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Case Report

CASE REPORT

Acute renal vein thrombosis and nephrotic syndrome in the setting of retroperitoneal fibrosis

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Abstract

Idiopathic retroperitoneal fibrosis (RPF) is a rare disease that involves non-specific inflammation and fibrosis surrounding the aorta. As a result, RPF is a challenging diagnosis to make; patients often seek medical attention after complications arise, such as nephrotic syndrome. The patient in our case report initially complained of flank pain. Laboratory evaluation revealed acute renal failure and nephrotic syndrome with substantial proteinuria. Multiple imaging modalities demonstrated a large, ill-defined, infiltrative retroperitoneal soft tissue mass that encased the great vessels. The patient was also noted to have acute left renal vein thrombosis. Although the patient was eventually diagnosed with RPF of unknown etiology, his clinical course is particularly unique given the rarity of the renal vein thrombosis. This case report adds a value to the medical community by helping to elucidate RPF and exposing its potentially life-threatening complications.

INTRODUCTION

Patients afflicted with nephrotic syndrome are predisposed to the development of deep venous thrombosis, particularly of the renal vein. Although uncommon, retroperitoneal fibrosis (RPF) has been associated with nephrotic syndrome in several case reports [1]. Idiopathic RPF is classified as 'chronic periaortitis', which is a broad term encompassing disorders of inflammation and fibrosis surrounding the aorta. Given the rarity of the disease and non-specific presentation, RPF can be difficult to diagnose. We present an unusual case report of RPF resulting in acute left renal vein thrombosis and clinically evident nephrotic syndrome.

CASE REPORT

A 59-year-old Caucasian male with chronic, controlled hypothyroidism and essential hypertension sought treatment for sub-acute left-sided flank pain and constitutional symptoms. The patient first presented to an outside facility, receiving renal sonography that revealed mild left-sided hydronephrosis. Subsequently, he was admitted to our hospital for ongoing work-up, where laboratory evaluation revealed acute renal failure and newly diagnosed nephrotic syndrome. The patient's serum creatinine level peaked at 1.39 mg/dl (baseline value was 0.7 mg/dl). The serum blood urea nitrogen was 31 mg/dl on admission. A spot urine sample contained over 10 g of protein per gram creatinine. The serum inflammatory markers were abnormally high; erythrocyte sedimentation rate was >119 mm/h, which is the highest measured value at our institution's laboratory. The C-reactive protein was 36.2 mg/l. Contrasted computed tomography imaging of the abdomen revealed an ill-defined, infiltrative retroperitoneal soft tissue mass (4.6 × 6.0 × 8.5 cm) that encased the abdominal aorta and inferior vena cava (Fig. 1). This same image also demonstrated expansion of the left renal vein with thrombus extending into the inferior vena cava. Serum protein electrophoresis showed a potential M-spike, but immunofixation was consistent with polyclonal spike. The renal failure partially resolved with intravenous hydration and supportive care measures. The left renal vein thrombosis was treated with systemic

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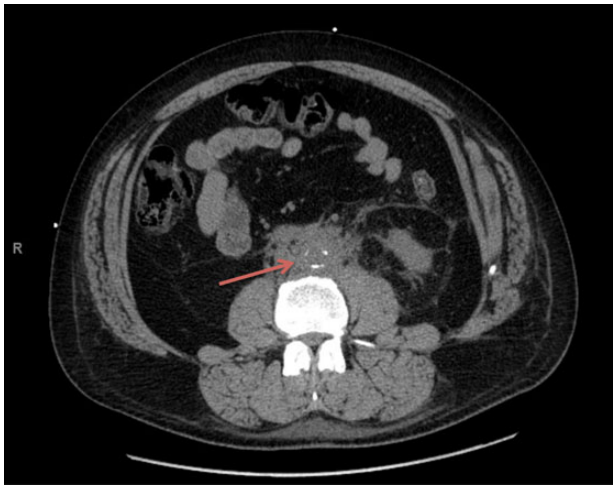

Figure 1: Infiltrative retroperitoneal soft tissue mass encasing the infrarenal aorta and inferior vena cava.

anticoagulation. The radiographic appearance of the soft tissue mass was concerning for RPF. However, due to the encroachment on the abdominal aorta and inferior vena cava, a biopsy was not feasible. The patient was discharged home in stable condition on warfarin therapy. Follow-up appointment with rheumatology showed increased proteinuria (16 g/gCr). Interestingly, the IgG4 subtype was found to be in the upper limits of normal, raising some suspicion for IgG4-related disease. Given the progression of the disease, the patient was empirically started on high-dose corticosteroid therapy (prednisone 60 mg daily); the patient was followed with serial inflammatory markers and computed tomography imaging. At the time of publication, the patient is demonstrating a favorable response to therapy, as inflammatory markers and measurements of the RPF are decreasing.

DISCUSSION

RPF is a rare condition characterized by inflammation and fibrosis in the retroperitoneal space. A population-based study in the Netherlands estimated the annual incidence of RPF to be 1.3 per 100 000 cases per year [2]. RPF is a known culprit of hydronephrosis and renal failure, given the ability of the soft tissue mass to entrap the ureters. RPF is generally an idiopathic disorder, which has been termed Ormond's disease, but it can also be secondary to the use of certain drugs, radiotherapy, malignancies, surgeries or infections [3]. Patients most commonly present for medical evaluation because of flank pain and weight loss [4]. There are no specific hematologic or biochemical abnormalities associated with RPF, making a laboratory-based diagnosis difficult. Given the systemic inflammation that is often present, erythrocyte sedimentation rate and C-reactive protein levels are frequently elevated. The extension of RPF is best visualized with a contrast-enhanced CT scan, though MR imaging is also helpful. IgG4-related disease includes autoimmune pancreatitis and a recently described subset of cases of RPF previously described as idiopathic [5]. The utility of measuring serum IgG antibodies still has a nebulous role in the setting of periaortitis; Moroni *et al.* [6] described such antibodies in both healthy patients and those afflicted with chronic periaortitis.

Currently, glucocorticoids are the mainstay of treatment for RPF [7]. However, more case reports with serial follow-up with a documented response to glucocorticoids are necessary to better assess treatment efficacy.

Known complications of RPF include hydronephrosis, renal failure and hypertension; although less common, there are case reports of bilateral iliac and caval venous thrombosis [8]. To the best of our knowledge, there are no previous case reports of acute renal vein thrombosis and nephrotic syndrome in the setting of RPF.

CONCLUSION

Renal vein thrombosis secondary to RPF is an unusual and life-threatening event. Given the nebulous clinical presentation, clinicians should remain keenly aware of RPF and its potentially fatal complications.

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CONFLICT OF INTEREST STATEMENT

None declared.

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