IgG-4 Related Retroperitoneal Fibrosis: A Rare Association with Riedel's Thyroiditis

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Case Presentation

A 53-year-old male with history of RT previously treated with isthmectomy for compressive symptoms relief who presented with one week of severe localized lower abdominal and suprapubic pain. He denied any fever, gastrointestinal symptoms, genitourinary symptoms, or weight loss. He was initially diagnosed with acute prostatitis and was treated with ciprofloxacin without improvement of symptoms, which prompted him the second visit to the emergency room. The physical exam demonstrated a flat, soft abdomen with normal bowel sounds and no palpable masses, but with diffuse tenderness across the lower abdomen, especially in the right lower quadrant and suprapubic region.

The patient underwent a non-contrast CT scan of the abdomen and pelvis with findings significant for an extensive, predominantly right-sided retroperitoneal mass encircling the aorta, inferior vena cava and proximal right ureter producing severe obstructive uropathy of the right kidney with massive distention of the pyelocaliceal system and proximal ureter and marked loss of renal cortex (Figure 1).

The patient underwent right ureteral stent placement, which partially resolved hydronephrosis and restored kidney function. The CT-guided biopsy of the retroperitoneal mass revealed fibro-inflammatory tissue without specific features (Figure 2). The immunohistochemistry staining was notable for IgG4 positive plasma cells (Figure 2) and CD68 positive histiocytes.

The patient was finally diagnosed with IgG4-related systemic fibrosclerosis. The additional lab testing showed normal LDH, uric acid and IgG4 levels. He was started on high-dose prednisone at 60 mg daily. Throughout this time, he developed acute renal failure requiring additional stent placement by urology as well as refractory pain necessitating the use of narcotics.

Over the next few months, along with continuing decrease in the size of the retroperitoneal mass on the follow-up CT (Figure 3), his analgesic requirements began to decline. His renal function improved and he was able to taper prednisone to a lower dose.

Given the presence of an IgG4 positive plasma cell infiltrate, retroperitoneal fibrosis and history of RT, the diagnosis of IgG4-related retroperitoneal fibrosis in a patient with Riedel's thyroiditis was made.

Case Discussion

IgG4-RD is an immune-mediated fibroinflammatory condition capable of affecting multiple organs. It is characterized by extensive fibrosis in various organs including the pancreato-hepato-biliary system, retroperitoneum, mesentery, aorta, salivary and lacrimal glands.

Retroperitoneal fibrosis in IgG4-RD can present with poorly localized pain in the back or lower abdomen, leg edema, or hydronephrosis from ureteral or prostate involvement.

Figure 1: Initial Imaging

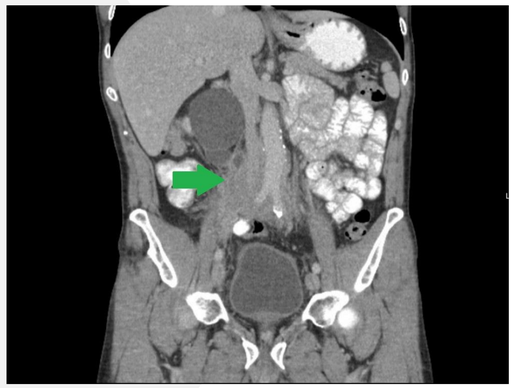

Figure 1: predominantly right-sided aortic Retroperitoneal encircling aorta and IVC

Figure 3: Follow-Up Imaging

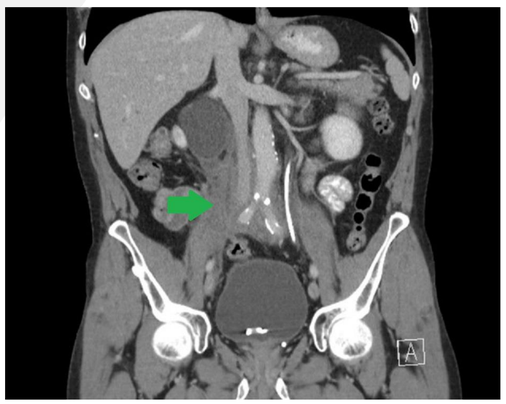

Figure 2: Decreased size (7.7 vs 8.1 cm) of mass

Figure 2: Pathology

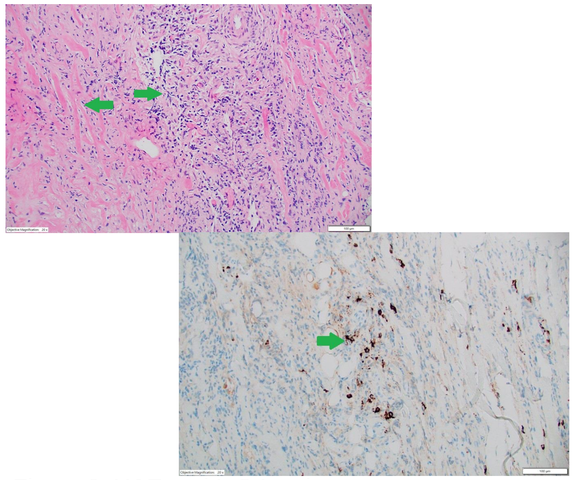

Figure 2: H&E and IgG4 stain

Figure 4: ACR/ELAR 2019 IgG4-RD Criteria

- Presence of Inclusion criteria AND
- Absence of exclusion criteria AND
- Inclusion criteria ≥20
  - Histopathologic findings
    - Lymphocytic infiltrate, IgG4/hpf, IgG4/IgG ratio
  - Serologic findings
  - Imaging findings

RT is considered an IgG4-RD of the thyroid gland. Approximately one third of patients with RT eventually manifest other signs of systemic fibrosis over a 10-year period, although there are only rare case reports of RT and RF. Recognition of the risk of developing other features of IgG4-RD in patients with RT is important. Prompt recognition of extrathyroidal IgG4-RD will aid early diagnosis and treatment.

Conclusion

- Presence of inclusion, absence of exclusion, and inclusion criteria = 26
- Met criteria to diagnose IgG4-RD
- Steroid treatment lead to decreasing fibrosis in surrounding retroperitoneal organs
- Likely represents a rare case of IgG4-RD RF in RT patient

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