Case Report: Renal Lypanchectesia with Nephritic Syndrome

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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.

Case Report

A 19 years old male resident of XYZ, no any known comorbidity, presented with complaints 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 dull ach, 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.

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 lymphangectesia 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 hydropnephrosis. 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 lymphangectesia with nephrotic range proteinuria and renal failure as case presentation.
Radiological evaluation started with ultrasonography showed bilateral increased renal size:
- Right: 11.9 X 5.0
- Left: 10.2 X 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.

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 T2WS and shows few internal septations. It measures approximately 14.0 x 6.0 x 12.0 cm in size. No abnormal enhancement is noted. No mass noted. The right kidney measures 11.5 x 3.0 cm and the left kidney measures 12.5 x 2.5 cm (Figure 1).
- Suspicion was Urinoma/Acute hemorrhage?

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 Lymphangectasia/Sub Capsular Fluid Collection with Nephrotic Syndrome (Figure 2).
Discussion

Lymphangectesia is a rare disease also known as abnormal lymphatics, described first in the middle of 19th century by Rudolf Virchow and it is caused by abnormal dilatation of lymphatic vessels. It's either congenital or acquired. Congenital variant usually presents with birth or in early childhood with severe complications related to respiratory failure. Commonly it 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 a rare clinical presentation of lymphangectesia and it is mostly confined to children. It is usually 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 a rare clinical presentation of 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 dilatation of any one or all perirenal, peripelvic and intrarenal lymphatic vessels. Peripelvic dilatations usually present 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 a rare clinical presentation of lymphangectesia and it is mostly confined to children. It is usually 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]. 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 proteinuria 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

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

References