

Spontaneous Resolution of Retroperitoneal Fibrosis in a Young Man

Shirley Shapiro MD^{1,2}, Shira Shoher MD^{1,2}, Dror Cantrell MD^{1,2}, and Micha J. Rapoport MD^{1,2}

¹Department of Internal Medicine C, Shamir Medical Center (Assaf Harofeh), Zerifin, Israel

²Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

*In memory of the dear Professor Rapoport, a doctor who believed in people.
Thank you for the part you took in my story.*

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Retroperitoneal infiltration, also known as retroperitoneal fibrosis (RPF), is a rare condition, which mostly occurs in men over the age of 40 years. This condition involves inflammation of the soft tissue of the retroperitoneal cavity, most commonly around the infrarenal abdominal aorta, iliac arteries, ureters, and abdominal organs. Clinical manifestations consist of severe pain in the lower back, abdomen, or flank, which may radiate to the inguinal region. Pain may be acute at the onset and can be mistaken for renal colic. Renal and ureteral involvement is common and can develop into acute kidney injury and hypertensive crises.

Idiopathic RPF is immune mediated, which may be isolated or associated with autoimmune diseases such as immunoglobulin G4-related disease (IgG4-RD). It may also be secondary to malignancies, infectious diseases, prior surgery, radiation, and medications. Treatment depends on the infiltration cause and is intended to relieve the obstruction and slow the progression of the fibrosis. RPF is generally progressive and irreversible. Spontaneous regression is uncommon despite various treatment modalities.

We describe a patient with idiopathic RPF and an unusual spontaneous recovery.

PATIENT DESCRIPTION

A 39-year-old man presented to the emergency department with a sudden and severe onset abdominal and flank pain accompanied by nausea and vomiting. His medical history was unremarkable apart from a prior abdominoplasty after weight loss. He declined smoking, alcohol consumption or illicit drug use, and recent treatment with any prescription medications or over the counter drugs. On admission, he was afebrile, and his vital signs were within normal limits apart from mildly elevated blood pressure of 148/73 mmHg. Physical examination revealed only a periumbilical tenderness and submandibular lymphadenopathy. Initial laboratory evaluation, including full chemistry, complete blood count, C-reactive protein (CRP), and urine analysis was within normal limits. An abdominal-pelvic computed tomography (CT) scan demonstrated multiple retroperitoneal findings including fat infiltration surrounding the aorta, its branches, and the inferior vena cava (IVC) with multiple enlarged lymph nodules up to 8 mm diameter without hydronephrosis or nephrolithiasis. These findings were suggestive of a diffuse inflammatory process compatible with aortitis, lymphoma, retroperitoneal fibrosis, and extramedullary plasmacytoma [Figure 1A]. The differential diagnosis of the CT scan findings included an inflammatory process of unknown etiology, aortitis, lymphoma, retroperitoneal fibrosis, and extramedullary plasmacytoma.

Extended diagnostics workup with blood and urine cultures and a full collagenogram including IGG4 levels, tumor markers, plasma, urine protein electrophoresis, and testicular sonogram did not reveal any abnormalities.

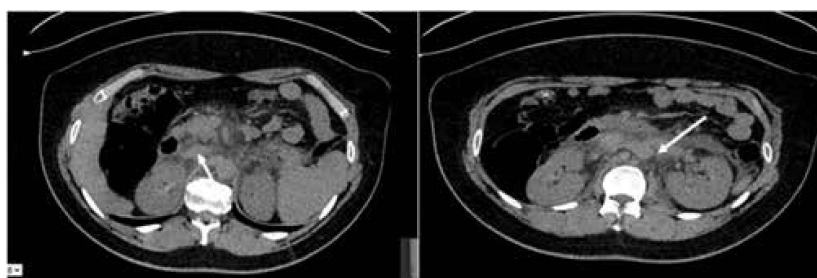
A second CT scan performed a few days after his admission demonstrated a vast retroperitoneal infiltration involving the aorta and the IVC, extensive lymphadenopathy, and overall worsening of the previous findings, but no signs of urinary obstruction [Figure 1B].

The patient was discharged, and a CT-guided retroperitoneal tissue biopsy was scheduled for 5 days later. However, on readmission, the patient reported resolution of all his symptoms. A third CT scan demonstrated regression of all retroperitoneal findings [Figure 1C]. A biopsy was canceled, and the patient continued the follow-up in the outpatient clinic. Three months later, a CT scan of the retroperitoneum was completely normal, and no complaints were reported on an annual follow-up visit.

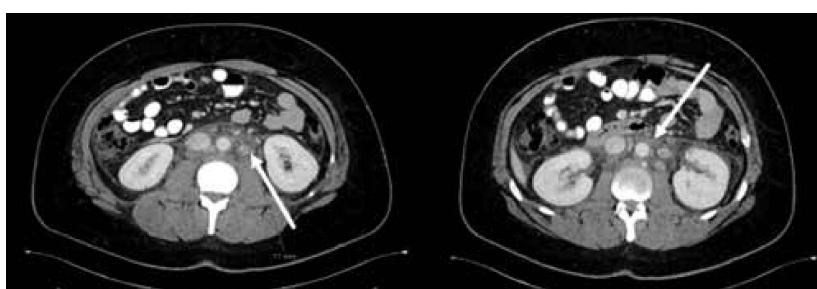
COMMENT

Our patient presented with classical findings compatible with RPF. An extensive diagnostic workup failed to demonstrate a secondary cause for his condition, suggesting an extensive primary idiopathic disease. It could be argued that in the absence of retroperitoneal tissue biopsy, a definite diagnosis of RPF cannot be made with certainty nor could a secondary cause be ruled. However, in the presence

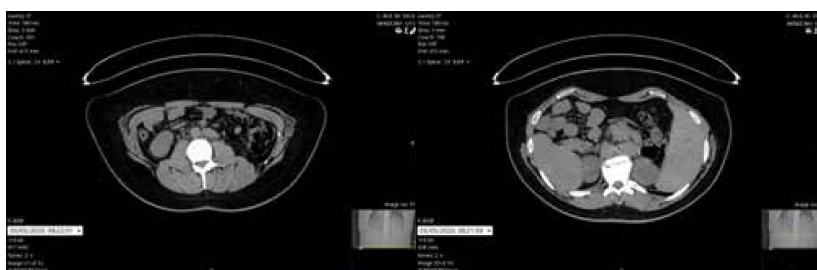
Figure 1. Abdominal-pelvic non-contrast CT scan
CT = computed tomography, IVC = inferior vena cava



[A] A first non-contrast CT demonstrating multiple retroperitoneal findings of fat infiltration surrounding the aorta, its branches, and the IVC with lymphadenopathy suggestive of a diffuse inflammatory process



[B] A second CT scan with contrast demonstrating a vast retroperitoneal infiltration involving the aorta and the IVC, extensive lymphadenopathy, and overall worsening of the previous findings but no signs of urinary obstruction



[C] A third non-contrast CT before the scheduled biopsy demonstrating complete regression of all retroperitoneal findings

of typical symptoms and radiographic findings, a biopsy may be considered unnecessary as 23% of patients are diagnosed based on radiographic findings alone without a biopsy [1].

Spontaneous regression of RPF is extremely rare. A review of the relevant literature revealed three cases of such spontaneous regression. Williams and colleagues [2] described a 54-year-old woman who presented with similar symptoms of elevated CRP and eryth-

rocyte sedimentation rate. Her CT scan was compatible with RPF, which was confirmed by retroperitoneal biopsy. She was treated with bilateral ureteric stenting due to a deterioration of her renal function but refused steroid treatment for fear of potential side effects. After 12 months her pain subsided, levels of inflammatory markers decreased, and the CT scan findings in the retroperitoneum completely resolved.

In two other cases described by

Kume and Kitamura [3] and Robbé and Dixon [4], spontaneous regression of fibrosis occurred in patients admitted with idiopathic RPF complicated by obstructive uropathy. Both patients were treated symptomatically with temporary nephrostomy to relieve urinary obstruction. There was no documented recurrence during the follow-up period [3-5]. These patients were not treated for their underlying disease, but rather for the complications of the disease underscoring the rarity of the spontaneous regression in our patient whose symptoms and imaging findings resolved without any intervention.

CONCLUSIONS

Spontaneous and complete recovery of RPF is possible and may occur in rare cases. Given repeated reports of spontaneous recovery and the problematic nature of the pharmacologic treatment, a watchful waiting strategy should be considered in the absence of a clear indication for conservative or surgical treatment.

Correspondence

Dr. S. Shapiro
Dept of Internal Medicine C, Shamir Medical Center
(Assaf Harofeh), Zerifin 70300, Israel
email: shirliutka@gmail.com

References

1. Kermani TA, Crowson CS, Achenbach SJ, Luthra HS. Idiopathic retroperitoneal fibrosis: a retrospective review of clinical presentation, treatment, and outcomes. *Mayo Clin Proc* 2011; 86 (4): 297-303.
2. Williams G, Sarkar B, Neilly JB, McKay GA. Spontaneous remission in a case of idiopathic retroperitoneal fibrosis. *Scott Med J* 2013; 58 (2): e7-9.
3. Kume H, Kitamura T. Spontaneous regression of bilateral hydronephrosis due to retroperitoneal fibrosis. *Scand J Urol Nephrol* 2001; 35 (3): 255-6.
4. Robbé IJ, Dixon AK. Spontaneous resolution of ureteric obstruction in perianeurysmal retroperitoneal fibrosis. *Br J Radiol* 1984; 57 (673): 92-5.
5. Caiafa RO, Vinuesa AS, Izquierdo RS, Brufau BP, Ayuso Colella JR, Molina CN. Retroperitoneal fibrosis: role of imaging in diagnosis and follow-up. *Radiographics* 2013; 33 (2): 535-52.