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Case Report and Review of the Literature

Renal Lymphangiectasia as an Unusual Cause of Perirenal Collections: A Case Report and Literature Review

Long Jiahuan Xanicia[1*] and Ng Kok Kit[2]

[1]Ministry of Health Holdings, Singapore

[2]Department of Urology, Changi General Hospital, Singapore

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ABSTRACT

Renal lymphangiectasia (RLM) is a rare condition characterized by dilatation of perirenal, parapelvic or intrarenal lymphatics. We report an unusual case of bilateral RLM in a 20-year-old Malay male who presented with 1 month duration of bilateral flank pain. Bedside ultrasound showed bilateral perinephric collections, which were further confirmed on computer tomography intravenous pyelogram (CT IVP). Other causes of perinephric collections were ruled out based on laboratory and radiological findings, and he was managed conservatively as for RLM with surveillance ultrasound kidneys at 3 to 6 monthly intervals.

Introduction

RLM is hypothesized to result from developmental malformation of the renal lymphatic tissues leading to obstruction and accumulation of lymph in the subcapsular region or hilum. Due to variable imaging findings, the differentials may be broad and diagnosis can be difficult. We aim to describe the clinical presentation, radiological findings, differentials and management of this condition using a case report and a review of the literature.

Case Report

A 20-year-old male presented to the emergency department for bilateral flank pain over 1 month. His past medical history includes L4/5 disc desiccation. Functionally, he is community ambulant and independent in activities of daily living. On examination, his abdomen was soft with tenderness over both flanks. Renal punch was equivocal bilaterally. Neurological examination of the lower limbs was unremarkable. His serum chemistry was as follows: white cell count 12.2×10^3/μL, haemoglobin 14.4g/dL, platelet count 210×10^9/L, C-reactive protein 25.9mg/L, creatinine 143μmol/L. Urinalysis showed 4 red cells per high power field, 2 white cells per high power field, 0 epithelial cells. Urine culture showed no bacterial growth. Bedside ultrasound in the emergency department showed bilateral peripheral collections. CT IVP revealed bilateral symmetrical perinephric fluid collections measuring 12mm thick with some free fluid within the retroperitoneum and pericolic gutters (Figure 1A). Hounsfield unit for the collections ranged <10. There was no urinary calculus, solid renal mass or hydronephrosis. The delayed excretory phase showed no contrast extravasation into the collections (Figure 1B), thereby ruling out the possibility of urinoma.

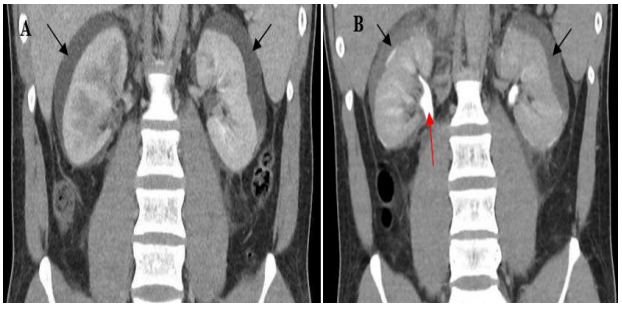

Figure 1: CT IVP **A)** medullary **B)** delayed/excretory phase. Red arrow showing ureter enhancement. 12mm perinephric collections (black arrows), non-enhancing in excretory phase.

[*]Correspondence to: Long Jiahuan Xanicia, Ministry of Health Holdings, Singapore; E-mail: xanicia@hotmail.com

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Additional urine studies included 24h urine protein of 0.22g/day and random urine protein of 0.11g/L, hence ruling out proteinuric nephropathy. The patient was offered aspiration of collections for fluid analysis, which he declined and instead opted for conservative management with surveillance US kidneys every 3-6 months. At the 9-month mark, there was an interval increase in the size of the left upper to mid pole collection to 6.3cm×5.3cm×2.0cm (Figure 2) and the patient was re-offered aspiration, which he declined as he was asymptomatic. At the 15-month mark, there was a further increase in the size of the above-mentioned collection to 6.8cm×7.1cm×2.1cm (Figure 3). The latest US kidneys at the 21-month mark showed a reduction in size to 7.3cm×5.0cm×1.9cm. The patient remained asymptomatic throughout his course of follow-up consults.

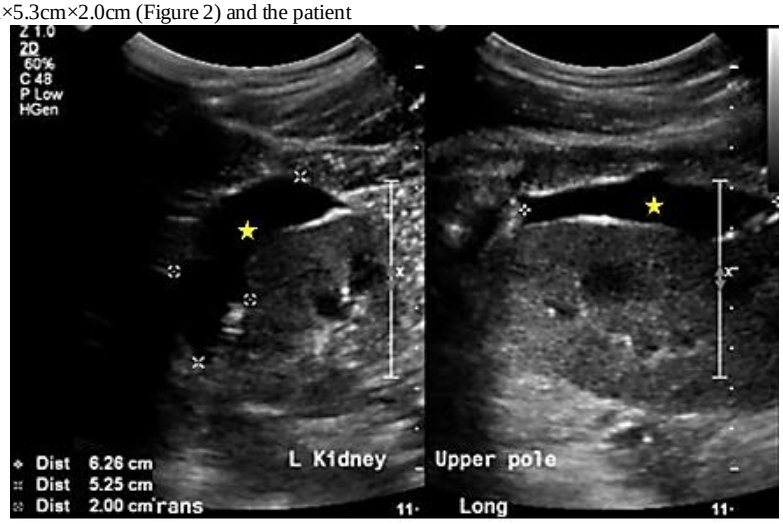

Figure 2: US kidneys at 9 months. 6.3cm×5.3cm×2.0cm left upper to mid pole collection.

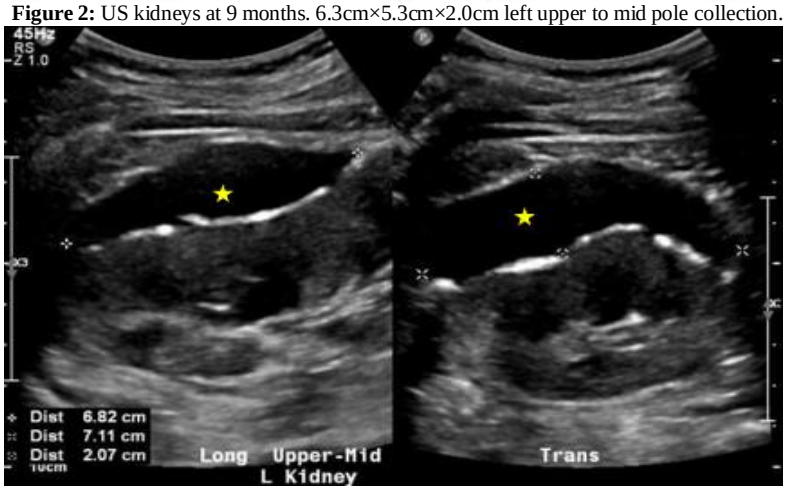

Figure 3: US kidneys at 15 months. 6.8cm×7.1cm×2.1cm left upper to mid pole collection.

Table 1: Presentation, imaging findings and management of previous case reports.

	Age	Sex	Comorbid	Presentation	Imaging findings	Complications	Management
Pandya VK *et al.* [1]	34	M	Nil	Bilateral flank pain	US: anechoic PN collections 33mm in width CT: PN collections of density 0-10HU	Nil	Conservative
Chen Z *et al.* [2]	34	F	Nil	Fever, right flank pain	US: dilatation of right PC space and right PN collection CT: bilateral polycysts in the renal sinus and right PN collection	Nil	Emergency exploratory surgery, followed by nephrectomy for uncontrolled hilar leakage
Choudhury S *et al.* [3]	16	M	Nil	Abdominal distension, pain	CT: subcapsular PN collections without PC communication, compression of PC system	HTN	Percutaneous drainage and sclerotherapy

Ashraf K *et al.* [4]	23	F	Para umbilical hernia	Abdominal distension and pain x1/12	US: ascites and bilateral dilatation of PC system CT abdomen: bilateral renal sinus collections 15HU	HTN, ascites	Conservative with diuretics and antihypertensive
Koc NS *et al.* [5]	30	M	Young HTN	Diagnosed as ADPKD at age 20 based on US kidney findings > MRI done 10y later to calculate kidney volume for tolvaptan treatment	MRI kidneys: multiple peripherally located cysts hyperintense on T2WI and hypointense on T1WI	HTN	Conservative
Umapathy S *et al.* [6]	49	M	Nil	Left loin pain x1/12	CT: 9 x 6.5 x 6.2 cm cystic lesion in left renal sinus distorting the PC system with PN fat stranding	Renal vein thrombosis	Conservative
Al-Dofri *et al.* [7]	22	M	Nil	Abdominal distension and pain, dyspnea x6/12	CT: renal sinus cysts 3HU ; PN collection 2HU; right pleural effusion and ascites 3HU	Pleural effusion, ascites	Conservative with diuretics
Pianezza *et al.* [8]	52	M	Pancreatic psuedocyst	Bilateral flank pain and gross haematuria	US: PN hypoechoic lesions with thin septations, loss of corticomedullary differentiation CT: multilocated pararenal cystic lesion, density of 0-10 HU, atrophic kidney MRI kidneys: non enhancing cystic lesion on T1WI; increased cortical and decreased medullary intensity on T2WI	Gross haematuria	Conservative
Blanc M. *et al.* [9]	58	M	Newly diagnosed HTN	Fatigue Hb 213g/L Hct 0.63	US: unilateral right PN anechoic collection with multiple septa CT: water-density right PN collection with lobulated contours extending to the renal hilum with renal parenchymal compression Renal scintigraphy MAG-3: renal function of 47% on the right kidney and 53% on the left	HTN, polycythemia	Percutaneous drainage followed by laparoscopic bilateral marsupialisation
Ali K. *et al.* [10]	50	F	Lupus nephritis s/p 3 renal transplants	Right lower quadrant pain 13 months following last renal transplant	US: multi-septated thin-walled fluid collection in the hilum separate from collecting system MRI kidney: non enhancing peripelvic multiseptated collection hypointense on T1 hyperintense on T2	Worsening kidney function post-transplant	Percutaneous drainage initially and then wide peritoneal fenestration and omentoplasty
Hamroun A. *et al.* [11]	34	M	ESRD 2' FSGS s/p renal transplant	Refractory ascites 10 years following kidney transplant	MR urography: multiple plurilobular fluid collections located at parapyelic, juxtacapsular, and perihilar regions	Inguinoscrotal hydrocele, bladder compression from mass effect	Conversion to mammalian target of rapamycin (mTOR) inhibitor Hydrocele managed conservatively

	35	M	ESRD 2' childhood HUS s/p renal transplant	Chronic pelvic pain with LUTS for 8 years	US: nephromegaly (16 cm), multiple peripyelic cysts, perihepatic, and perisplenic ascites MR urography: nephromegaly (729cc) pericapsular collection and edematous infiltration of perirenal fat	Bladder compression from mass effect resulting in LUTS	Mofetil mycophenolate was switched to mTORi
Dawidek M. T. *et al.* [12]	55	F	ESRD 2' microscopic polyangitis s/p renal transplant	New onset ascites 4 years post transplant	US kidneys: loss of corticomedullary differentiation with subcapsular fluid densities	Ascites, umbilical and laparoscopic port incisional hernias	Allograft nephrectomy with concurrent hernia repair

PN: Perinephric; PC: Pelvicalyceal; HU: Hounsfield Units; HTN: Hypertension; Hb: Haemoglobin; Hct: Haematocrit; ESRD: End Stage Renal Disease; FSGS: Focal and Segmental Glomerulosclerosis; HUS: Hemolytic Uraemic Syndrome; LUTS: Lower Urinary Tract Symptoms

Discussion

Renal lymphangiectasia (RLM) is an uncommon cause of perirenal and parapelvic collections. It can present unilaterally or bilaterally and has no age or gender predilection [6]. The familial nature of the disease has been reported [13]. Its pathogenesis is hypothesized to result from developmental malformation of the renal lymphatic tissues. The lymphatic drainage of the kidney, renal capsule, and the perinephric region intercommunicate through lymphatic trunks within the renal sinus [6]. These lymphatics drain into the paraaortic, paracaval, and inter-aortocaval lymph nodes [6]. In RLM, there is impairment in the drainage of larger renal sinus lymphatic trunks with resultant dilatation of peripelvic and perinephric and intrarenal lymphatics [6]. It is usually asymptomatic and incidentally diagnosed on radiological imaging as perinephric or parapelvic fluid collections. When symptomatic, it may be associated with pain, abdominal distension, hematuria, hypertension, and less commonly deterioration of renal function, renal vein thrombosis, polycythemia and pleural effusion (see Table 1 for a summary of different presentations of RLM from previous case reports).

Differentials commonly include but are not limited to [5, 6, 8, 9]:

i. Polycystic kidneys
ii. Hydronephrosis
iii. Urinoma
iv. Cystic nephroma
v. Lymphoma
vi. Nephroblastomatosis

These conditions may be difficult to differentiate from RLM using a single imaging modality, and hence the need for correlation of clinical history, biochemical parameters and additional imaging modalities beyond ultrasound (Table 2).

Table 2: Summary of differentials for RLM and their imaging findings.

Differentials	**US findings**	**Contrasted CT findings**	**MRI findings**
Polycystic kidneys	Massive enlarged kidneys Multiple anechoic cysts of varying sizes Distortion of normal renal parenchyma	Cysts with fluid attenuation can be found in the cortex/hilar space/subcapsular space No enhancement	MRI rarely indicated, unless for workup for complicated/complex cysts with the need to rule out malignant component on the background of impaired renal function
Hydronephrosis	Dilation (not displacement) of pelvis and calyces	Opacification of collecting system in delayed phase from the filling of contrast Source of obstruction identifiable at renal pelvis/ureter/vesicoureteric junction/ bladder	MRI rarely indicated
Urinoma	Thin walled anechoic collection contouring any portion of the renal tracts	Urinary leakage in delayed phase due to direct contrast extravasation from the urinary tract	T1: hypointense T2: hyperintense
Multilocular cystic nephroma	Multilocular anechoic mass originating from kidney -- claw or beak-shape of adjacent renal parenchyma Septal vascularity	Encapsulated well-circumscribed mass with near-water HU Enhancing septa and no excretion of contrast agent into the cyst No nodular or solid enhancement	T1: variable signal, depending on the protein or blood products of the cysts T2: hyperintense

Lymphoma	Internal vascularity of mass	Soft tissue attenuation with enhancement Associated with: splenomegaly, retroperitoneal lymphadenopathy	T1: hypointense T2: iso- or hyperintense
Nephroblastomatosis	Enlarged diffusely hypoechoeic kidneys	Poorly enhancing soft tissue lesion with adjacent normally enhancing renal parenchyma	T1: low-signal-intensity nodules T2: low-signal-intensity nodules

The features of RLM on US and CT depend on the site and extent of lymphatic obstruction (see Table 1 for variations of radiological findings of RLM from previous case reports). When mainly smaller intrarenal lymphatics are blocked, diffuse enlargement of kidneys may be seen without cystic spaces [4]. Loss of cortico-medullary differentiation may be seen in more chronic cases [4, 8]. On CT, collections may be accompanied by the presence of septa. The attenuation of these collections lies in the range of fluid [1, 7-9]. Imaging findings of perirenal and parapelvic involvement sparing the renal parenchyma are highly suggestive of RLM as opposed to other conditions involving pathology of the renal parenchyma. MRI is not commonly performed as US and contrast-enhanced CT often suffice in ruling out differentials, and any value of performing MRI would be academic. MRI typically shows hyperintense collections on T2W images with reversal of corticomedullary intensity due to fewer small lymphatics in the mid-cortex and absence of lymphatics in the medulla [5, 8, 14].

Asymptomatic collections can be managed conservatively. Diuretics can be used to control ascites and anti-hypertensive medication is used to control hypertension when present [4, 7]. When collections are very large and causing pressure symptoms, percutaneous drainage may be performed and fluid analysis typically shows chylous fluid with lymphocytic predominance with small amounts of fat globules, proteins and high renin levels in the fluid [3, 4, 6, 9, 14]. Histological examination of fluid may also play an important role if there is a suspicion of malignancy. RLM is positive for lymphatic endothelial immunomarkers such as D2-40 and is characterized by cortical dilated endothelial-lined spaces without glomerular or tubular abnormalities [10]. Percutaneous drainage, despite being easy to perform, is often only a temporary measure and cannot be considered a definitive solution because it is inevitably followed by recurrence and might also be difficult to achieve complete drainage in the presence of septations [2, 3, 9, 14]. Definitive surgical interventions reported in the literature include marsupialisation, sclerotherapy and nephrectomy (Table 1) [2, 3, 9, 15, 16]. Marsupialisation has been reported to be complicated by extensive hemorrhage mandating nephrectomy [14, 16]. Possible reasoning would be that lesions in the renal hilum can be difficult to isolate and surgical manipulation at the hilum can lead to unintentional nicking of hilar vessels. Sclerotherapy in conjunction with percutaneous drainage can conversely avoid such complications. Povidone-iodine can be used as a sclerosant as it is homogeneously distributed, long-acting and minimally irritative to the urinary tract, which prevents stricture formation [3, 15]. Nephrectomy is not commonly performed and is reserved for recurrent collections or uncontrolled intraoperative bleeding [4, 17]. Existing literature on RLM mainly describe the condition in native kidneys, and it is rare in the context of renal transplant. The diagnosis of RLM should be considered in renal transplant patients with ascites after all other sources have been ruled out [10-12]. Renal lymphangiectasia with high output ascites may ultimately require treatment with allograft nephrectomy [12].

Conclusion

In conclusion, we present a case of renal lymphangiectasia, which was managed conservatively with surveillance imaging as the patient was asymptomatic. Despite interval increase in the size of perinephric collections from the 9-month to 15-month mark, the patient did not undergo any invasive intervention as he was asymptomatic, and subsequent reduction in the size of collections was noted in the most recent US kidneys. Awareness of this condition will be helpful in differentiating it from other renal cystic diseases and other causes of perirenal collections, which is key in guiding appropriate management and avoiding over-investigating and subjecting the patient to unnecessary procedures. Nevertheless, surveillance scans and follow-up are recommended in all patients due to the risk of complications such as renal failure and hypertension, which might only present at a later stage. Percutaneous drainage is the first line of management in less severe symptomatic cases but has a lower success rate in larger lesions with multiseptation. Marsupialisation, sclerotherapy and nephrectomy are reserved for more symptomatic cases associated with larger cysts or multiple recurrences. Lack of sufficient data in literature makes a comparison in functional outcomes by different surgical techniques difficult.

Conflicts of Interest

None.

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