Radiographical resolution of renal lymphangiomatosis following cardiac transplantation

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Renal lymphangiomatosis is a disease characterized by abnormal formation of perirenal lymphatic vessels that fail to communicate with other retroperitoneal lymphatics; as a result, perirenal lymphatics dilate and form cysts. While typically an asymptomatic incidental finding, renal lymphangiomatosis rarely presents as flank or abdominal pain, ascites, impaired renal function, hypertension, hematuria, or proteinuria. Here we present the first known case of renal lymphangiomatosis found to spontaneously resolve following cardiac transplantation.

Key Words: renal lymphangiomatosis, perirenal cyst, cardiac transplant, lymphatic drainage

Introduction

Renal lymphangiomatosis, also known as a benign cystic lymphangioma, or renal lymphangiectasia, was first described in 1889 and has also been referred to as renal hygroma, peripelvic lymphangectasia, renal peripelvic lymphangectasia, retroperitoneal lymphangectasia, and renal lymphangectasia which account for less than 1% of all lymphangiomas within an age range of 8-74 years and without gender predilection.[1] It is regarded as an extremely rare congenital developmental malformation of the intrarenal or perirenal lymphatic tissue.[2] This developmental malformation is believed to be the result of failure of developing lymphatic tissue to establish normal communications with the rest of the lymphatic system resulting in abnormal cystic dilations.[3] This disorder is most often discovered incidentally during routine clinical imaging which when other disease manifestations are ruled out, is sufficient to make the diagnosis.[1] Rarely, renal lymphangiomatosis can present symptomatically secondary to extrinsic compression.[4] In this case, we present the first known report of renal lymphangiomatosis to spontaneously resolve following heart transplantation.

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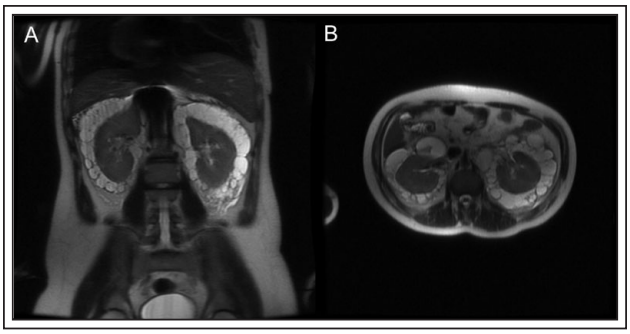

Figure 1. Pre-transplant non contrasted coronal **(A)** and axial **(B)** T2WI fat-sat MRI images.

Case report

A 21-year-old Caucasian male with severe complex congenital heart disease secondary to heterotaxy syndrome with dextrocardia, double outlet right ventricle, atrioventricular septal defect, and congenital pulmonary stenosis along with malpositioned great arteries presented for urologic consultation for asymptomatic bilateral perinephric soft tissue swelling. The finding was discovered incidentally upon routine imaging prior to being listed for heart transplantation secondary to his heart failure with an ejection fraction (EF) of 10.

On physical exam, the patient appears normal, but with perioral cyanosis and impressive clubbing of the digits. Vital signs demonstrated a normotensive blood pressure of 107/80 with a heart rate of 115 and a pulse oxygen saturation of 81%. Abdominal exam revealed a soft non-distended abdomen with no palpable tenderness or organomegaly, but an obvious apical pulse in the fifth right intercostal space on his chest. The remainder of his systemic examination including genitourinary exam was otherwise benign. Laboratory analysis was significant only for severe polycythemia with a hemoglobin level of 21.4. Renal function was

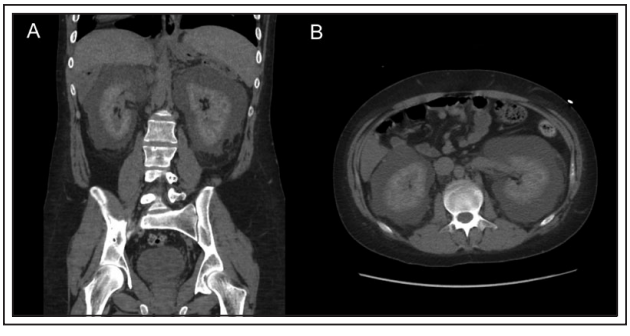

Figure 2. Pre-transplant non contrasted coronal **(A)** and axial **(B)** CT images.

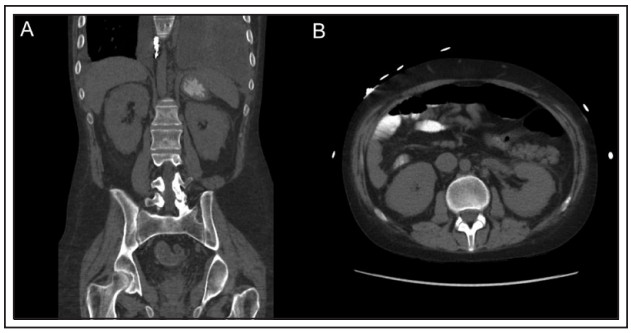

Figure 3. Post-transplant non contrasted coronal **(A)** and axial **(B)** CT images.

normal with, serum creatinine of 1.1, blood urea nitrogen (BUN) of 25, and glomerular filtration rate (GFR) > 60. Radiographs available for review included ultrasound (U/S), computed tomography (CT), and magnetic resonance imaging (MRI).

Ultrasonography of the kidneys revealed an 11.8 cm right kidney and a left kidney 13.3 cm with no collecting system dilatation. There were numerous bilateral perinephric cysts suggestive of renal lymphangiomatosis (image not shown). Non-contrasted CT imaging demonstrated situs ambiguous of the abdominal organs, a retroaortic left renal vein, and bilateral perirenal water attenuation of the tissues surrounding the kidneys, Figure 1a and 1b. Furthermore, both kidneys show heterogeneously increased cortical density concerning for medical renal parenchymal disease. Non contrasted coronal T2-weighted MRI imaging, Figure 2a and 2b, exhibited bilateral normal size and position of the kidneys with bilateral poor corticomedulllary differentiation and innumerable cystic lesions involving the perinephric spaces. There was no evidence of parenchymal renal cysts or collecting system dilation. Universally, the imaging studies were all consistent with bilateral renal lymphangiomatosis. Several months later, a post-cardiac transplantation non-contrast abdominal CT scan was performed for unrelated testing. Strikingly, the CT scan revealed significant reduction in the perirenal cysts compared to the prior imaging, Figure 3a and 3b.

Discussion

Renal lymphangiomatosis is an extremely rare developmental disorder affecting normal drainage of the lymphatic system surrounding the kidney. It can form peripelvic and perirenal unilocular/multilocular cystic masses on one or both kidneys.[5] There is scant literature concerning this diagnosis over the past quarter century.[6] Here we present the only known case of renal lympangiomatosis where the cystic dilations

decreased in size following cardiac transplantation.

Renal lymphangiomatosis is largely diagnosed incidentally from imaging studies. On rare occurrences, symptomatic individuals present with flank or abdominal pain, ascites, impaired renal function, hypertension, hematuria, or proteinuria. Hypertension is thought to be secondary to renal vessel extrinsic compression by the lymphangioma.[4] If hematuria or proteinuria are observed, this is felt to be secondary to chyluria rather than true medical disease.[2]

Imaging studies reveal several common findings. U/S shows anechoic regions with sharply defined septae. Hemorrhage and debris that may be present within the unilocular or multilocular cysts appear as fine echoes. Cysts found on CT or MRI scans have similar density to water. On MRI, cysts appear hypointense on T1 weighted images and hyperintense on T2 weighted images.[7]

The differential diagnosis for renal lymphangiomatosis includes: polycystic kidney disease, urinoma, hydronephrosis, renal lymphoma with perirenal involvement, tuberous sclerosis, von Hippel-Lindau disease, and nephroblastomatosis. Other diagnostic modalities such as fine needle aspiration of chylous material, lymphoscintigraphy, histological staining and analysis are available but rarely necessary.[6,8] When asymptomatic, the appropriate management is observation with serial radiographs. When symptomatic treatment is required this can include injection of sclerosing agents (ethanol, bleomycin), percutaneous drainage, or cyst marsupialization.[1]

In our patient, we observed him with periodic imaging due to his complete lack of symptoms. Follow up images after cardiac transplantation stunningly revealed decreased cystic size around both kidneys. This finding points to perirenal cysts being dynamic structures rather than static, inactive fluid-filled sacs. This finding suggests that overall fluid load may play a pivotal role in the formation of the cysts. For example, increased fluid load in pregnancy may cause an increase in cystic size.[9] Improvement in cardiac function (pre-transplant EF: 10 to post-transplant EF: 57) may have improved total body fluid equilibrium that resulted in such drastic changes. Poor ejection fraction in general leads to increased fluid retention in interstitial tissue. This can lead to compensatory dilation of lymphatic vessels. Therefore, these abnormal perirenal cysts in renal lympangiomatosis may act as a reservoir to store the excess fluid. Following cardiac transplantation, the improved ejection fraction likely decreased overall capillary filtration into interstitial tissue preventing an overload of fluid into the lymphatics. Improved arterial pulsations might also aid in improved lymphatic flow.[10] As a result, overall drainage through lymphatic vessels likely improved and decreased cystic size. Understanding the mechanism of abnormal lymphatic flow may lend insight into the pathophysiology of symptomatic individuals affected by renal lymphangiomatosis. □

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