Kumar et al., J Nephrol Ther 2018, 8:5
DOI: 10.4172/2161-0959.1000318

Journal of Nephrology & Therapeutics

Case Report Open Access

Case Report: Renal Lyphangectesia with Nephritic Syndrome

Kishore Kumar*, Shakeel Khan, Sohail Tanveer and Ahmed Shamim Khan

Department of Nephrology, Pakistan Institute of Medical Sciences, Islamabad, Pakistan

***Corresponding author:** Kishore Kumar, Department of Nephrology, Pakistan Institute of Medical Sciences, Islamabad, Pakistan, Tel: +92 51 9261170; E-mail: smciankishore@gmail.com

Received Date: July 17, 2018; **Accepted Date:** September 1, 2018; **Published Date:** September 9, 2018

Abstract

A very rare condition of lymphatic vessels called lymphangectesia is under study since mid of nineteenth century after the association with some vital organ abnormality lead to increased mortality and morbidity. This is an interesting fact that made clinicians to work more in this field. We also have diagnosed a rare suspected case of perirenal lymphangectesia with nephrotic syndrome when a young 19 years old boy admitted to us with repeated history of perirenal fluid collection for 4 years, a chylous fluid rich in lymphocytes on chemical examination with progressive worsening of renal failure and proteinuria. The radiographic studies done were ultrasonography of abdomen, followed by CT scan on first presentation and MRI with contrast on 3rd repeated presentation; show the perirenal fluid rim with intact kidneys. On the third presentation we have done some extensive workup, searched in literature and then after all parameters calculated we reached at a diagnosis of perirenal lymphangectesia with nephrotic syndrome progressed to chronic renal disease.

Keywords: Perirenal fluid; Lymphangectesia; Nephrotic syndrome

Introduction

Lymphangectesia is rare disease of abnormal lymphatic vessels dilatation results in lymphatic blockade or lymphatic accumulation of various organs, described first in middle of 19th century by Rudolf Virchow and it is caused by abnormal dilatation of lymphatic vessels [1]. It is either congenital (present by birth) or acquired (present in children or adults). Congenital variant usually presents with birth or in early childhood caused by abnormal lymph vessels formation associated with severe complications, mostly related to respiratory failure, other mostly involve are liver and rarely kidneys. Acquired variant presents in child hood or adulthood, it is due to secondary dilatation of lymphatics by either blockage or damage of existing lymphatics most commonly after radiation or extensive surgical interventions, and involved organs are skin, intestine and others. As result of new collaterals there are usually abnormal accumulations of lymph in these dilated lymphatics and represent as cystic swellings in various organs.

Lymphangectesia involving kidneys is again a very rare only 1% of all and if it is associated with renal impairment then it makes a case very special because till date only few of cases of renal lyphangectesia with renal impairment in the form of nephrotic range proteinuria and chronic kidney disease have been reported [2,3]. Most common type of renal lymphangectesia is peri renal (extra-renal) others also present as peri pelvic (intra-renal). Peri-renal (extra-renal) represent as repeated perirenal fluid collection in the form of a thin fluid rim around the kidney which represent as page kidney usually misdiagnosed as extra renal mass or hydronephrosis. Peri pelvic (intra-renal) lymphangectesia represent as cystic fluid collection which again miss diagnosed as polycystic kidneys [4].

Here we are going to present a case of perirenal lyphangectesia with nephrotic range proteinuria and renal failure as case presentation.

Case Report

A 19 years old male resident of XYZ , no any known comorbidity, presented with complains bilateral flank pain with dull nature and progressed with time over 10 days resulted burning maturation and low grade fever of 98°F and increasing facial puffiness. On evaluation there was more than 4 years history of repeated perirenal collection with symptoms of dullach, lowgrade fever and mild pedal edema. There was no history of hematuria, renal stone and. He underwent 2 times perirenal fluid aspiration followed by twice antibiotic treatment with Ceftrixone 2 g for 14 days+Amikacin 25 mg for 7 days along with NSAIDS and Furesimide 20 mg, once completed ATT 2 years back. On further evaluation it was noted the he underwent radiological evaluation every time with ultrasonography pre and post fluid aspiration. And fluid work up showed only increased lymphocytes count.

On detailed examination there was Blood Pressure of 140/90 mmHg, Pulse of 85b/min, Respiratory Rate of 18/min and moderate pitting pedal edema with mobile, soft, non-tender swelling in bilateral renal area.

Biochemical lab results shows progressive increase in serum creatinine from 1.23 at first presentation to 2.6 on last presentation along with progressive decrease in Hb from 12.4 to 10.8 and serum cholesterol of 326 but other parameters were almost within normal limits (measurement unit was mg/dl). Urinary evaluation also showed increasing protein urea 1+ on first presentation to 3+ and 24 hour protein quantification was <200 mg on first presentation to 5.5 g on last presentation.

Serological workup was as serum complement levels were as C3 90{80-185} and C4 15{10-53} and ANA was negative.

Blood and urine cultures were negative and both blood+fluid PCR DNA MTB was also negative.

Radiological evaluation started with ultrasonography showed bilateral increased renal size;

- Right: 11.9 X 5.0
- Left: 10.2X 4.9

Then done CT scan whole abdomen which resulted as:

- Fluid attenuating area filling entire perirenal space on right side noted causing indentation of lateral border of right kidney.
- Sickle shaped fluid attenuating area is noted in left perirenal space. Perirenal stranding is appreciated.
- Suspicion was lymphangectasia/hemorrhage/abscess.

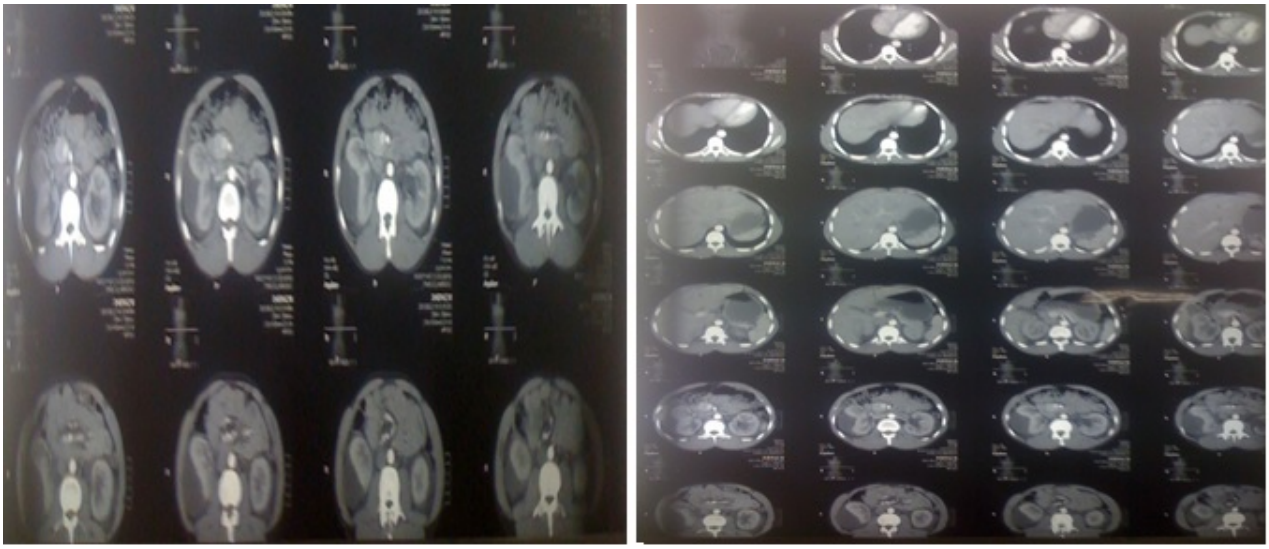

Figure 1: CT scan abdomen with special section taken of renal area (Transverse and Longitudinal).

MRI done rule out any possible mass growth which resulted as;

- A large abnormal signal intensity area is noted in the right perirenal region surrounding the kidney at its posteriolateral aspect and pushing it anteromedially. It appears hypo intense on T1WS and hyper intense on T2WSand shows few internal septations. It measures approximately 14.0 × 6.0 × 12.0 cm in size. No abnormal enhancement is noted. No mass noted. The right kidney measures 11.5 × 3.0 cm and the left kidney measures 12.5 × 2.5 cm (Figure 1).
- Suspicion was Urinoma/Acute hemorrhage?

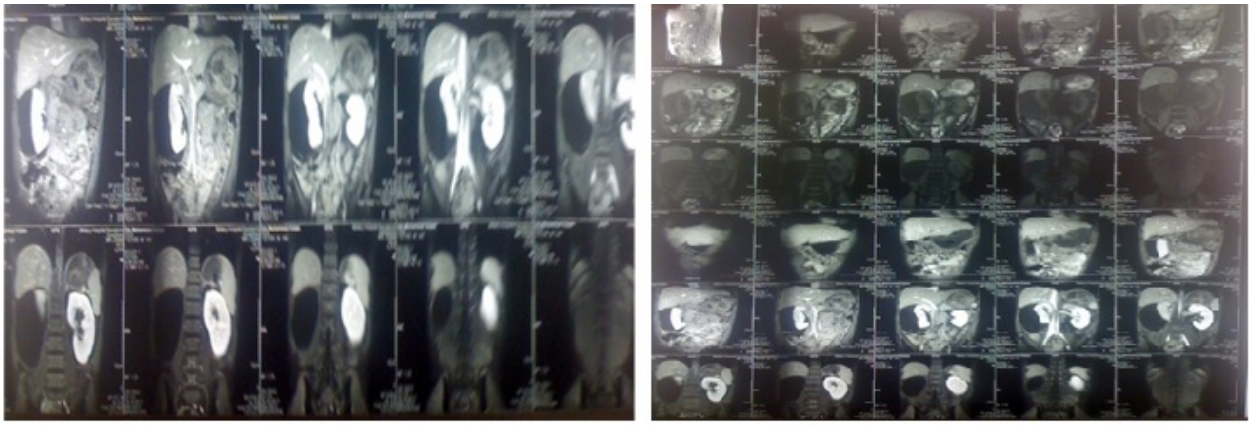

Figure 2: MRI abdomen with section pointing renal collection (t1ws and t2ws).

Fluid cytology was done which sowed increased levels of lymphocytes (due to limited facility detailed examination was not possible).

On the basis of repeated perirenal fluid collection and available laboratory we labeled it as Renal Lymphangiectasia/Sub Capsular Fluid Collection with Nephrotic Syndrome (Figure 2).

Discussion

Lymphangectesia is rare disease also known as abnormal lymphatics, described first in middle of 19th century by Rudolf Virchow and it is caused by abnormal dilatation of lymphatic vessels1. It's either congenital or acquired. Congenital variant usually presents with birth or in early childhood with severe complications related to respiratory failure a most, is both primary and secondary; mostly involve lungs, liver and more rare kidneys. Acquired variant presents in child hood or adulthood, it is due to secondary dilatation of lymphatics by either blockage or damage of existing lymphatics most commonly after radiation or extensive surgical interventions, and involved organs are skin, intestine and others. As result of new collaterals there are usually abnormal accumulations of lymph in these dilated lymphatics and represent as cystic swellings in various organs.

Renal lymphangectesia is very rare form and only 1% of all lymphangectesia patients have renal lymphangectesia and again only few of them are actually diagnosed separately because of its radiographic appearance, which is similar to cystic lesions in kidney [2,3]. Renal lymphangectesia characterized by abnormal and ectatic lymphatic vessels within and around the kidneys [5]. There may be dilation of any one or all perirenal, peripelvic and intrarenal lymphatic vessels. Peripelvic dilatations usually represents as pelvic cysts called as intrarenal lymphangectesia and prirenal dilatation results in fluid collection in perirenal area called as extrarenal lymphangectesia. Although only a few of cases are reported worldwide of renal lymphangectesia most of them are associated with renal vein thrombosis but its association of nephritic syndrome is very rare that only three to four cases are diagnosed with it [4,6].

Renal lymphangectesia with nephrotic syndrome is very rare presents more commonly in children because a few can grow to adulthood. As only a few have first presentation in adult hood, reason may be misdiagnosis of disease or overlook of this rare variant as a cause of nephrotic syndrome. These patients can present with repeated preirenl fluid collections without any significant pathological cause mostly after renal vein thrombosis, uncontrolled hypertension, radiation exposure or obstruction of venous and lymphatics during normal pregnancy related hydronephrosis. These patients can also present first time with mild to severe flank pain, hematuria, and renal insufficiency or repeated UTIs.

Diagnosis is renal lymphangectesia usually clinical and make only after suspicion in patients with repeated symptoms of renal impairment and nephrotic range protienurea with radiographic presence of perirenal fluid collection [7-9]. Confirmation is usually made after fluid analysis, as chylous appearance with increased lymphocytes without any infective cause. Renal biopsy is usually not performed due to chances of chylous or lymphatic fluid leakage [10,11].

There is no any specific treatment for renal lymphangectesia and usually it's not necessary to treat every case until there is associated complication in the form of protienurea or progressive renal impairment. If required it is usually symptomatic with diuretics and anti-hypertensive and in complicated cases, percutaneous drainage and marsupialization [12] and if unilateral then nephrectomy is recommended.

Conclusion

Lyphangectesia is always suspected in patients with repeated perirenal fluid collection with progressive renal impairment without any available observed cause.

References

1. Ferreira CR, Sibre V, Schultz R, de Melo AM, Ibidi SM, et al. (2015) Congenital generalized lymphangiectasia: A rare developmental disorder for non-immune fetal hydrops. Autops Case Rep 5: 27-33.
2. Wani NA, Mir F, Gojwari T, Qureshi UA, Ahmad R (2010) Subscapular fluid collection: An unusual manifestation of nephrotic syndrome. Am J Kidney Dis 56: 181-184.
3. Riehl J, Schmitt H, Schäfer L, Schneider B, Sieberth HG (1997) Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrol Dial Transplant 12: 1701-1703.
4. Pandya VK, Shah MK, Gandhi SP (2016) Bilateral Renal Lymphangiectasia. J Clin Diagn Res 10: TD01-TD02.
5. Pianezza ML, Mokhtassi A, Wu L, D'A Honey RJ. (2006) Case report: renal lymphangiectasia. Can J Urol 13: 3204-3207.
6. Merguerian PA, Sargent SK, Dunn JL (1995) Unilateral lymphangiectasis of the kidney: an unusual cause of renal enlargement in an infant. J Urol 153: 447-449.
7. 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.
8. Varela JR, Bargiela A, Requejo I, Fernandez R, Darriba M, et al. (1998) Bilateral renal lymphangiomatosis: US and CT findings. Eur Radiol 8: 230-231.
9. Llorente JG, García AD, Sacristan JS, Chicharro GN (2002) Renal lymphangiectasia: radiologic diagnosis and evolution. Abdom Imaging 27: 637-639.
10. Kutcher R, Mahadevia P, Nussbaum MK, Rosenblatt R, Freed S (1987) Renal peripelvic multicystic lymphangiectasia. Urol 30: 177-179.
11. Ashraf K, Raza SS, Ashraf O (2007) Renal lymphangiectasia. Br J Radiol 80: e117-e118.
12. Schwarz A, Lenz T, Klaen R, Offermann G, Fiedler U, et al. (1993) Hygroma renale: pararenal lymphaticcysts associated with renindependent hypertension (Pagekidney). Case report on bilateral cysts and successful therapy by marsupialization. J Urol 150: 953-957.