Letter to the Editor

Renal lymphangiomatosis: literature analysis on research progress and presentation of four cases

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Introduction

Renal lymphangioma is a rare disease (1-3), being more of a malformation rather than a true tumor. It is caused by an inability of the renal lymphatic system to communicate with the normal lymphatic system, resulting in the lymphatic dilation of the kidney, renal capsule, and perirenal area and the formation a single or multilocular cystic mass. Due to the relative rarity of the disease, the current disease understanding is based on a few case reports and multiple case series reports. The misdiagnosis rate of renal lymphangioma is high, as it is easily confused with other renal cystic diseases (e.g., hydronephrosis, polycystic kidney disease, and parapelvic cyst) (4-6). Imaging plays an important role in its diagnosis and differential diagnosis. Although renal lymphangioma is itself benign, studies (7-11) have found that it may cause more serious complications (e.g., hypertension, renal vein thrombosis, renal insufficiency, glomerulonephritis, and ascites). The natural history of renal lymphangioma is largely unknown. Only reports by Meredith *et al.* (12) and Antonopoulos *et al.* (13) and our first report of 3 related cases (2 sisters and their father) support the hypothesis of family disease heredity. Thus, to further inform clinical diagnosis and treatment, we selected 4 case reports, conducted a retrospective analysis on published Chinese and international articles on this rare disease; described the clinical and laboratory characteristics, imaging manifestations, treatment methods, and other factors of renal lymphangioma; and classified them accordingly.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). This retrospective study was approved by the Institutional Ethics Committee of Women's Hospital School of Medicine Zhejiang University (approval number: IRB-20220046-R). Written informed consent was provided by the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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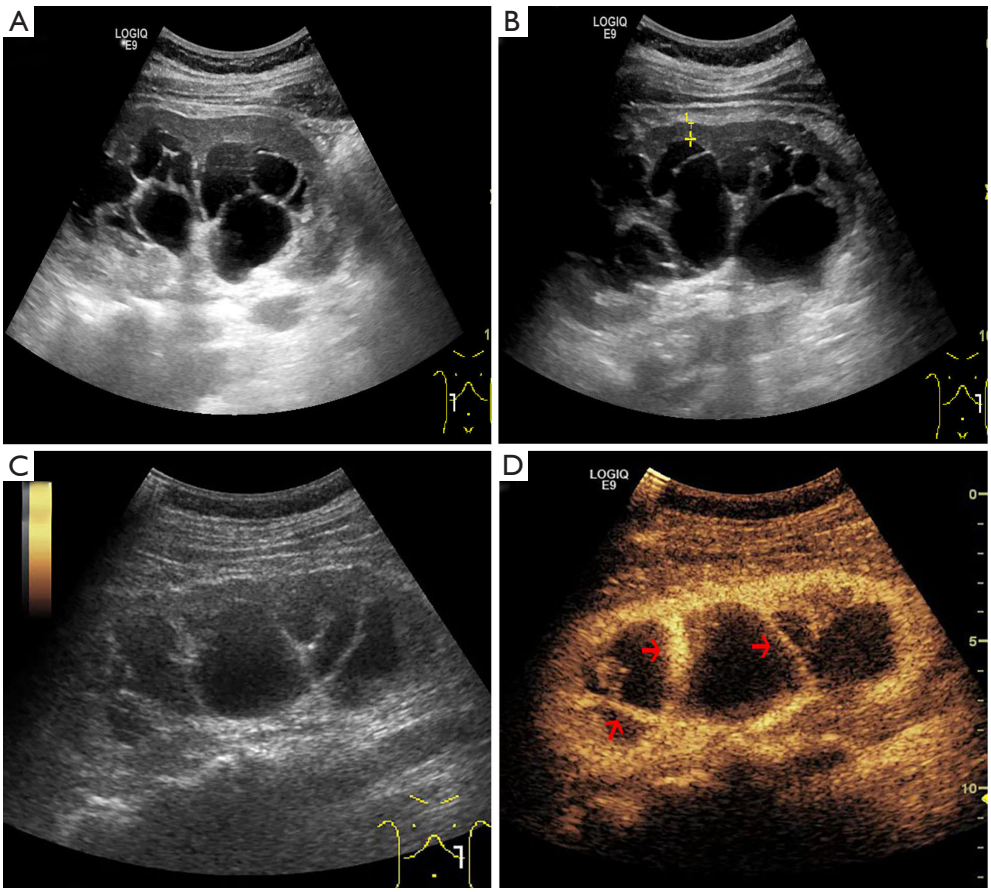

Figure 1 A 38-year-old female (patient A) with known renal lymphangiomatosis (case 1). (A,B) A multilocular cystic area resembling hydronephrosis was found in the renal sinus, with a fine wall and slight thickening in some areas. The renal cortex was locally thin in both kidneys. (C,D) CEUS appearance of renal lymphangioma: the capsule walls and septa are filled with contrast agent (arrows). CEUS, contrast-enhanced ultrasound.

Case 1

Patient A, A 38-year-old female, was found to have hydronephrosis during routine physical examination. Consequently, the patient presented to the Women's Hospital School of Medicine Zhejiang University for further diagnosis. Ultrasonographic findings showed that the volume of both kidneys had increased, and multilocular cystic areas similar to hydronephrosis were found in the renal sinus with a fine wall and slight thickness in some areas and no obvious communication between the capsules and local thinning of the cortical areas of both kidneys (*Figure 1A,1B*). For further confirmation, contrast-enhanced ultrasound (CEUS) was performed, the results of which showed that no contrast agent had filled in the cystic lesions and a little contrast agent had filled in the capsule wall and septa (*Figure 1C,1D*). These results suggested renal lymphangioma. Enhanced computed tomography (CT) examination was not performed because the patient was allergic to the iodine contrast agent. An investigation of the patient's family history revealed that her sister and father had a positive history of renal cysts. Upon our request, all family members were examined by ultrasound (*Figure 2*). No new cases were found in addition to the patient's sister and father who had the same renal cystic lesion. During the follow-up period, patient A retained good and stable renal function without obvious discomfort, so she did not receive any treatment.

Case 2

Patient B (sister of patient A) is 37 years old and showed no obvious abnormality in physical and laboratory examinations. A 2-dimensional (2D) ultrasound

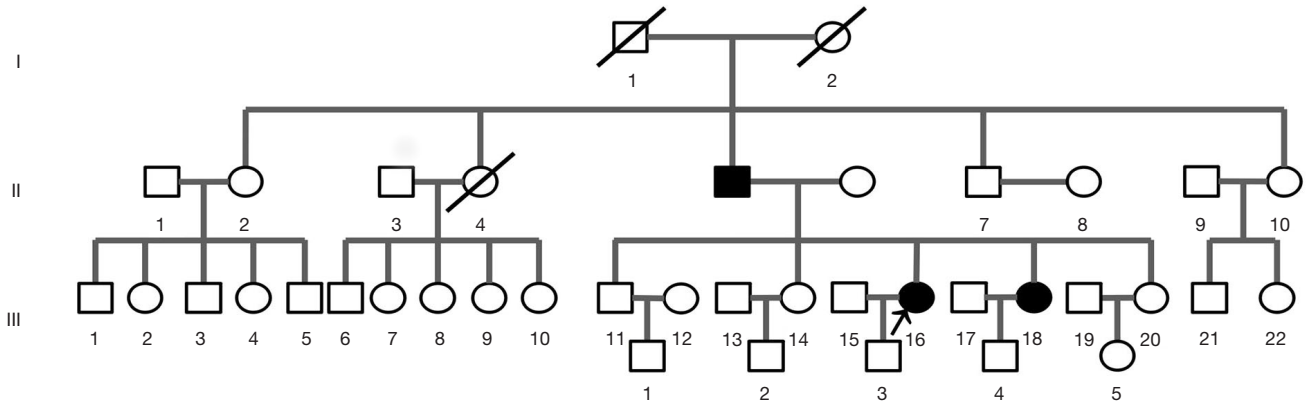

Figure 2 Family atlas of patient A (case 1). ↗ , proband; □○, normal; ■●, patient; /, deceased.

(*Figure 3A,3B*) and CEUS (*Figure 3C,3D*) showed that the appearance of both kidneys was similar to that of patient A. A plain CT scan showed multiple low-density cystic lesions and scattered small cystic hypodensities in the bilateral renal sinuses and around the left kidney, respectively (*Figure 3E*). The CT enhancement showed that the low-density cystic lesions were not filled with contrast agents in either the arterial or delayed phase (*Figure 3F*). CT urography showed the enlargement of both kidneys, multiple low-density cystic lesions filling the renal sinuses, and compression of the renal pelvis and calyces on both sides (*Figure 3G,3H*). The renal function index of patient B was in the normal range, she self-reported no obvious discomfort, and the lesion did not increase significantly during the follow-up, so she did not receive any treatment.

Case 3

Patient C was the father of patients A and B. In the family investigation, the renal ultrasound imaging manifestations of patient C were similar to those of the 2 sisters (*Figure 4A,4B*). The ultrasound examination of other organs showed no cystic changes, he self-reported no obvious discomfort symptoms, the routine examination of renal function and urine showed no obvious abnormalities, and there were no urinary tract complications, so he did not receive any treatment. There was no further increase in the range of lesions during follow-up.

Case 4

A 30-year-old woman presented with hypertension for more than 3 years and presented to the doctor with low back pain 6 years ago. The physical examination showed grade I pitting edema of both lower limbs. The laboratory and imaging results were as follows: routine urine tests included urine occult blood ++, urine protein +++, a white blood cell count increase to 55.8 cell/μL (range, 0.0–28.0 cell/μL), a red blood cell count increase to 13.3 cell/μL (range, 0.0–3.0 cell/μL), a bacterial count increase to 820.1 cell/μL, and chyluria +; meanwhile, renal function findings included a serum creatinine level increase to 109.6 μmol/L and a creatinine clearance rate decrease to 36.6 mL/min. The ultrasonographic appearances (*Figure 5A,5B*) were as follows: a multilocular cystic area in both renal sinuses and perirenal area; ultrasound prompts showing bilateral renal enlargement, a multilocular cystic area in both renal sinuses and the perirenal area, and multiple fine septa in the perirenal cystic area. In addition, the renal sinus appearance was similar to that of hydronephrosis, and renal lymphangioma was considered. The CEUS (*Figure 5C,5D*) revealed that both renal cortices were well filled with contrast agents, renal sinuses and perirenal cystic areas were not filled with contrast agents, and the capsule wall was filled with contrast agent. The results of enhanced CT (*Figure 5E,5F*) were as follows: the parenchyma of both kidneys were thinned and significantly enhanced, there was copious fluid accumulation in the perirenal and renal sinus, the structures of renal pelvis and calyces on both sides were difficult to distinguish, and exudation was observed in the pararenal space on both sides. The diagnosis was renal lymphangioma. During hospitalization, she experienced sudden and transient total amnesia. Skull magnetic resonance imaging (MRI) + diffusion weighted imaging (DWI) showed multiple ischemic infarcts in the right thalamus and pedunculus

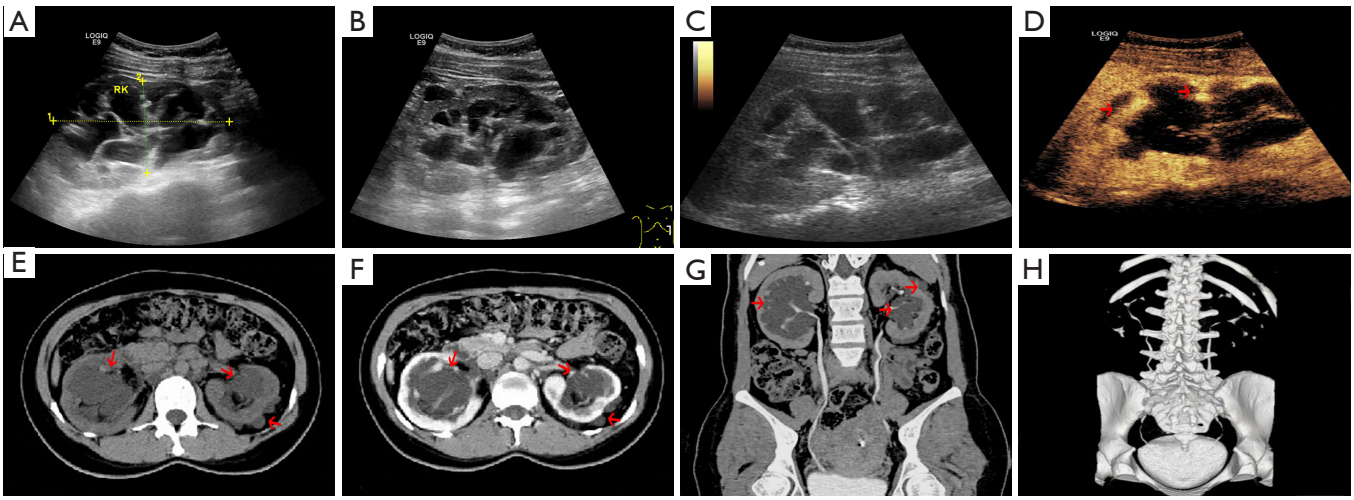

Figure 3 A 37-year-old female (patient B) with known renal lymphangioma (case 2). (A,B) A multilocular cystic area resembling hydronephrosis was found in the renal sinus, with a fine wall and slight thickening in some areas. The renal cortex was locally thin in both kidneys. (C,D) CEUS appearance of renal lymphangioma: the capsule walls and septa are filled with contrast agent (arrows). (E) CT plain scan showing cystic dilatation in the renal pelvis of both kidneys and around the left kidney (arrows). (F) Enhanced CT showing a small amount of fluid around the left kidney and bilateral hydronephrosis (arrows). (G,H) CT urography showing a small amount of fluid around the left kidney (arrows); distortion of renal calyces due to the cystic change of renal sinus. RK, right kidney; CEUS, contrast-enhanced ultrasound; CT, computed tomography.

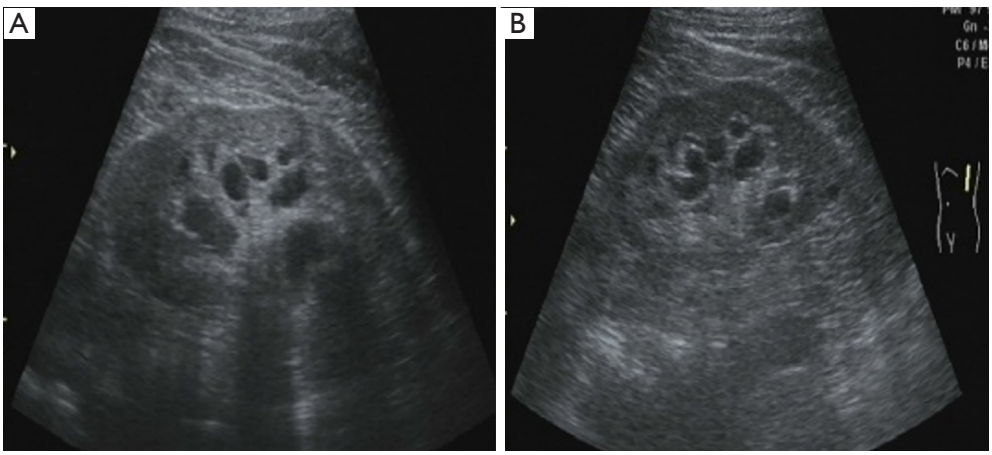

Figure 4 A male (patient C) with known renal lymphangioma (case 3). (A) Ultrasonographic findings of the right kidney. (B) Ultrasonographic findings of the left kidney.

cerebri. Brain magnetic resonance angiography (MRA) and cervical CT angiography (CTA) showed a small aneurysm in the ophthalmic segment of the left internal carotid artery. The department of interventional surgery recommended close observation of the patient's condition, and digital subtraction angiography (DSA) was not considered for the time being. The urology department suggested conservative treatment with regular follow-up. She was followed up for 7 years because of recurrent urinary tract infections. Laboratory and imaging examinations showed that the white blood cell count had increased to 848.5 cell/μL (range, 0.0–16.9 cell/μL), the bacterial count increased to 6,346.1 cell/μL (range, 0.0–130.7 cell/μL), and the serum creatinine level increased to 163 μmol/L, all of which were higher than before. Ultrasound and CT examinations showed that the cystic lesions in the perirenal

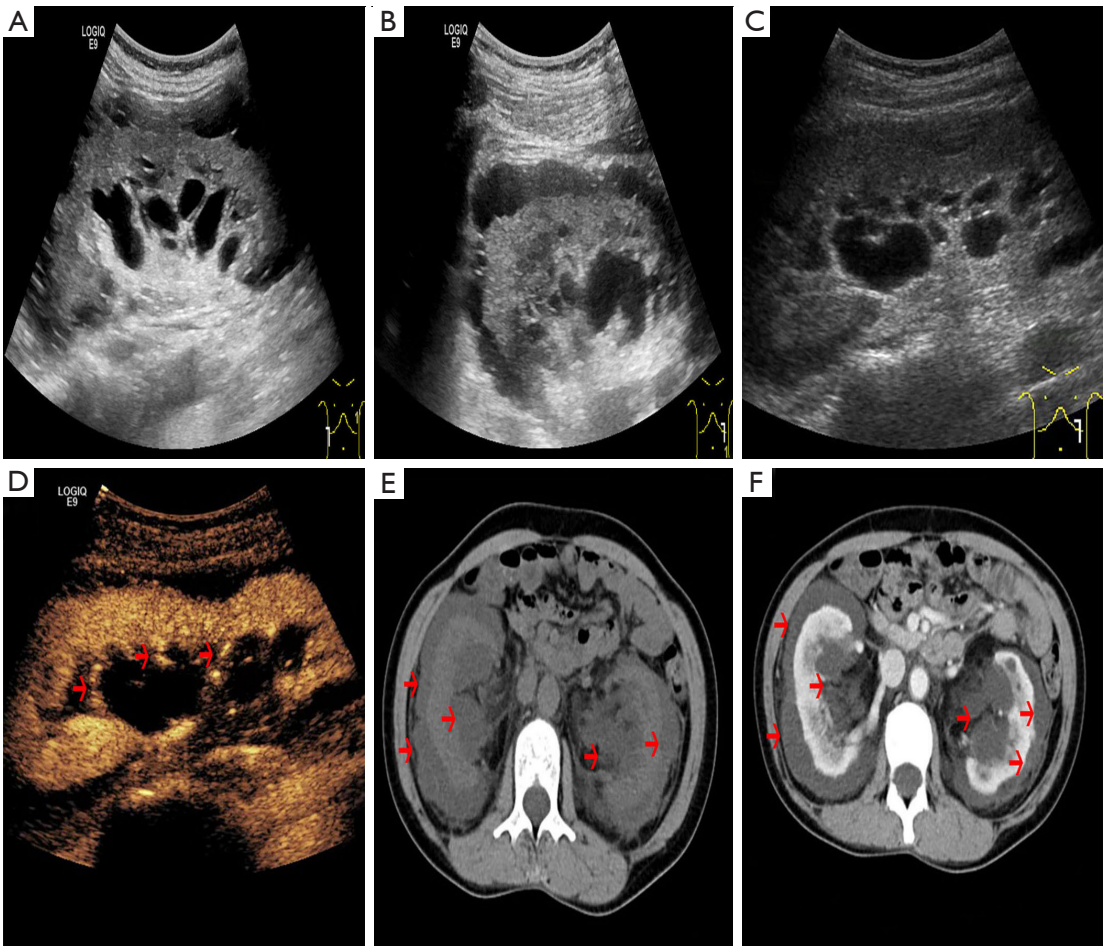

Figure 5 A 30-year-old female with known renal lymphangioma (case 4). (A) Ultrasonographic findings of the right kidney. (B) Ultrasonographic findings of the left kidney. (C,D) CEUS began filling at the 9th second, and reached the peak value at 13th second, and contrast medium filling can be seen in part of the capsule wall and septum (arrows). (E) CT plain scan showed cystic dilatation in the renal pelvis and around both kidneys (arrows). (F) Enhanced CT showed a large amount of fluid around the kidneys and a large amount of fluid within the kidney pelvis (arrows). CEUS, contrast-enhanced ultrasound; CT, computed tomography.

and renal sinuses on both sides had enlarged since their last observation. The patient's renal indicators gradually deteriorated. In addition, the lymphangioma was located in the renal sinus structure and could not be treated surgically. Conservative treatment (e.g., reducing blood pressure and protecting the kidney) was accepted after discussion with the patient.

Literature review

Relevant medical literature (156 and 684 in Chinese and English languages, respectively) about lymphangioma published since 1928 were searched. According to the inclusion and exclusion criteria, 89 patients in 81 articles were analyzed for summary and tabulation (*Table 1*). Moreover, 2 reviewers reviewed each article in detail.

By literature review, understanding and mastering the imaging characteristics can help the physician improve the diagnosis of renal lymphangioma, so as to formulate an effective and comprehensive treatment plan to avoid unnecessary surgery due to misdiagnosis.

Description of renal lymphangioma

Renal lymphangioma is also known as renal lymphangiectasia (14,15) because the disorder is characterized by involvement

Table 1 Information summary of 89 cases of renal lymphangioma

Characteristic	Summarized results
Gender (M:F)	53:36
Age	Newborn–87 years
Localization (bilateral:unilateral)	56:33
Clinical symptoms, n (%)	
Flank pain	27 (30.34)
Hypertension	26 (29.21)
Hematuria	17 (19.10)
Proteinuria	7 (7.87)
Abdominal pain	29 (32.58)
Palpable lump	12 (13.48)
Anasarca	3 (3.37)
Classify, n (%)	
Perinephric	32 (35.96)
Peripelvic	11 (12.36)
Perinephric + peripelvic	39 (43.82)
Special type	7 (7.87)
Image examination, n (%)	
US	74 (83.15)
CEUS	3 (3.37)
IVU	9 (10.11)
CT scan	68 (76.40)
CT enhancement	61 (68.54)
MRI	19 (21.35)
MRU	2 (2.25)
Lymphoscintigraphy	1 (1.12)
Clinical treatment, n (%)	
Nephrectomy/partial nephrectomy	30 (33.71)
Open biopsy	6 (6.74)
Aspiration	13 (14.61)
Puncture	11 (12.36)
Conservative treatment	23 (25.84)
Not mentioned	8 (8.99)

M, male; F, female; US, ultrasound; CEUS, contrast-enhanced ultrasound; IVU, intravenous urography; CT, computed tomography; MRI, magnetic resonance imaging; MRU, magnetic resonance urography.

of the renal lymphatic system, including perinephric, pararenal, and intrarenal lymphatics (16,17). It is a cystic area with localized lymphatic stasis due to developmental obstruction of regional lymphatic drainage, rather than a true tumor or a hamartoma (18,19). Moreover, 70% of lymphangiomas occur in the head and neck, and bleeding can cause dysphagia and discomfort. In addition, 25% of lymphangiomas occur in the chest wall and limbs (20). Furthermore, 5% of lymphangiomas occur in the internal organs and are more commonly seen in the intestine and mesentery. However, renal lymphangioma is rare.

Clinical manifestations of renal lymphangioma

A total of 81 literatures were reviewed, including 89 patients, with a male-female ratio of 1.46:1. The disease can occur at any age, from newborns to those 87 years old, with a higher incidence rate at 20–50 years old. Age distribution supports the notion that lymphangiomatosis is a congenital malformation. Disease growth is either slow or the incubation period of the disease is very long because it occurs in a relatively safe area of the body. Thus, it is commonly discovered late (1). Most of the lesions involve both kidneys. Patients manifest different clinical symptoms, including abdominal pain, lower back pain, hypertension, hematuria, palpable mass, proteinuria, edema, and others, due to the different sizes and positions of lymphatic sinuses. However, small cysts are generally asymptomatic and are mostly found accidentally during the examination. Larger cysts may cause a dragging feeling due to the compression of adjacent structures, resulting in lower back and abdominal pain (21). The 4 most common clinical symptoms are abdominal pain, lower back pain, high blood pressure, and hematuria (*Table 1*).

Diagnosis of renal lymphangioma

Renal lymphangioma is divided into capillary lymphatic, cavernous, and cystic types in clinical pathology because it has the characteristics of involving the perinephric, pararenal, and intrarenal lymphatics (21). Thus, it can be divided into microcyst, large cyst, and mixed types according to the clinical and histological features (22). The disease is divided into 4 types based on its imaging findings (*Figure 6*): perinephric and peripelvic, perinephric, peripelvic (similar to the hydronephrosis type), and special types (including

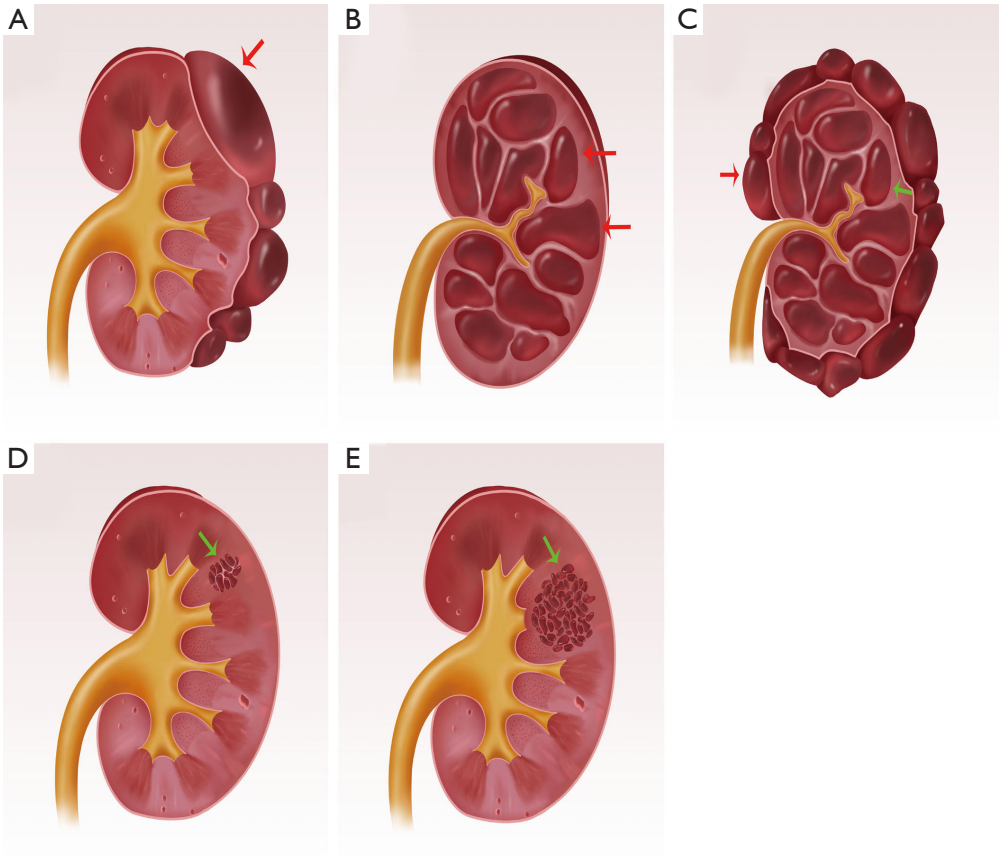

Figure 6 A pattern map of renal lymphangioma based on imaging findings. (A) Perinephric type. (B) Peripelvic type. (C) Perinephric + peripelvic type. (D) Special type 1. (E) Special type 2. The arrows indicate the lesions.

special types 1 and 2). Among the 89 articles screened, imaging examination was the main means of screening: 74 cases were examined by ultrasound, 68 cases were examined by plain CT scanning, 61 cases were examined by CT enhancement, 19 cases were examined by MRI, 9 cases were examined by intravenous urography (IVU), 3 cases were examined by CEUS, 2 cases were examined by MR urography (MRU), and 1 case was examined by radionuclide lymphography. The characteristics, advantages, and disadvantages of various imaging examinations of the disease are shown in *Table 2*. Statistics show that (*Table 1*) ultrasound and CT can be used as the primary means for the diagnosis of the disease because they have no side effects and are easy to operate. However, CEUS or MRI can provide further information regarding tumor size and extension. Before 2002, renal lymphangioma was diagnosed by biopsy and surgical pathology, after which time it was diagnosed according to imaging and clinical manifestations so that the patients could be conservatively treated (15,23,24). According to literature reports, MRI of the disease can manifest as the presence of fluid or fluid-filled structures in the retroperitoneum adjacent to the great vessels at the level of the renal hilum and crossing the midline (16,25-27). This manifestation is thought to be the dilation of the renal lymphatic vessels into the larger retroperitoneal lymphatic vessels. Moreover, this sign further confirms the diagnosis of renal lymphangiectasis.

In addition, a special type tends to occur in childhood. Two kinds of ultrasound manifestations were noted. Special type 1 (*Figure 6D*) appears as hyperechoic nodules with clear or fuzzy boundaries (18,28). Its MRI shows hyperintense signal intensity on T2-weighted images, and the dynamic contrast-enhanced images show centripetal enhancement. This may be caused by the slow infusion of the contrast agent into dilated lymphatics. These MRI features can help to differentiate a solid masslike lymphangioma from renal cell carcinoma, angiomyolipoma, and other conditions (18). Under normal circumstances, the lesion size depends on

Table 2 Various imaging findings of renal lymphangioma and their strengths and limitations

Summarize	IVU	US	CT	MRI	MRU
Imaging features	Normal or enlarged renal volume, compression, and distortion of the renal pelvis and calyceal system	Multilocular cystic structures are seen around the kidney and (or) next to the renal pelvis, which is not connected, multiple diaphragms are seen, and blood flow signals may or may not be seen on the septum	The cystic wall and septum of the lesion appear unevenly enhanced. After enhancement, the compressed and thinned residual renal cortex and the compression of the collecting system can be distinguished	T1 low signal, T2 high signal, and special type (similar to hamartoma) T2 show high-signal intensity and centripetal enhancement	Irregular long T1 and long T2 signals were seen around the kidney, ureter, and renal hilum vessels, with low signal separation within, and the lipid pressure sequence was high signal
Strengths	Sensitive to small lesions of the renal pelvis	Noninvasive, real-time imaging	High-density resolution	High resolution of soft tissue; multisequence imaging	Obstruction degree can be determined
Limitations	People with poor kidney function cannot use it	Susceptible to intestinal gas interference	Large radiation dose	Long scan time; motion artifacts; not suitable for microvascular imaging	Is not independently qualitative; low spatial resolution

IVU, intravenous urography; US, ultrasound; CT, computed tomography; MRI, magnetic resonance imaging; MRU, magnetic resonance urography.

the lymphatic obstruction location. Diffuse bilateral renal lymphangiectasis will occur if the large lymphatics draining the kidney through the renal pedicle are blocked. Moreover, blocking the smaller intrarenal lymph may lead to localized renal lymphangioma (17). Special type 2 (*Figure 6E*) refers to the diffuse change of the echo of the renal collecting system (14,29). Unlike with a true tumor, the echo of the collecting system is enhanced, the CT manifestations are nonspecific, the kidney is enlarged, and the boundary of the collecting system is unclear. However, the reversal of cortical medulla signal intensity in half-Fourier acquisition single-shot turbo spin-echo (HASTE) sequences in MRI urinary imaging may be an important finding (15,25). It seems that this performance type can be explained and distinguished from other performances. However, further confirmation is still needed. The enhanced echogenicity of the collecting system may be caused by the tiny, dilated lymphatic vessels scattered in the renal tubules and blood vessels (17).

Differential diagnosis of renal lymphangioma

The most common differential diagnosis of renal lymphangioma and the diseases with the highest misdiagnosis rate are polycystic kidneys and hydronephrosis (4-6). Adult polycystic kidney disease is an autosomal-dominant genetic disease, most cases of which are accompanied by multiple cysts in the liver, pancreas, and other organs. The renal parenchyma is filled with several cysts of different sizes, with the cysts not being connected with the renal pelvis or calyces. In addition, the renal cortex appears as a cyst, whereas in renal lymphangioma, the renal cortex appears to be normal. Hydronephrosis manifests as the dilation of the renal calyces and communication with the renal pelvis and the ureter (2). Renal lymphangioma should also be differentiated from parapelvic cysts. A CT examination is the most reliable diagnostic method for parapelvic cysts, showing that the cysts are located in the hilum and separate from the normal renal parenchyma. The lowest density halo formed by renal sinus fat around the cyst is the characteristic feature of the disease. The CT value is 0–20 Hounsfield units (HU), and the CT value changes little before and after enhancement. However, the CT findings of renal lymphangioma are cystic lesions under the renal capsule or in the renal parenchyma, or diffuse distribution around the kidney, with uniform density and clear boundary, and the CT value of cystic lesions is generally higher.

However, comparing renal lymphangioma with cystic kidney cancer, multilocular cystic kidney tumors, Wilms tumors, mesodermal renal tumors, and clear cell renal sarcoma is difficult because 5–15% of renal malignant tumors will have multilocular cystic changes (30). However, clinical history, normal biochemical indicators, and typical imaging findings can identify some of the aforementioned

diseases. Only a few complex and atypical multilocular cystic structures are difficult to distinguish from malignant tumors, and accurate judgments need to be made by biopsy and postoperative pathology.

One of the most current accurate diagnostic methods is immunohistochemical staining, which can be used to determine the origin of the flat cells that line up the saclike structure. According to the literature, lymphatic endothelial cells lack immunoreactivity, whereas lymphoid tissues have been shown to contain factor VIII-related antigen (14). Not reacting to AE1, AE3, or 5D3 suggests that the cells are not of epithelial origin (14). Immunoreactivity with the factor VIII and Ulex europaeus antibodies that recognize endothelium helped to differentiate between endothelial and epithelial linings.

Treatment and prognosis of renal lymphangioma

Most cases cannot be accurately diagnosed by imaging examinations due to the rarity of lymphangioma and lack of disease awareness (1). The data group of this study showed that 19 patients with renal lymphangioma underwent invasive methods (11, 5, and 3 cases underwent radical renal surgery, open biopsy, and fine-needle aspiration, respectively) from 1928 to 2002 for disease diagnosis. Patients who were symptomatic but not eligible for surgery received percutaneous drainage. However, this treatment only temporarily relieves the symptoms, and most cases treated in this way will relapse (24). Some patients are misdiagnosed with malignant lesions, leading to nephrectomy or marsupialization (1). Marsupialization may cause massive bleeding (31) and ultimately result in nephrectomy because the lesions often involve both sides. Reports in the literature have also shown that the area of the lymphangioma in the other kidney will increase and cause complications in a short period after the removal of 1 kidney (24). In addition, in 2 cases (8,24), it was reported that the lesion area increased sharply 2 months and 6 years after puncture drainage, respectively. Recurrence was reported in 1 case (5) after 8 months of follow-up nephrectomy, and only 1 case report (30) mentioned an 8-mm lesion being successfully removed by retrograde ureteroscopy without recurrence after 24 years of follow-up. For the patients in our current report, 3 members of the family in case 1 were not treated and were only followed up. The renal function was stable for 13 years. In case 4, the patient had renal insufficiency, and after conservative treatment, experienced repeated hematuria during the follow-up period, had increased creatinine value, and had gradually deteriorating renal function, which indicates that the occurrence and development of the disease is a gradual process (24). Therefore, patients without obvious disease symptoms can choose conservative treatment, and patients with symptoms causing obvious discomfort due to the acceleration of the disease can choose ultrasound-guided percutaneous drainage or surgical treatment according to their specific situation. Antihypertensive drugs and diuretics can be used for symptomatic treatment for patients with hypertension and abdominal effusion (32).

Complications of renal lymphangioma

Complications of the disease include hematuria (27-31), ascites (8,10,24,25), hypertension (33-35), renal vein thrombosis (24), and renal impairment (24,36), among others. The cause of hypertension may be the pressure of renal parenchyma by perirenal effusion, which affects the insufficiency of blood perfusion in renal tissue and leads to increased secretion of renin, which results in the increase of blood pressure. This is also known as the Page kidney phenomenon (31). The formation of renal vein thrombosis may be the final result of the gradual expansion of the renal lymphatic vessels (24). Partial natural improvement of the disease was reported in a pair of pregnant sisters (12) and a newborn (29), suggesting that the disease is self-limited.

Outlook

Few molecular pathological studies are currently available on renal lymphangioma, and the etiology and pathogenesis of the disease are not completely clear. A review of the literature found that genetic evidence of the disease is limited, and many sporadic cases were noted. Blesinger *et al.* (37) have suggested that somatic mutations may be the cause of the lesion. The *PIK3CA* mutation that encodes the catalytic subunit of the PI3K enzyme was found in 74–94% of patients with lymphangioma (37). These mutations are also common in cancer, showing high expression, but not common in normal lymphatic channels. However, whether *PIK3CA* mutations alone can produce lymphangioma is unclear. In another article (38), the family members of a 15-year-old girl with left renal lymphangioma disease and a previous history of congenital polycythemia, were examined. Consequently, the renal lymphangioma disease may be related to the *D126Y* and *D92Y* mutations in the von Hippel-Lindau (*VHL*) gene. However, after consulting

the relevant information, congenital polycythemia is a genetic disease. According to international reports, this type of disease is related to the *VHL* gene (39). Thus, the correlation with renal lymphangioma should be excluded. It is believed that with the continuous advancement of medical standards, the development of molecular biology, and the support of big data, there will be future breakthroughs in the diagnosis and treatment of the disease.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-314/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). This retrospective study was approved by the Institutional Ethics Committee of Women's Hospital School of Medicine Zhejiang University (approval number: IRB-20220046-R). Written informed consent was provided by the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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