Renal lymphangiectasia is a rare disorder where perirenal, parapelvic, and/or intra-renal lymphatics are dilated. The clinical presentation of renal lymphangiectasia can range from asymptomatic to renal failure. Ultrasound, computed tomography and magnetic resonance imaging have been used for the diagnosis. Management of such cases varies from conservative to nephrectomy. We report three cases of pediatric renal lymphangiectasia that were managed by recurrent sclerotherapies and medical supportive treatment at our institution. A literature review is also presented. To the best of our knowledge, there are 83 reported cases in the literature, 60 adults and 23 pediatrics.

Methods

Three cases of renal lymphangiectasia that were managed at our institution were reviewed, and demographic, clinical, and follow-up data were collected. The PubMed database was searched for the disease with all known nomenclature. Due to the rarity of the pathology in the pediatric population, we opted to include the adult population as well to compare both populations and thus expand the review of the literature. Any case younger than 21 years of age was considered pediatric. Out of 104 publications returned from the search, 83 articles were available for review with data related to presentation and treatment modalities. Descriptive data was generated. Continuous data was expressed as means and categorical data was reported as percentages. The Fisher exact test was used to calculate two-tailed $P$ values in the comparisons of categorical data.

Case 1

A 6-year old girl presented to a local hospital with abdominal distension and high blood pressure. Abdominal ultrasonography showed bilateral lymphangiectasia, also known as lymphangioma or lymphangiomatisis, is a miscommunication between the lymphatic vessels resulting in abnormal dilation of the vessels. Renal lymphangiectasia is a rare disorder that can affect the perirenal, peripelvic, and/or intrarenal lymphatic vessels. Renal lymphangiectasia is now the term most often used for the disease. To our knowledge, five cases of renal lymphangiectasia have been reported in Saudi Arabia and 104 worldwide. We report three cases at King Faisal Specialist Hospital and Research Centre managed for renal lymphangiectasia over the last 15 years, and present a review of the literature.
nephromegaly with cystic changes and ascites. The patient was referred to our hospital for further examination. Our work-up showed normal blood and urine parameters including urea and creatinine. A renal mercaptoacetyltriglycine scan showed right kidney split function at 68% and left kidney at 31%. A computed tomography (CT) scan of the abdomen showed large multiloculated cystic structures in both kidneys, mainly within the renal cortex, likely representing lymphangiectasia (Figure 1). No extra-renal extension was noted.

The patient was initially treated with a peritoneal catheter that drained over 1 liter of clear yellow fluid, resolving the ascites. The ascitic fluid results were WBC $38 \times 10^6$, lymphocytes 61%, glucose 6 mmol/L, lactic dehydrogenase (LDH) fluid:serum ratio=35/275=0.12, and total protein fluid:serum ratio=4/62=0.06. The high lymphocyte contents of the ascitic fluid were indicative of a lymphatic nature. Over a 2-year course of treatment, the patient was managed with a total of six sessions of sclerotherapy bilaterally targeting individual cysts sequentially. Sclerosing agents used were singular in four sessions, and in combination with other substances in two sessions: 98% alcohol 10 mL (two sessions), doxycycline 100 mg/10 mL (four sessions), lipiodol 3% 4 mL (two sessions). Sclerotherapy sessions were given based on follow-up imaging in intervals of a median of 3 months over 2 years. Laparoscopic exploration was performed with unroofing of the cyst and omental patch repair of the left kidney for the treatment of the recurrent large cysts. On a couple of occasions, a peritoneal drain was inserted for a few weeks to drain the ascites and was removed following patient stability. The patient went into remission with stable relatively small cysts with no ascites for 2 years. After that, she presented with ascites that was drained for 6 weeks and then resolved. Since then, she was stable with normal renal function and followed up with a nephrology team with stable bilateral renal lymphangiectasia (Figure 2). The patient was eventually lost to follow-up at age 18 years.

**CASE 2**

A 2-year-old boy, the brother of the girl in case 1, presented with right flank pain. The patient had a normal blood workup and blood pressure but ultrasound revealed cystic changes. Ascitic fluid revealed WBC $29 \times 10^6$, lymphocyte 72%, glucose 5.8 mmol/L, LDH fluid:serum ratio=22/288=0.08, and total protein fluid:serum ratio=5/68=0.07, indicating a transudate with a lymphatic nature. A diagnosis of unilateral extensive renal lymphangiectasia was made by CT scan. The patient was treated with right radical nephrectomy, and when last seen at our institution 5 years after surgery had normal renal function.

**CASE 3**

A 12-year-old boy was referred to our institution with left-sided pleural effusion and a provisional diagnosis of polycystic kidney disease. He complained of progressive abdominal distension and flank pain 3 months prior to his referral. On presentation, the patient had...
high blood pressure. Ultrasound and CT scan chest and abdomen showed bilateral renal lymphangiectasia with ascites and left pleural effusion. The pleural effusion was due to leak from the peritoneal cavity to the hemithorax (Figure 3). Left-sided thoracocentesis and paracentesis showed a clear yellowish fluid. Ascitic fluid revealed WBC 152×10^6, lymphocyte 82%, glucose 5.3 mmol/L, LDH fluid:serum ratio=16/322=0.05, and total protein fluid:serum ratio=6/79=0.08, indicating a transudate with a lymphatic nature.

The patient was then managed with four sclerotherapy sessions over a period of 3 years targeting bilateral renal cysts sequentially. Sclerosing agents used were singular (three sessions) and in combinations (one session): 98% alcohol 10 mL (three sessions), doxycycline 100 mg/10 mL (two sessions), bleomycin 15 mg (one session). Sclerotherapy sessions were given based on follow-up imaging on intervals of a median of 3 months over 2 years. An abdominal drain was left after sclerotherapy sessions on two occasions, 1 week for one and 2 weeks for the other, to drain the ascites. Due to recurrence of the left-sided pleural effusion associated respiratory distress, left thoracoscopic decortication was performed. Afterwards, the patient went into remission, and was kept on antihypertensive medications.

Six years after the diagnosis, the patient was diagnosed with chronic kidney disease stage G3b/A1 and was last followed up at the pediatric nephrology clinic. His bilateral renal lymphangiectasia was stable with no apparent leak (Figure 4).

**LITERATURE REVIEW**

A total of 83 cases were reviewed. There was no clear epidemiological data about the incidence of renal lymphangiectasia. The disease has been reported in children and adults with no gender predominance. Renal lymphangiectasia can occur in one kidney or both (Table 1). Some patients (16%) with renal lymphangiectasia were diagnosed incidentally. This may indicate an under-reported issue because it can be asymptomatic. The most common symptoms are abdominal pain, abdominal distension (ascites), and hypertension (Table 2). Ascites was the only difference in presentation that was statistically significant between the pediatric and adult age groups. In one case report, hypertension was caused by renal compression from the cystic collections, and drainage of the peri-pelvic fluid showed drastic improvement of blood pressure. In our cases, drainage of ascites and sclerotherapy of the cysts did not improve the hypertension. Management of renal lymphangiectasia can be expectant (follow up), medications such as antihypertensives, diuretics, and pain killers, drainage and sclerotherapy or surgical resection. Out of the 83

![Figure 3. An axial abdominal and chest CT with contrast cut showing massive left pleural effusion associated with mediastinal shift to the right side. Both kidneys are enlarged and widening of the renal hilum is noted. Both kidneys are surrounded by loculated-like cystic fluid collection causing a pressure effect mainly in the upper pole of the right kidney. Moderate ascites is noted (Case 3).](image1)

![Figure 4. An axial abdominal CT with contrast cut, after a sclerotherapy session over a year course, showing interval decrease in size and thickness of the right perinephric lymphangiectasia component with improvement of the mass effect. In addition, interval improvement of the abdominal ascites is noted (Case 3).](image2)
cases, regular follow up and total nephrectomy were the most common treatments (Table 3).

**DISCUSSION**

Renal lymphangiectasia is a rare and benign condition that accounts for 1% of all lymphangiomas. The pathophysiology of renal lymphangiectasia remains unclear. Familial predisposition has been reported in two cases.10,11 One article suggested that the pathophysiology of the disease occurs due to inflammation of renal lymphatic vessels, which leads to scar formation and obstruction of the efferent lymphatic vessels.12 On the other hand, in a case series, two patients developed severe rejection episodes and renal lymphangiectasia after renal transplantation from a pediatric donor. This was presumed to be an inherent result of interrupted development of the lymphatics.13 In our cases, there were no lymphatic abnormalities or extension to other organs elsewhere in the body. Associations of renal lymphangiectasia with renal vein thrombosis, chronic myeloid leukemia, polycythemia, and Gorham disease has been reported occasionally.6,14-20 Exacerbation of renal lymphangiectasia during pregnancy has been described in the literature.10,21

The disease has been reported in pediatric and adult patients. Our review showed that there were no differences in gender, clinical presentation or management options. The only difference was that more pediatric patients present with ascites than adults. The latter may reflect the severity of the disease, hence its early presentation with rupture of these cysts in the abdomen early in its course.

Imaging modalities such as sonography, contrast CT, and magnetic resonance imaging have been used in diagnosing renal lymphangiectasia. Detection of cystic lesions with otherwise normal renal parenchyma suggests the diagnosis. It can present as a solid mass with gradual enhancement on contrast enhanced CT. Cross-sectional imaging can show the presence or absence of ascites, pleural effusion, and help excluding cardiovascular, renal, and hepatic pathologies.22 Differential diagnosis of renal lymphangiectasia includes polycystic kidney disease, hydronephrosis, and cystic renal tumors.13 As the renal lymphangiectasia advances, a large isolated perinephric fluid collection can be seen and needs to be differentiated from hematomas, abscesses, urinomas, and nephrophathies that have been associated with floating kidneys.23,24 Management of asymptomatic patients is usually achieved through follow-up as the disease is benign in nature. Hypertension, ascites, and pain can be controlled with antihypertensives, diuretics, and pain killers, respectively.23,24 The location of the fluid collections guides the management approach. Paraneoplastic fluid aspiration and cyst decortication may minimize the compressive effects on the kidney and improve blood pressure. Nevertheless, they have been shown to be less effective in intrarenal lymphangiectasias.25 Percutaneous drainage can also be done to relieve compression related symptoms and is considered useful when the patient is not eligible for surgery.

| Table 1. Demographic data from the literature review. |
|-----------------------------------------------|
| **Category**                | **Pediatric (23 cases)** | **Adult (60 cases)** | **P value** |
|-----------------------------------------------|
| Median (range) age at presentation* (years) | 3.5 (0.1-20)             | 39.0 (22-87)        | -           |
| Gender (M:F)             | 15:8 (65%:35%)           | 29:31 (49%:51%)     | .22         |
| Unilateral/bilateral      | 11:12 (48%:52%)          | 25:33 (42%:58%)     | .8          |

*A newborn in one study was considered 1 month of age.

| Table 2. Presenting symptoms from the literature review. |
|-----------------------------------------------|
| **Symptom**                | **Pediatric (23 cases)** | **Adult (60 cases)** | **P value** |
|-----------------------------------------------|
| Ascites                     | 9 (39)                   | 8 (13)               | .015        |
| Hypertension                | 7 (30)                   | 13 (22)              | .4          |
| Abdominal/flank pain or discomfort | 7 (30)           | 29 (48)              | .21         |
| Fever                       | 3 (13)                   | 4 (7)                | .39         |
| Pleural effusion            | 3 (13)                   | 3 (5)                | .34         |
| Hematuria                   | 2 (8)                    | 10 (17)              | .5          |
| Asymptomatic                | 2 (8)                    | 12 (20)              | .33         |
| Renal impairment            | 2 (8)                    | 6 (10)               | .99         |

*Data are n (%).

| Table 3. Management modalities from the literature review. |
|-----------------------------------------------|
| **Management**                | **Pediatric (23 cases)** | **Adult (60 cases)** | **P value (<.05)** |
|-----------------------------------------------|
| Total nephrectomy                  | 6 (26)                   | 15 (24)              | .99         |
| Partial nephrectomy                | 2 (8)                    | 4 (7)                | .67         |
| Percutaneous drainage              | 5 (22)                   | 13 (21)              | .99         |
| Percutaneous drainage with sclerotherapy | 2 (8)               | 3 (4)                | .61         |
| Marsupialization                   | 0 (0)                    | 2 (3)                | .99         |
| Medications                        | 5 (22)                   | 12 (20)              | .99         |
| Follow-up                          | 5 (22)                   | 15 (24)              | .99         |

*Data are n (%).
However, the procedure yields a temporary effect as recurrence is a common event.\textsuperscript{21}

Percutaneous drainage with sclerotherapy has been shown to be a safe and efficient technique for the treatment of renal lymphangiectasia.\textsuperscript{26,27} From our experience, it needs many sessions of sclerotherapy because of the complexity of the disease and high rate of recurrence. Sclerotherapy is contraindicated in parapelvic cysts and those connected to the collecting system as there is a risk of sclerosis and obstruction of the collecting system by leakage of the sclerosing agent from the cysts to the collecting system.\textsuperscript{28} Percutaneous drainage is the treatment of choice for postoperative lymphoceles.\textsuperscript{29} However, management of renal lymphangiectasia with only percutaneous drainage can cause a high relapse rate in especially larger or multiseptated lesions. In reported cases using percutaneous drainage with sclerotherapy, doxycycline was used in one pediatric patient and showed no fluid re-accumulation with normal renal function at follow-up.\textsuperscript{30} In another study using povidone-iodine sclerotherapy approach in two adult patients, the patient underwent povidone-iodine washing for 18 days with resolution of symptoms and the patient was relapse-free at follow-up.\textsuperscript{26}

In conditions where recurrent collections and leakage in the retroperitoneum occur, cyst marsupialization into the peritoneum was done. Marsupialization was also used in conditions where the cyst was peripherally located.\textsuperscript{31} This intervention, however, is not always favored due to the increased tendency of intraoperative bleeding and nephrectomy. Nephrectomy is spared for conditions like uncontrolled intraoperative bleeding and recurrent complicated symptoms. Unfortunately, renal lymphangiectasia is sometimes treated with total nephrectomy with a presumptive diagnosis of a malignant lesion.\textsuperscript{8,32-35} We believe that nephrectomy should be used as a last option after failure of sclerotherapy and for unilateral disease. Moreover, nephrectomy may be considered for uncontrolled complications such as infection, bleeding, and suspicion of neoplastic changes. Out of the reported management modalities, there is no data to support the ultimate preference or success of one modality over another as the case reports are singular or in small numbers, which precludes the ability to draw any conclusions regarding effectiveness.

In conclusion, renal lymphangiectasia is a very rare disease. Its management is very challenging especially if it is bilateral. We believe that expectant management for asymptomatic patients with close monitoring of renal function and blood pressure is appropriate. Symptomatic patients should be managed conservatively with repeated drainage and sclerotherapy sessions while reserving nephrectomy to complicated cases or failure of sclerotherapy, especially in the presence of unilateral disease. Renal function needs to be monitored closely for a long time, especially in bilateral cases.
REFERENCES

1. Elbanna KY, Almutairi BM, Zidan AT. Bilateral renal lymphangiectasia: radiological findings by ultrasound, computed tomography, and magnetic resonance imaging. J Clin Imaging Sci. 2015;5:26. Published 2015 Jan 30. doi: 10.4103/2156-7514.150449

2. Llorente JG, García AD, Sacristán JS, Chicharro GN. Renal lymphangiectasia: radiologic diagnosis and evolution. Abdom Imaging. 2002;27(6):637-639. doi: 10.1007/s00261-001-0147-z

3. Ramseyer LT. Case 34: renal lymphangiectasia. Radiology. 2001;219(2):442-444. doi:10.1148/radiology.219.2.01ma17442

4. Karkouche R, Rocher L, Guettier C, et al. Bilateral renal lymphangiomatosis: imaging and histopathologic findings. Abdom Imaging. 2013;38(4):858-862. doi: 10.1007/s00261-012-9977-0

5. Jorge MR, Juan LA, Natalia AS, Melissa UV, Maurício M. Renal Lymphangiectasia. MDCT and MRI findings. Rev Colomb Radiol. 2011;23(3):1-8

6. Leite AF, Venturieri B, de Araújo RG, Silva EJ, Elias Junior J. Renal lymphangiectasia: know it in order to diagnose it. Radiol Bras. 2011;22(3):1-8.

7. Pandya VK, Shah MK, Gandhi SP, Patel HV. Bilateral Renal Lymphangiectasia. J Clin Diagn Res. 2016;10(9):TD01-TD02.

8. Blanc M, Schmutz G, Belzile F, Sabbagh R. Renal lymphangiectasia presenting with hypertension and polycythemia. Can Urol Assoc J. 2014;8(3-4):E163-E166. doi:10.5489/cjua.1596

9. Hauser H, Mischinger HJ, Beham A, et al. Cystic retroperitoneal lymphangiomas in adults. Eur J Surg Oncol. 1997;23(6):322-326. doi:10.1016/s0748-7983(97)90777-0

10. Meredith WT, Levine E, Ahlstrom NG, Grantham JJ. Exacerbation of familial renal hypertension and polycythemia. Can Urol Assoc J. 2016;10(9):TD01-TD02.

11. Kim JK, Ahn HJ, Kim KR, Cho KS. Renal lymphangiectasia in the Transplanted Kidney. Case Series and Literature Review. Transplantation. 2020;104(1):172-175. doi: 10.1097/TP.0000000000002745

12. Rietj H, Schmitt H, Schäfer L, Schnei- der B, Sieberth HG. Retropertoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrol Dial Transplant. 1997;12(8):1701-1703. doi:10.1093/ndt/12.8.1701

13. Rastogi R, Rastogi UC, Sarikwal A, Rastogi V. Renal lymphangiectasia associated with chronic myeloid leukemia. Saudi J Kidney Dis Transpl. 2010;21(4):724-727.

14. Renacci RM, Bartolotta RJ. Gorham disease: lymphangiomatosis with massive osteolysis. Clin Imaging. 2017;41:83-85. doi:10.1016/j.clinimag.2016.10.007

15. Burton IE, Sambrook P, McWilliam LJ. Secondary polyphagocytosis associated with bilateral renal lymphocelees. Postgrad Med J. 1994;70(825):515-517. doi:10.1136/pgmj.70.825.515

16. Vignetti D, Sverzut JM, Peraldi MN. Perirenal fluid collections and monoclonal gammopathy. Nephrol Dial Transplant. 2012;27(1):448-449. doi:10.1093/ndt/gfr433

17. Shaheen M, Hilgarter KA, Hawes D, Badve S, Antony AC. A Mexican man with “too much blood”. Lancet. 2003;362(9386):806. doi:10.1016/S0140-6736(03)14291-2

18. Bazari H, Attar EC, Dahi DL, Uspot RN, Colvin RB. Case records of the Massachusetts General Hospital. Case 3-2010. 2010;25:366-370. doi:10.1093/nejmsat/s003-000550

19. Ozturk M, Deren O, Akata D, Ahkan O, Ozem H, Durukan T. Renal lymphangiectasia during pregnancy: management with percutaneous drainage. Eur Radiol. 2001;11(1):37-40. doi:10.1007/s00330-000-0071-0

20. Kim JK, Ahn HJ, Kim KR, Cho KS. Renal lymphangioma manifested as a solid mass on ultrasonography and computed tomography. J Ultrasound Med. 2002;21(2):203-206. doi:10.7863/jum.2002.21.2.203

21. Asfah K, Raza SS, Ashraf O, Memon W, Bano S, Yadav SN, Chaturvedi S, Garga A. Lymphatic disorders after renal transplantation: imaging appearance on sonography, CT and MRI. Int Urol Nephrol. 2002;34(8):669-670. doi:10.1023/A:1020569707912

22. Ashraf K, Raza SS, Ashraf O, Memon W, Alogaili R, et al. Renal lymphangiectasia: imaging appearance on sonography, CT and MRI. Int Urol Nephrol. 2002;34(8):669-670. doi:10.1023/A:1020569707912

23. Chen Z, Qi L, Zhang Z, Hu Z, Fan B. Renal lymphangiomatosis: a rare differential diagnosis for autosomal recessive polycystic kidney disease in pediatric patients. Radiol Case Rep. 2016;12(1):70-72. Published 2016 Dec 23. doi:10.1016/j.racr.2016.11.016

24. Wadhwa P, Kumar A, Sharma S, Dogra PN, Hemal AK. Renal lymphangiomatosis: imaging and management of a rare renal anomaly. Int Urol Nephrol. 2017;49(1):365-368. doi:10.1007/s10045-016-0392-0

25. Murray KK, McLellan GL. Renal peripelvic lymphangiectasia: appearance at CT. Radiol. 1997;180(2):455-456. doi:11.1148/radiol.1802.2068311

26. Ratti M, Ammar L, Zennaro F, et al. Renal lymphangiectasia. Pediatr Radiol. 2004;34(8):669-670. doi:10.1007/s00247-004-0115-0

27. Simonot SC, Saltzman DA, Brennon W, et al. Cystic renal lymphangiectasia: a distinctive clinicopathologic entity in the pediatric age group. Pediatr Pathol Lab Med. 1997;17(2):293-301.