Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis

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Introduction

Lymphangiectasia is regarded as a developmental malformation of the lymphatic system, effecting generalized or localized dilatation of the lymphatics. In retroperitoneal lymphangiectasia these changes occur in the kidneys and the retroperitoneal tissue. If the smaller intrarenal lymphatics become dilated, considerable enlargement of the kidneys may occur. Dilatation of larger peripelvic and perirenal lymph channels can be visualized as cystic masses in ultrasound and computer tomographic scans. In these cases it may be difficult to differentiate between lymphangiectasia and autosomal dominant polycystic kidney disease (ADPKD) or autosomal recessive polycystic kidney disease (ARPKD). In order to establish the diagnosis of retroperitoneal lymphangiectasia, additional liquid structures that coat the retroperitoneal vessels are suggestive of the diagnosis [1].

Complications of retroperitoneal lymphangiectasia have been reported only in rare cases. Blood pressure may be increased and haemorrhagic complications may occur. In the current report we describe a case of retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis.

Case report

In 1985, a 22-year-old male presented at our clinic for evaluation of pain in the right flank region. There was no evidence of hereditary kidney disease in his family history. Haematuria had been found during the medical examination for entry to military service. The patient’s blood pressure was 130/85 mmHg. His renal function was normal and there were no signs of proteinuria.

The ultrasound examinations and computed tomography (CT) revealed multiple bilateral peripelvic and perirenal cysts, enlargement of the right and shrinkage of the left kidney. A fluid specimen from these perirenal cysts was aspirated via exploratory puncture and identified as lymph fluid.

Angiography revealed renal arteries of normal calibre, while none of the main renal veins on either side could be visualized during the venous phase. On the right serpinginous veins in the perinephric space were diagnosed as collateral vessels.

Another follow-up was performed in 1996. The patient was asymptomatic and there were no signs of arterial hypertension. The laboratory tests performed during the current investigation revealed the following values: white blood cells count 6.5 × 10^9 g/l, haemoglobin 176 g/l, haematocrit 50%, platelet count 221 × 10^9 g/l, serum sodium 137 mmol/l, potassium 4.1 mmol/l, calcium 2.5 mmol/l, phosphate 0.65 mmol/l, total proteins 75 g/l, urea 7.2 mmol/l, serum creatinine 105 μmol/l, endogenous creatinine clearance 131 ml/min, C-reactive protein <5 mg/l, Quick 90%, partial thromboplastin time 27 s. Urine specimens contained no leukocytes or erythrocytes. The urinary protein extraction was 32 mg/24 h; the blood gas analysis was normal.

Immunological tests (ANA, Anti-dDNA, AMA, ANCA) were all negative. Antithrombin III, protein C, and protein S were within normal range. The lupus anticoagulant test and activated protein C resistance (APC-R) test were negative. The serum protein electrophoresis was normal.

Ultrasound and CT findings (Figures 1 and 2) remained unchanged from those established in 1985. MAG-3 scintigraphy revealed slightly delayed tubular secretion but normal perfusion of the enlarged right kidney. Perfusion of the left kidney was greatly reduced. The total tubular clearance was decreased to 224 ml/min (normal 443–664 ml/min), and the right kidney provided 95% of the total clearance.

In colour-coded Doppler sonography, the main renal veins could not be visualized, and communication between the right intrarenal veins and abundant serpinginous collateral vessels in the perinephric space was
Fig. 1. CT scan demonstrating liquid masses in the renal sinus and perirenal spaces. Enlargement of the right kidney and shrinkage of the left kidney can also be observed.

Fig. 2. Contrast CT scan of the abdomen with enhancement of the right kidney. The left renal parenchyma is shrunken. Multiple venous collaterals can be seen in the right perirenal space.

demonstrated, indicating a gradual onset of the renal vein thrombosis. No collateral veins were visible on the left side.

Summing up the data, we found that this patient had a retroperitoneal lymphangiectasia affecting both kidneys, associated with bilateral renal vein thrombosis. Peripelvic and retroperitoneal venous collaterals had developed in the right retroperitoneum, resulting in a normal renal function of the right kidney. Atrophy of the left kidney developed in the absence of collateral veins. In the 11 years of follow-up, renal function has remained stable and no further thrombotic events occurred.

Discussion

Dilatation of the lymphatics located in the kidneys and the retroperitoneal tissue have been reported as renal hygroma [2,3], lymphangiomatosis [1,4,5], and retroperitoneal lymphangiectasia [6–9]. In these cases, peripelvic, intra- and perirenal cysts corresponding to
dilated lymph channels were found. The cause of these changes, which may be uni- or bilateral, is not obstruction, but ectasia of the retroperitoneal lymphatics [10]; therefore the preferred term is retroperitoneal lymphangiectasia.

In the common minor variant, retroperitoneal lymphangiectasia manifests as peripelvic cysts (synonyms: renal peripelvic multicystic lymphangiectasia [7], polycystic disease of the renal sinus [11]). Due to their position in the renal sinus, these cysts are easy to detect by means of ultrasound, and their flat, fanlike shape makes them distinguishable from the more common parenchymal cysts. Although their shape gives them a characteristic appearance, it is not always possible to differentiate them accurately from a dilated collecting system. Lack of communication between the individual cystic components is suggestive of the diagnosis of peripelvic lymphangiectasia.

The major variant of retroperitoneal lymphangiectasia, which affects the entire kidney and retroperitoneal tissue, is an uncommon manifestation that is often associated with significant enlargement of the kidneys. The specific cause of the renal enlargement generally cannot be determined by diagnostic imaging. However, concomitant findings such as the peripelvic, perirenal, or retroperitoneal cysts mentioned above are suggestive of the diagnosis of a renal or retroperitoneal lymphangiectasia. Depending on the patient’s age, polycystic kidney disease, multicystic dysplasia, multicellular cystic nephroma and Wilms’ tumour must be considered in the differential diagnosis [12].

In our patient, retroperitoneal lymphangiectasia was diagnosed by means of imaging techniques and by supplementary analysis of aspirated cystic fluid. Other case reports conclude that the lymphangiectasia can become quite extensive and ultimately lead to ‘decompensated lymphangiectasia’ with massive lymph collections in pre-existing cavities [5]. Although marsupialization of the larger lymphatic cysts can be a useful therapeutic measure in this case, therapeutic intervention is not required in the most cases. No reports of disturbed renal function or urinary flow obstruction were found in the literature.

Renal and retroperitoneal lymphangiectasia may effect a renin-dependent hypertension secondary to mechanical compression and compromised perfusion of the kidney (Page’s kidney) [2,3]. These cases can be effectively treated by nephrectomy, cystectomy, percutaneous drainage, or marsupialization [2]. Additional complications include proteinuria, haematuria, and haemorrhage into lymphatic cysts [5,9,10,13,14].

In our patient, bilateral renal vein thrombosis was found in association with retroperitoneal lymphangiectasia. The findings ruled out the most known causes of bilateral renal vein thrombosis. Although the co-occurrence of retroperitoneal lymphangiectasia and renal vein thrombosis have never been reported before, we must assume that there is a causal connection between the two diseases. Presumably due to its expansile nature, primary (dilatative) lymphangiectasia may pave the way for renal vein thrombosis. In our case, right renal vein thrombosis developed with a gradual onset. Collateral formation resulted in the normal renal function of the right kidney, while an atrophic kidney developed in the availability of venous collaterals on the left.

Secondary (obstructive) lymphangiectasia, on the other hand, is known to occur subsequent to irradiation and surgical intervention [15,16]. However, no reports on the possible development of retroperitoneal lymphangiectasia as a sequel to idiopathic renal vein thrombosis were found in the literature.

References

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