CLINICAL VIGNETTE

Primary Retroperitoneal Fibrosis Causing Obstructive Uropathy

Samir Bhalla, MD and Tina Kapadia, DO

Case Presentation

A 47-year-old male was directed to the hospital with abdominal pain and abnormal outpatient laboratory studies. He has morbid obesity treated with semaglutide with 100lb weight loss over the past 18 months. He reports midline abdominal pain radiating to the bilateral lower quadrants for several weeks. Outpatient labs revealed acute renal failure with creatinine 8.88 mg/dL (0.60-1.30 mg/dL), BUN 73 mg/dL (7-22 mg/dL), and potassium of 5.5 mmol/L (3.6 - 5.0 mmol/L). Basic metabolic panel two months prior had completely normal renal function. He was referred to the emergency department for expedited evaluation of renal failure. On admission he was afebrile, but hypertensive, 196/115 mm Hg, with a normal pulse and room air oxygen saturation. Physical examination noted central abdominal tenderness without guarding or rebound, as well as 1+ lower extremity edema. CT imaging of the abdomen and pelvis revealed a 6.6 x 3 x 8 cm retroperitoneal mass abutting the anterior margins of the lower aorta and moderate bilateral hydronephrosis (see Figure 1). MRI of the abdomen and pelvis confirmed the soft tissue mass within the retroperitoneum draping anteriorly along the abdominal aorta and extending along the proximal common iliac arteries. The patient underwent urgent bilateral nephrostomy tube placement, with gradual normalization of renal function. Surgical consultation was obtained, and biopsied the retroperitoneal mass. Pathology demonstrated sclerotic fibrotic tissue with chronic lymphoid inflammation without evidence of malignant cells. Additional laboratory studies included elevated immunoglobulin G-4 level of 184 mg/dL (1-123 mg/dL), erythrocyte sedimentation rate 48 (0-20 mm/hr), and C-reactive peptide 41.40 mg/dL (0.00 - 0.90 mg/dL). Rheumatology consultation was obtained, and the patient was started on daily oral prednisone 40mg with plan for repeat imaging in several months.

Discussion

Retroperitoneal fibrosis (RPF) is a rare disease, which usually presents with abdominal, lower back and/or flank pain as the chief complaint.[1] Epidemiological data show a male predominance, with typical onset between 40 and 50 years. Complications can arise from compromise of other retroperitoneal structures such as ureters, causing obstructive uropathy as in this patient.[2] IVC compression can lead to venous stasis and deep venous thrombosis. Primary, or idiopathic, retroperitoneal fibrosis represents 70% of cases. It is thought to arise as a multifocal periaortic fibroinflammatory response from chronic aortitis and often affects other aortic areas.[1] RPF associated with immunoglobulin G-4 related disease accounts for about 40-60% of idiopathic cases.[3] Secondary retroperitoneal fibrosis can result from: medications, ergots, dopamine agonists, biological agents; malignancies (carcinoid, lymphoma and sarcomas), radiation therapy, and certain infections including tuberculosis.[1]

Initiation of treatment is needed to relieve compression/ obstruction of affected organs such as ureters and vascular structures. Glucocorticoids are the mainstay of treatment, usually oral prednisone dosed at 0.6 mg/kg/day.[4] Other immunosuppressive agents such as rituximab, methotrexate, or mycophenolate mofetil are used in patients with contraindications to steroids. Serial radiographs are often used to assess response to treatment. Inflammatory markers may also respond, but are not as reliable as imaging.

This patient was also taking semaglutide for weight loss. As of this submission, there are no published case reports linking semaglutide to retroperitoneal fibrosis. However, this medication is still quite new, with increasing utilization.

Conclusion

We present a rare case of primary retroperitoneal fibrosis likely resulting from immunoglobulin G-4 related disease. He presented with abdominal pain and obstructive renal failure. While this patient was taking semaglutide, currently there are no published associations between semaglutide and RPF.

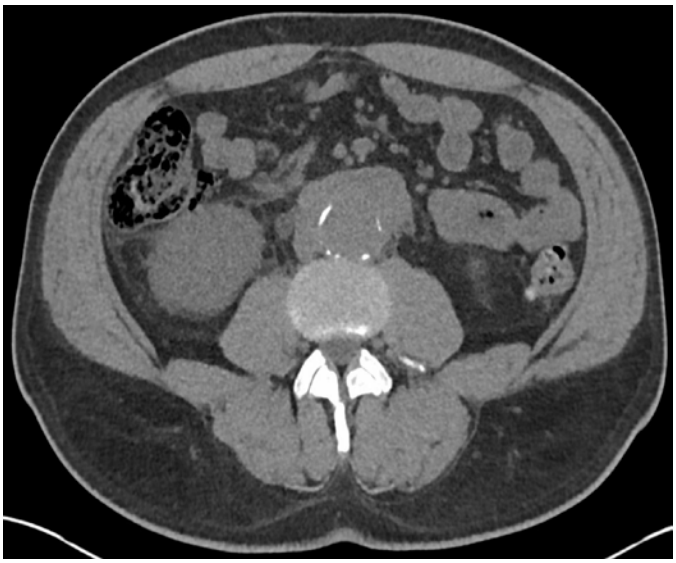

Figure 1: Cross sectional CAT imaging showing a large irregular retroperitoneal mass with resultant bilateral hydronephrosis.

REFERENCES

1. **Vaglio A, Salvarani C, Buzio C**. Retroperitoneal fibrosis. *Lancet*. 2006 Jan 21;367(9506):241-51. doi: 10.1016/S0140-6736(06)68035-5. PMID: 16427494.
2. **Baker LR, Mallinson WJ, Gregory MC, Menzies EA, Cattell WR, Whitfield HN, Hendry WF, Wickham JE, Joekes AM**. Idiopathic retroperitoneal fibrosis. A retrospective analysis of 60 cases. *Br J Urol*. 1987 Dec; 60(6):497-503. doi: 10.1111/j.1464-410x.1987.tb05028.x. PMID: 3427331.
3. **Stone JH, Zen Y, Deshpande V**. IgG4-related disease. *N Engl J Med*. 2012 Feb 9;366(6):539-51. doi: 10.1056/NEJMra1104650. PMID: 22316447.
4. **Fenaroli P, Maritati F, Vaglio A**. Into Clinical Practice: Diagnosis and Therapy of Retroperitoneal Fibrosis. *Curr Rheumatol Rep*. 2021 Feb 10;23(3):18. doi: 10.1007/s11926-020-00966-9. PMID: 33569638.