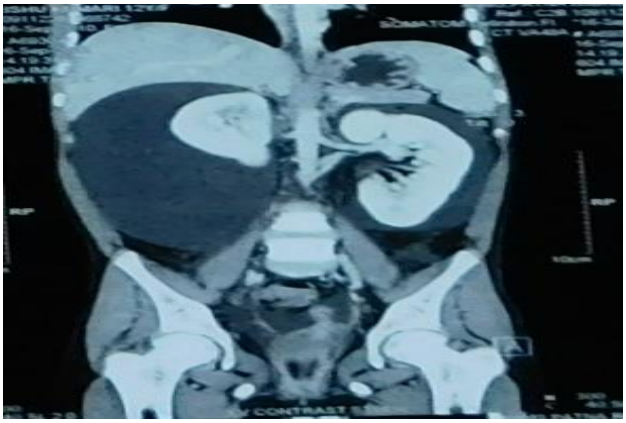

Figure 1: Coronal view of CECT abdomen and pelvis- of unilocular cysts enclosing bilateral kidneys.

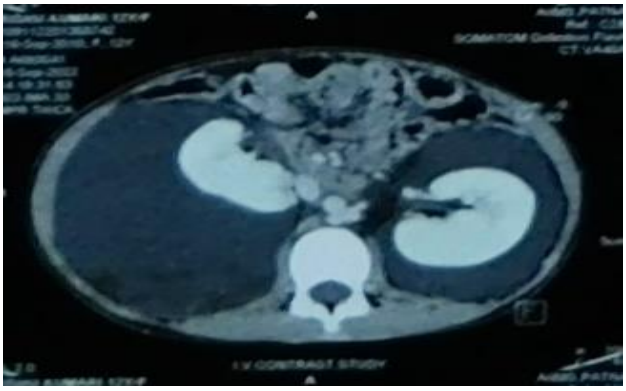

Figure 2: Axial view of CT abdomen of the bilateral peri-renal cysts.

Patient was then planned for exploratory laparotomy after proper preoperative preparation. Intra-operatively bilateral perirenal cyst were identified, circumscribing both the kidneys completely (Figure 3 and 4). Clear fluid was aspirated from cyst and sent for analysis. Excision of bilateral cysts was done and sent for histopathological evaluation. Fluid analysis revealed normal creatinine, LDH, total cholesterol, total protein, triglycerides, urea levels. Post-operative period was uneventful and child was discharged on postoperative day 5. Histopathological examination revealed Bilateral perirenal lymphatic cyst-formed by fibro collagenous cyst wall-lined by flattened epithelium. Sub epithelium shows moderate chronic inflammatory infiltrate along with lymphoid aggregates. No signs of malignancy were seen.

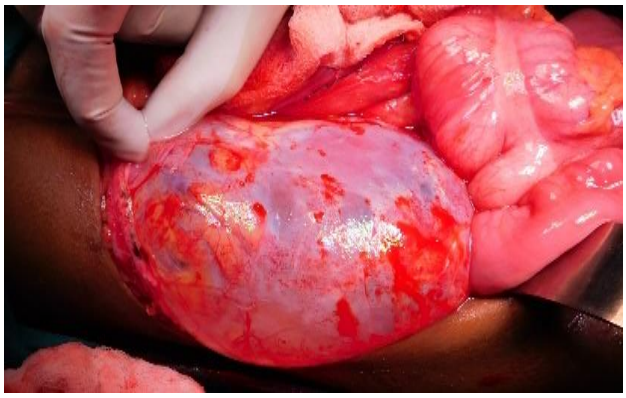

Figure 3: Intraoperative picture-revealing left side peri-renal cyst.

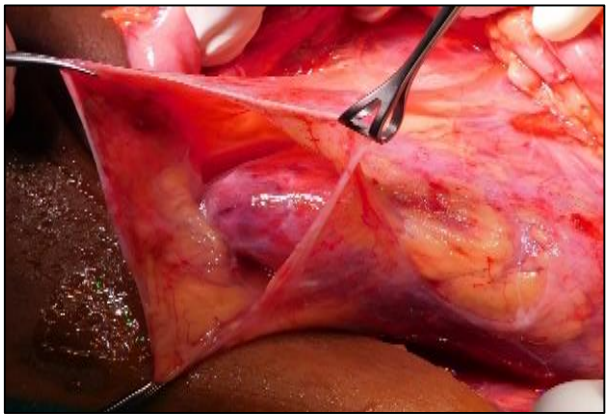

Figure 4: Intraoperative picture- revealing the cyst circumferentially encloses the kidney and kidney is peeping out of cyst after opening cyst.

The child was followed up post-operatively for a period of 12 months and there was no evidence of recurrence.

DISCUSSION

Perirenal lymphatic malformation is a rare entity.[3] 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. It results from dilatation of lymphatic ducts around the kidneys and the formation of unilocular or multilocular cystic mass. The other type of renal lymphatic malformations is Para pelvic or intra renal and is thought to be caused by the obstruction of the

renal pedicle lymphatics.[4] 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, haematuria, and proteinuria.[4] 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 bilateral category.

The diagnosis of renal lymphatic malformation is made by clinical history with typical imaging finding from ultrasonography and CT. The diagnosis cannot be confirmed by analysis of perinephric fluid. However, differentials like urinoma and abscess can be excluded thus giving a clue to the diagnosis. Renal lymphatic aspirates surprisingly not milky or chylous, as in our case. This is because lymphatic ducts of renal system are outside the mesenteric drainage pathway. It contains only sporadic cells, mostly lymphocytes and small amounts of fat and protein.[6] Imaging (ultrasonography and CT) plays an important role in the diagnosis of renal lymphangiectasis.[7]

There are various treatment modalities listed in literature. And since this a rare diagnosis. It is difficult to interpret the procedure or treatment of choice. Various modalities tried are percutaneous drainage-results in recurrence, injection of sclerosant, marsupialisation of cyst-so that fluid is absorbed into peritoneal cavity, excision of cysts and nephrectomy in decompensated cysts.[8-10]

In our case, the child presented with right sided pain abdomen and lump. On evaluation she was diagnosed as right perirenal lymphangioma with bilateral nephromegaly (right>left). The case was further discussed with radiologist and was planned for pigtail catheter insertion in view of unilocular cyst with clear fluid as content. But pigtail catheter insertion did not resolve the cyst. The fluid only kept forming again and again. Hence, we decided to remove the PCN and go ahead with a definitive procedure. We followed up the child and this time to see she had developed bilateral disease. Even left lesion was very much similar and smaller compared to right sided lesion. Intraoperatively it was interesting to see bilateral retroperitoneal cysts which had formed circumferentially enclosing both kidneys individually, which were unilocular and without any septations and contained clear fluid and kidney as a content of the cyst. We drained cyst contents and excised the cyst wall in-toto and sent for further analysis. We also observed enlarged kidneys after excision of cyst.

Child is monitored for 12 months and remains well, with resolution of symptoms and without any evidence of recurrence on examination and ultrasound. Hence stating excision of cyst is the definitive treatment of bilateral perirenal macro cystic lymphatic malformation with no risk of recurrence.

CONCLUSION

We are presenting this case for its rarity and unusual presentation. Macro cystic lymphatic malformations are common diagnosis but around the kidneys, there are only a very few cases reported in literature. The prognosis of the cystic lymphatic malformation is excellent.

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