Pediatric Nephrology (2019) 34:1543–1544
https://doi.org/10.1007/s00467-019-4198-1

CLINICAL QUIZ

A child with bilateral multiple renal cysts presenting with ascites and pleural effusion: Answers

Ayşe Ağbaş[1] · Bağdagül Aksu[1] · Güzide Doğan[2] · Türkan İkizceli[3] · Hatice Nilgün Duru Selçuk[4] · Murat Elevli[4]

Received: 6 January 2019 / Accepted: 9 January 2019 / Published online: 21 February 2019

Answers

1. What is your diagnosis?
   Ultrasound and abdominal MRI findings were consistent with renal lymphangiomatosis (RL).

2. Is further investigation or intervention required for the diagnosis?
   The diagnosis of RL can be made according to characteristic radiologic findings; further investigation is usually not required.

3 What is the treatment and prognosis?
   The diagnosis of RL can be made according to characteristic radiologic findings; further investigation is usually not required.

Discussion

We report here the unusual case of a young girl with renal cysts who presented with ascites and pleural effusion. She was evaluated by the Pediatric Gastroenterology Unit for the differential diagnosis of ascites having transudate features with few lymphocytes. She was then referred to the Pediatric Nephrology Unit for the investigation of multiple subcortical renal cysts with a differential diagnosis of autosomal recessive polycystic kidney disease. Following evaluation of the renal imaging studies, ultrasound and magnetic resonance imaging (MRI), and a review of the literature, Renal lymphangioma (RL) was diagnosed. The patient was put on a diuretic treatment (spironolactone), in accordance with reports in the literature of similar cases where both ascites and pleural effusion had improved with diuretic treatment. At the time of this publication, she had been on diuretic treatment for 12 months, with no reported side effects and normal renal function. Her renal cysts have regressed; the diameter of the largest cyst decreased from 45 to 35 mm. In addition, ultrasound examination revealed no ascites or pleural effusion. To the best of our knowledge, this is the youngest pediatric case reported that improved with medical treatment without any side effects.

Renal lymphangioma is a rare and benign renal malformation. It is thought that a developmental malformation of the intrarenal, perirenal, and peripelvic lymphatics causes accumulation of lymph as parenchymal edema, subcapsular and/or hilum cysts [1, 2]. This condition has also been termed renal lymphangioma, peripelvic lymphangiectasia, renal sinus polycystic disease, and renal hygroma [3]. The age of the patient at presentation varies and there is no sex predilection. A few familial cases have also been reported [4, 5]. Autosomal recessive polycystic kidney disease is the most common differential diagnosis in pediatric patients, especially when ascites is present. Other differential diagnoses include autosomal dominant polycystic kidney disease, Wilm's tumor, lymphoma, and urinoma [1].

Most cases of RL are asymptomatic and diagnosed incidentally. In symptomatic cases, the most common symptoms are abdominal or flank pain and abdominal distension. Weight loss, fatigue, palpable abdominal mass, hematuria, hypertension [6, 7], polycythemia [6, 8], and

This refers to the article that can be found at https://doi.org/10.1007/s00467-019-4196-3-x.

✉ Ayşe Ağbaş
yurtayse@hotmail.com

[1] Department of Pediatric Nephrology, Haseki Education and Research Hospital, University of Health Sciences, Istanbul 34140, Turkey

[2] Department of Pediatric Gastroenterology, Haseki Education and Research Hospital, University of Health Sciences, Istanbul, Turkey

[3] Department of Radiology, Haseki Education and Research Hospital, University of Health Sciences, Istanbul, Turkey

[4] Department of Pediatrics, Haseki Education and Research Hospital, University of Health Sciences, Istanbul, Turkey

renal vein thrombosis [9] have also been described. Complications of RL include infection, rupture, or hemorrhage of the cyst. Renin-dependent hypertension may be seen due to compression of the renal parenchyma and compromised renal perfusion as in Paget's kidney, which can be reversible or markedly improved following management of the cysts [6, 7]. Ascites and pleural effusion have also been documented [2, 8, 10, 11]. Renal function is usually preserved; however, renal failure has been reported in a few cases [12].

The diagnosis of RL can be made according to characteristic radiologic findings. The radiologic features of RL on unltrasound (US), MRI, or computed tomography (CT) are well defined [3, 13]. RL can be unilateral/focal or bilateral/diffuse. There are two radiologic manifestations [14]. In the first pattern, cystic lesions in the renal sinus may be seen. The second pattern reveals lobular perinephric fluid with multiple septations and less apparent renal sinus cysts. Ultrasonography examination may reveal enlarged kidneys, increased renal parenchymal echogenicity, loss of corticomedullary differentiation, and anechoic cysts. On CT; thin-walled, fluid-filled cysts with attenuation on the cyst walls are evident. The cysts on MRI appear as hypointense without enhancement on post-contrast images in T1-weighted and hyperintense in T2-weighted images. Diagnostic aspiration of the cysts or ascites can be performed but is not necessary for diagnosis. The characteristic of the aspirated fluid is usually the same as those for transudate (non-chylous and unremarkable for urea, protein, triglyceride, and lactate dehydrogenase (LDH)). The fluid may however have elevated renin and lymphocyte content [2, 7, 15].

Treatment is not required for the majority of asymptomatic cases. Diuretic treatment has proven effective in cases with ascites and pleural effusion [8, 10, 11]. Although recurrence is possible, percutaneous aspiration and sclerotherapy of the cysts have been successfully performed in symptomatic cases [16]. Marsupialization of the perirenal collection has also been reported [6, 7]. A partial or complete nephrectomy should be preserved for severe cases [7].

In summary, RL is a rare benign malformation. Awareness of this condition can prevent unnecessary investigations and interventions.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

References

1. Varela JR, Bargiela A, Requejo I, Fernandez R, Darriba M, Pombo F (1998) Bilateral renal lymphangiomatosis: US and CT findings. Eur Radiol 8:230–231. https://doi.org/10.1007/s003300050368
2. Jeon TG, Kong do H, Park HJ, Kim S, Park WY, Kim SD, Ha HK (2014) Perirenal lymphangiomatosis. World J Mens Health 32:116–119. https://doi.org/10.5534/wjmh.2014.32.2.116
3. Bagheri MH, Zare Z, Sefidbakht S, Nabavizadeh SA, Meshksar A, Roozbeh J, Salehipour M (2009) Bilateral renal lymphangiomatosis: sonographic findings. J Clin Ultrasound 37:115–118. https://doi.org/10.1002/jcu.20488
4. Gopaluni S, Gibson M, Mohteshamzadeh M (2014) An unusual case of familial cystic kidney disease. Clin Kidney J 7:484–485. https://doi.org/10.1093/ckj/sfu075
5. Sarikaya B, Akturk Y, Bekar U, Topaloglu S (2006) Bilateral renal lymphangiomatosis mimicking hydronephrosis: multidetector CT urographic findings. Abdom Imaging 31:732–734. https://doi.org/10.1007/s00261-005-8014-y
6. Blanc M, Schmutz G, Belzile F, Sabbagh R (2014) Renal lymphangiectasia presenting with hypertension and polycythemia. Can Urol Assoc J 8:E163–E166. https://doi.org/10.5489/cuaj.1596
7. Schwarz A, Lenz T, Klaen R, Offermann G, Fiedler U, Nussberger J (1993) Hygroma renale: pararenal lymphatic cysts associated with renin-dependent hypertension (Page kidney). Case report on bilateral cysts and successful therapy by marsupialization. J Urol 150:953–957. https://doi.org/10.1016/S0022-5347(17)35660-4
8. Nassiri AA, Lotfollahi L, Bakhshayeshkaram M, Kiani A, Haghighi S, Alavi Darazam I, Rashidfarokhi F (2015) Renal lymphangiectasia: a curious cause of pleural effusion. Tanaffos 14:213–216
9. Riehl J, Schmitt H, Schafer L, Schneider B, Sieberth HG (1997) Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrol Dial Transplant 12:1701–1703
10. Al-Dofri SA (2009) Renal lymphangiectasia presented by pleural effusion and ascites. J Radiol Case Rep 3:5–10. https://doi.org/10.3941/jrcr.v3i10.317
11. Arora R (2016) Bilateral renal lymphangiectasia-an unusual cause of abdominal distension and flank pain in a child. Quant Imaging Med Surg 6:466–469. https://doi.org/10.21037/gims.2016.07.04
12. Cadnapaphornchai MA, Ford DM, Tyson RW, Lum GM (2000) Cystic renal lymphangiectasia presenting as renal insufficiency in childhood. Pediatr Nephrol 15:129–131. https://doi.org/10.1007/s00460000413
13. Restrepo JM, Amaya JEL, Sepúlveda NA, Vélez MU, Massaro M (2011) Renal lymphangiectasia: MDCT and MRI findings. Rev Colomb Radiol 22:1–8
14. Gupta R, Sharma R, Gamanagatti S, Dogra PN, Kumar A (2007) Unilateral renal lymphangiectasia: imaging appearance on sonography, CT and MRI. Int Urol Nephrol 39:361–364. https://doi.org/10.1007/s11255-006-9039-z
15. Valerio M, Meuwly JY, Tawadros C, Jichlinski P (2012) Percutaneous drainage and sclerotherapy as definitive treatment of renal lymphangiomatosis. Can Urol Assoc J 6:E3–E7. https://doi.org/10.5489/cuaj.11034
16. Kashgari AA, Ozair N, Al Zahrani A, Al Otibi MO, Al Fakeeh K (2017) Renal lymphangiomatosis, a rare differential diagnosis for autosomal recessive polycystic kidney disease in pediatric patients. Radiol Case Rep 12:70–72. https://doi.org/10.1016/j.radcr.2016.11.016