Exceptional Cases

Perirenal fluid collections and monoclonal gammopathy

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Abstract
We report here a case with secondary polycythaemia, monoclonal gammopathy of undetermined significance and renal lymphangiectasis revealed by renal failure. Renal failure was probably linked to renal compression by fluid collections. Renal lymphangiectasis is a rare but has already been described in the literature. In addition, its association with a monoclonal paraprotein and polycythaemia seems to be a new clinical entity recently reported in only one patient.

Keywords: chronic renal failure; monoclonal gammopathy; renal lymphangiectasis

Background
Renal lymphangiectasis is a rare disease, and even more rarely causes renal failure. Two patients have been reported with renal lymphangiectasis associated with polycythaemia [1, 2]. Recently, one patient with monoclonal gammopathy of undetermined significance (MGUS), renal failure related to renal lymphangiectasis, ascites, pleural effusion and polycythaemia has been published as a new clinical entity [3]. Here, we report a new case with MGUS, polycythaemia and renal failure possibly linked to renal compression.

Case report
The patient was a 49-year-old man of Sub-Saharan African origin diagnosed with chronic renal insufficiency on 2006. He had a past history of recurrent bilateral pleural effusion and ascites since 2006. Computed tomography scans performed in 2006 did not show kidney anomalies. He was also diagnosed with polycythaemia and a small IgA lambda paraprotein (2 g/L). Polycythaemia appeared to be secondary. Test for V617F mutation in the Janus kinase 2 gene was negative. Serum erythropoietin level was 78 mIU/mL for a normal expected range between 4 and 16 mIU/mL. He was treated with five therapeutic phlebotomies in 2006. With regard to the IgA lambda paraprotein, skeletal radiographies, bone marrow aspiration and electromyogram were normal. Testing for urinary Bence Jones protein was negative. IgA paraprotein was 5.06 g/L. A new electro-myogram and skeletal magnetic resonance imaging (MRI) were normal. Serum level of vascular epithelial growth factor (VEGF) was 247 pg/mL (<500 pg/mL). Ophthalmologic examination was normal. CT of the abdomen showed compressive perirenal collections of fluid without pyelocaliceal distension (Figure 1). Because of increased serum creatinine, drains were placed in the perirenal collections with the use of CT guidance. Nine hundred millilitres of a clear fluid was drained from the right collection and 800 mL from the left. Fluid analysis revealed a protein level of 2 g/L, lactate dehydrogenase of 22 U/L, amylase of 15 U/L, potassium of 4 mmol/L, cholesterol of 0.03 mmol/L and triglycerides of 0.02 mmol/L. No cells were found and chylomicrons were absent from the samples. Bacteriology was negative with regard to mycobacteria. About 1000 mL a day was drained from each collection for the next 3 days. Renal function improved and serum creatinine was 119 μmol/L on the third day. The drains were then removed. Serum creatinine slowly increased to 130 μmol/L and has remained stable ever since. Renal function impairment due to bilateral perirenal compressive fluid collections was considered because of the compressive CT aspect of the kidneys and the partial improvement after fluid removal. A CT performed 1 month after the drainage showed collections recurrence but the stability of the renal function did not lead us into considering a marsupialization. Moreover, a kidney biopsy was not performed to explore chronic renal dysfunction since urinary sediment had always been inactive and proteinuria testing negative. The prolonged compression of the kidneys may explain the chronic renal dysfunction.

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Renal lymphangiectasis is a rare and benign disease related to renal lymphatics disorder. About 50 cases have been reported [4]. The disease is also known as renal lymphangiomatosis, renal lymphangioma, peripelvic lymphangiectasia, hygroma renale and polycystic disease of the renal sinus. Its aetiology remains unclear. It is speculated that lymphatic drainage of the kidney is blocked due to malformative impaired drainage of the renal sinus lymphatic trunks where lymphatic drainage of the kidney, the renal capsule and perinephretic tissues intercommunicate. It has been diagnosed in adults and children [5] with occasional familial forms [6].

It can be identified by symptoms such as abdominal pain, renin-dependant hypertension [7], ascites, gross or microscopic haematuria, palpable abdominal mass, weight loss or fatigue. Renal dysfunction is less common. Many patients are asymptomatic. It exacerbates during pregnancy [6].

Image findings by CT and ultrasound consist typically in cysts or fluid collections within the renal sinus and the perirenal space. On MRI, cysts are hypointense on T1W images, hyperintense on T2W images and do not enhance. They can be limited only to the renal sinus. Peri-aorto-caval and peripelvic cysts are possible. It is often bilateral [4].

Aspiration shows lymphatic fluid with rare cells and small amounts of protein, chylomicron and triglyceride. When nephrectomy or kidney biopsy are performed, histological findings reported are within dilated spaces in the cortex containing little or no protein or cells. Dilated lymphatics, dilated glomeruli, an increased of small lymphatic vessels density and an extent to the periglomerular space can be seen. Positive predominant staining for D2-40 (a molecule expressed by lymphatic endothelium) of dilated spaces has been described, confirming their lymphatic origin [1, 3, 4].

Treatment is usually conservative using diuretics for ascites and antihypertensive drugs but nephrectomies and marsupializations were performed in symptomatic cases with success [8].

The association of renal lymphangiectasis and secondary polycythaemia has been reported twice [1, 2] but in our case, renal lymphangiectasis appeared after polycythaemia. Our patient also had an IgA lambda paraprotein without evidence for POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, M-protein and Skin abnormalities), in particular a normal serum VEGF level. A very similar case was recently published [1]. However, it concerns an IgG kappa paraprotein. It is very unlikely for polycythaemia to be secondary to renal ischaemia since it appeared before renal fluid collections. Renal lymphangiectasis and pleural effusion may be consequences of the paraprotein by an unknown mechanism, maybe a VEGF-like activity. The aetiology of polycythaemia remains to be elucidated.

In conclusion, the syndrome which associates renal lymphangiectasis, polycythaemia and the presence of a monoclonal paraprotein without myeloma criteria seems to correspond to a new clinical entity. Further studies are needed to determine if the paraprotein has a direct role (through the VEGF pathway or another one) in the genesis of lymphangiectasia.

Conflict of interest statement. None declared.

References


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Fig. 1. Contrast-enhanced coronal (A) and transverse (B) CT slices showing bilateral compressive perirenal fluid collections.