Renal Lymphangiectasia- Critical Assessment Of A Rare Case And Review Of Literature.

Dr Nishant Raj[1]*, Dr.Rajasbala Dhande[2], Dr.Nikita Bora[3]

*[1] Junior Resident, Department Of Radiodiagnosis, Acharya Vinobha Bhave Rural Hospital, Sawangi (Meghe), Wardha, Maharashtra, India-442001 Email Id- Nishantraj88@Gmail.Com
[2] Hod And Professor, Department Of Radiodiagnosis, Acharya Vinobha Bhave Rural Hospital, Sawangi (Meghe), Wardha, Maharashtra, India-442001, Email Id-Dhanderajas@Gmail.Com
[3] Junior Resident, Department Of Radiodiagnosis, Acharya Vinobha Bhave Rural Hospital, Sawangi (Meghe), Wardha, Maharashtra, India-44200. Email Id- Drnikitabora@Gmail.Com

***Corresponding Author**- Dr. Nishant Raj

*Junior Resident, Department Of Radiodiagnosis, E-Mail:- Nishantraj88@Gmail.Com, +91-8670255587
Doi: 10.47750/pnr.2022.13.S05.141

Abstract

Renal Lymphangiectasia (RLM) is a very infrequent benign lymphatic disorder. RLM is often misdiagnosed for other disorders such as cystic renal masses, usually polycystic kidneys. Although parenthetically found in different cases, RLM is a probable cause of renal failure and hypertension in undiagnosed victims. The etiology of RLM is unclear and results from the formation of cystic masses and dilatation of perinephric lymphatic channels. In the renal sinus, lymphatic channels connect the perirenal tissues and the renal capsule the kidney's lymphatic system. Developmental malformation in the above-mentioned channels is one of the hypotheses presumed to describe the etiopathogenesis of RLM. The other hypothesis presumes the inflammation of the renal perinephric leads to the dilatation and obstruction of lymphatics.

CASE REPORT:

30-year-old female came with complains of swelling in bilateral lower limbs and distension of abdomen since 1 year, She had on and off bilateral flank pain since past 6 months and there was history of intermittent whitish discoloration of urine which has been on and off since past 15 years. She had aggravated similar symptoms 5 years back during her pregnancy. There was no history of fever, facial puffiness, decreased urination, trauma, loss of weight or decreased appetite. On examination, edema was seen in bilateral lower limbs, the abdomen was distended but soft with no guarding or rigidity, Bowel sounds were present. Rest of the systemic examination was unremarkable. Her labs were significant for increased 24 hour urinary protein: 1615 gm, Urine protein/creatinine ratio was increased: 9.1, ESR (Erythrocyte sedimentation rate) was elevated: 72, Kidney and Liver function tests were within normal limits.

On ultrasound, following imaging findings were seen: Normal size kidneys with increase cortical echogenicity and loss of cortico-medullary junction. Multiseptated anechoic collections in perinephric region of maximum width 30mm.No evidence of calculus or hydronephrosis on either side. [Figure 1] Free fluid was seen in abdomen and pelvis suggestive of ascites. [Figure 2]

She further underwent a contrast enhanced computed tomography scan of the abdomen which revealed following findings: Multiseptated collections with fluid density of 0-10 HU in perinephric regions of both kidneys. No evidence of soft tissue or fat density mass lesion seen. Both kidneys show normal parenchymal enhancement and contrast excretion with no leakage of contrast within the perinephric collections. Both ureters were normal in course and calibre. [Figure 3]

These features were in keeping with bilateral renal lymphangiectasia. Confirmation of diagnosis was done by perinephric fluid aspiration and cytology of fluid which revealed sterile chylous fluid containing majority of lymphocytes and small amount of fat.

REVIEW OF LITERATURE:

Literature investigation on PubMed shows five different cases of RLM in pediatric-age patients. In 1995, the first case of unilateral RLM was reported by Merguerian *et al*. in an infant. Merguerian *et al*. settled that RLM should be taken as a differential clinical diagnosis of unilateral renal enlargement among children.[1] After two years, Simonton *et al*. described a similar situation of unilateral RLM in a child aged two years. The child presented flank mass, massive ascites, and hypertension. [2] Cadnapaphornchai *et al*. subsequently reported another case of a child aged 7 years with bilateral RLM. The child presented with renal insufficiency. [3] In a publishment by Sanchez *et al*., another case of unilateral RLM was reported in a 4-month-old female infant as an incidental search. [4]In 2012, a pediatric patient with bilateral RLM was reported by Vasquez *et al*. and reinforced the significance of using radiological conclusions to identify the pathology

of RLM. [5]The etiopathogenesis of RLM is categorized by ectatic endothelial-lined spaces within the cortex with no tubular or glomerular abnormalities and with sparing of the medulla, unclear etiopathogenesis (Arora, 2016). [6] The literature search reveals that RLM is rare, and there is a need to focus on evidence-based clinical conclusions to examine and identify the pathology of RLM.

Approximately, of all lymphangiomas, RLM accounts for 1%. Rarely, cases of RLM are reported as a condition of lymphatic abnormality. The primary factor that causes lymphatic deformity is the poor communication between peri-pelvic and peri-renal lymphatic channels with the primary lymphatics. Occasionally, cases of RLM have been reported in children and adults of both sexes. RLM can be bilateral, unilateral, diffuse, or focal. RSL is asymptomatic and may not reveal specific signs and symptoms [5]. The failure of drainage by the lymphatic ducts into the main retroperitoneal lymphatics leads to the formation of multilocular or unilocular masses in pelvic sinuses or perinephric spaces. RLM is an incidental condition and does not present specific symptoms like abdominal distension, flank pains, altered renal function, hypertension, and hematuria. Pregnancy aggravates the symptoms of RLM. Therefore, RSL is an asymptomatic condition that is incidental and its diagnosis requires imaging conclusions to avoid differential diagnosis. RLM is a very infrequent benign lymphatic disorder. RLM is often misdiagnosed for other disorders such as cystic renal masses, usually polycystic kidneys. Although parenthetically found in different cases, RLM is a probable cause of renal failure and hypertension in undiagnosed victims [6]. The etiology of RLM is unclear and results from the formation of cystic masses and dilatation of perinephric lymphatic channels. The etiopathogenesis of RLM is categorized by ectatic endothelial-lined spaces within the cortex with no tubular or glomerular abnormalities and with sparing of the medulla, unclear etiopathogenesis. In the renal sinus, lymphatic channels connect the perirenal tissues and the renal capsule the kidney's lymphatic system. Developmental malformation in the above-mentioned channels is one of the hypotheses presumed to describe the etiopathogenesis of RLM. The other hypothesis presumes the inflammation of the renal perinephric leads to the dilatation and obstruction of lymphatics.

In most of the reported cases, victims of RSL are asymptomatic and their detection is an incidental finding for unrelated imaging studies carried out. Weight loss, flank pain, abdominal pain, fatigue, and haematuria are commonly presented signs and symptoms in patients with symptomatic cases. However, advanced cases of RSL manifest with renal insufficiency and ascites. Usual complications are hypertension, renal vein thrombosis, intracystic hemorrhage, superimposed infection, and obstructive uropathy.

Asymptomatic cases require imaging studies to produce striking conclusions to the diagnosis. Unrelated imaging studies are carried out to detect an incidental search. Imaging studies and features encompass various diagnostic modalities such as CT, US, and magnetic resonance imaging (MRI) to facilitate and allow for an evidence-based diagnosis. However, the findings and conclusions of the imaging studies may be more scrutinized with chylous fluid collected from perirenal fluid for a confident clinical diagnosis. The extent and distribution of the involved lymphatics confirm the imaging studies and features [5]. Ultrasound features and studies reveal enlarged or normal size of kidneys without cortico-medullary differentiation. Again, the ultrasound features disclose increased parenchymal echo texture. In the peri-pelvic or perinephric regions, the ultrasound studies show multi-septated thin-walled collections of fluid. Cases of RLM that involve small intra-renal, lymphatics are disclosed as mild diffuse increased size of the kidney without cystic space. Again, the cases reveal echogenic renal parenchyma that is diffuse with poor differentiation of corticomedullary on the hyperintense signal in the cortex & hypointense signal in the medulla that presented corticomedullary inversion.

However, peripelvic cysts can be traced in RSL cases where lymphatics are involved in the renal sinus. The peripelvic cysts are better viewed on US imaging features since they are enlarged. The presence of enlarged cysts can lead to blockage of the collecting system, a factor that leads to hydronephrosis. Septated perinephric fluid collection is caused by capsular lymphatic dilatation.

The rarity of RSL leads to the existence of differential diagnoses. The common differential diagnoses include polycystic kidney disease, hydronephrosis, urinomas, abscesses, multiloculated cystic nephroma, nephroblastomatosis, and lymphoma [7]. Nephroblastomatosis and lymphoma indicate characteristics of tender and soft tissue masses on imaging modalities. However, on imaging features, the two above-mentioned differential diagnoses are enhanced on MRI and CT and display internal vascularity on US imaging studies. Polycystic kidney disease shows a collection of non-communicating cysts that are spread in renal parenchyma without cysts like in other organs such as the pancreas, liver, and spleen. Hydronephrosis displays a collecting system that is distended with the presence or absence of dilated ureter. Multiloculated cystic nephroma is categorized by a focal multiseptated mass of cysts that arise from the kidney with a clawed shape that is adjacent to the renal parenchyma. Abscesses and urinomas can be distinguished on a proper basis by imaging history of a thick wall that enhances contrast eructation due to delayed phases in urinoma.

CONCLUSION:

RLM is an elusive condition because of its undetermined etiopathogenesis. Although parenthetically found in different cases, RLM is a probable cause of renal failure and hypertension in undiagnosed victims. The etiology of RLM is unclear and results from the formation of cystic masses and dilatation of perinephric lymphatic channels. The etiopathogenesis of RLM is categorized by ectatic endothelial-lined spaces within the cortex with no tubular or glomerular abnormalities and

with sparing of the medulla, unclear etiopathogenesis. Therefore, there is a need to emphasize the requirement for radiological conclusions for identifying the etiopathogenesis of RLM.

IMAGES:

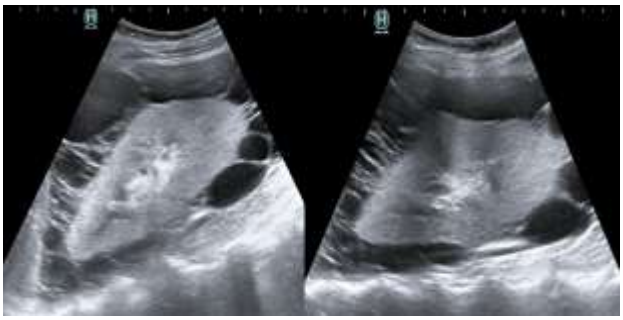

Figure 1: Axial grey scale image of bilateral kidneys showing normal sized kidneys with increase cortical echogenicity and loss of cortico-medullary junction. Multiseptated anechoic collections in perinephric region of maximum width 30mm.No evidence of calculus or hydronephrosis on either side.

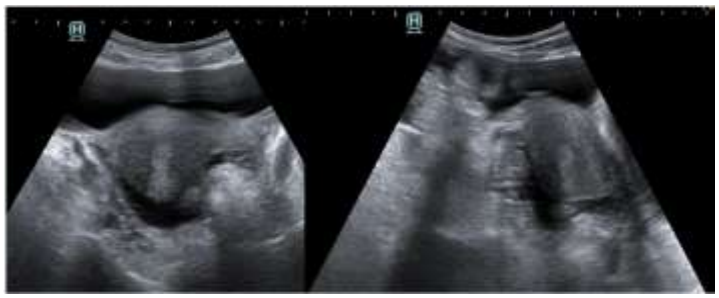

Figure 2: Axial grey scale ultrasound image showing free fluid seen in pelvis suggestive of Ascites.

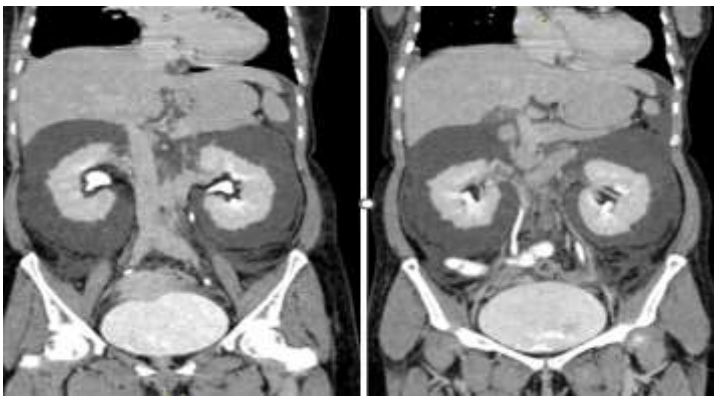

Figure 3: Coronal Contrast enhanced Computed Tomography Scan showing multiseptated collections with fluid density of 0-10 HU inperinephric regions of both kidneys. No evidence of soft tissue or fat density mass lesion seen. Both kidneys show normal parenchymal enhancement and contrast excretion with no leakage of contrast within the perinephric collections. Both ureters were normal in course and calibre.

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