



## **BILATERAL PERINEPHRIC LYMPHANGIOMATOSIS –A CASE REPORT**

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<p><b>Article Info</b> <i>Received 15/08/2015</i> <i>Revised 27/08/2015</i> <i>Accepted 02/09/2015</i></p> <p><b>Key words:</b> Perinephric Lymphangiomas, CT scan, Kidney.</p>	<p><b>ABSTRACT</b> Perinephric lymphangiomas is a rare benign malformation of the lymphatic system. It may be confused with various forms of renal cystic diseases and urinomas. Characteristic USG and CT scan findings help in diagnosis. We present a case of bilateral perinephric lymphangiomas in a 32 year old female patient.</p>
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### **INTRODUCTION**

Perinephric lymphangiomas (PL) is a rare benign renal disorder that has been reported in both children and adults [1,2]. In this disease, the perirenal space and renal sinuses are filled by unilocular or multiseptated cystic masses that may demonstrate extrarenal extension [3,4]. This is due to failure of renal lymphatic drainage into the retroperitoneal lymphatics, subsequently causing dilatation of the ducts and formation of cystic spaces in the perirenal and renal sinus regions.

Biochemical analysis of percutaneous needle aspiration of chylous fluid is the mainstay of diagnosis. However, USG and CT scan findings are also very characteristic that allow definitive diagnosis. We report a case of bilateral perinephric lymphangiomas in a 32 year old female patient. The clinical features, diagnosis and characteristic imaging findings are reviewed.

### **CASE REPORT**

A 32 year old female patient presented with vague abdominal pain and loss of appetite since 2 months. There was no significant past history or family history. On physical examination, she was well built and nourished with normal blood pressure. Per abdomen examination was unremarkable. Complete urine examination and blood biochemical investigations were within normal limits. Ultrasound evaluation of abdomen was done at the department of radiodiagnosis showed bilateral septated, multiloculated, symmetrical perirenal collections with internal echoes. Both kidneys were normal in size and corticomedullary differentiation was maintained.

CT scan of the abdomen showed bilateral hypodense collections with attenuation varying from 5-15v HU in the perirenal and peripelvic regions. Normal corticomedullary differentiation was seen. Normal enhancement of the cortex was seen. Delayed scans failed to reveal contrast extravasation into perinephric space or



any change in attenuation of the fluid thus ruling out communication with pelvicalyceal system.

An ultrasound guided aspiration of the fluid was done. The fluid was turbid and pale yellow in colour.

Microscopic examination of the fluid revealed lymphocytes confirmed the diagnosis of bilateral perinephric lymphangiomatosis.

**Figure 1. Ultrasound image of kidney showing perinephric hypoechoic collections with thin septations**



**Figure 2. Axial plain CT image showing perinephric fluid density collections. Underlying kidney is normal**



**Figure 3. Axial CECT scan showing non enhancing fluid collection in bilateral perinephric regions**



## DISCUSSION AND CONCLUSION

PL is an extremely rare developmental disorder of lymphatic system surrounding the kidneys. The frequency of lymphangiomas are 75% in head and neck, 20% in axillary region and 5% at other less common sites [5].

Retroperitoneal / renal lymphangiomas account for approximately 1% of all lymphangiomas [6]. This is due to the failure of drainage of lymphatics into retroperitoneal lymphatic system [7]. The lymphatic channels dilate to form uni/ multilocular cystic masses and may be seen unilaterally or bilaterally.

It may be asymptomatic or present with nonspecific symptoms like flank pain, abdominal distension, hematuria, fever, hypertension [7-9]. This condition is found in children and adults [10, 11]. Familial association has been described [12]. Complications of undiagnosed/untreated cases include hematuria, ascites, hypertension, renal vein thrombosis, and impairment of renal function.

The diagnosis of PL can be confirmed with aspiration of chylous fluid from perinephric fluid collections [13]. However, with characteristic imaging findings a definite diagnosis of perinephric lymphangiomatosis can be made.

USG shows multiloculated, cystic perirenal and arapelvic collections with or without thin septae. The kidneys may be normal or reduced in size [3].

Prior to the advent of CT scan the diagnosis was usually made at the time of exploratory laparotomy or after nephrectomy. On CT scan, this appears as well defined hypodense collections in the peripelvic or perinephric space with or without demonstrable septations with normal renal parenchyma. No abnormal enhancement is seen within the collections [3, 4]. On MRI, multiple hyperintense collections with septae on T2W images with reversal of the corticomedullary intensity, which is due to an anatomic variation in the lymphatics is seen [13]. Treatment is not usually necessary for asymptomatic, incidentally diagnosed cases. Complicated cases may be



treated with nephrectomy, percutaneous drainage or marsupialisation [14].

## CONCLUSION

Perinephric lymphangiomas can present with a number of nonspecific symptoms. However, it can present as an incidental finding for which nothing needs to be done till patient remains asymptomatic. The close differential

diagnosis is perinephric urinoma which can be ruled out with a contrast enhanced CT scan with delayed cuts. However, biochemical examination of aspirated fluid is also vital in arriving at a definite diagnosis.

**ACKNOWLEDGEMENT: NIL**

**CONFLICT OF INTEREST: NIL**

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