IgG4 Gastropathy: Hemorrhagic Gastropathy as the Culminating Presentation of Undiagnosed IgG4-Related Disease

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Background

- IgG4 related disease (IgG4-RD) is a newly recognized, immune-mediated fibro-inflammatory disease with IgG4-positive plasma cell infiltration (10-200 cells/hpf)[1,2]
- Serum IgG4 is often elevated (60-70% of cases, although not required for diagnosis)[2]
- Diagnosis is based on histology (lymphoplasmacytic infiltrate, focal fibrosis (storiform pattern), obliterative phlebitis), laboratory findings, and clinical context[1,2]
- IgG4-RD can affect almost any organ, including but not limited to, the pancreatic-biliary system, salivary/lacrimal glands, retroperitoneum, liver, aorta, lymph nodes, and rarely, the gastrointestinal tract[1-3] (8 cases of isolated gastric lesions)[4-6]
- Treatment includes glucocorticoids or other immunosuppression (rituximab, methotrexate, azathioprine)[1,2]

Case Presentation

- 81-year-old woman with past medical history of Sjogren's disease, myelodysplastic syndrome, and chronic myelomonocytic lymphoma presented to the emergency department for melena. She denied abdominal pain, nausea, or vomiting. No NSAID or alcohol use. Patient was hemodynamically stable. Physical exam unremarkable except for melena confirmed on digital rectal exam.
- Reported history of pancreatitis (records unavailable), fine needle aspiration of pancreatic head and tail (ten years prior) showed predominantly acute inflammatory cells, necrotic tissue debris and benign ductal epithelial cells, respectively
- Salivary gland biopsies (ten years prior) showed benign mucinous gland tissue and multiple reactive lymphoid follicles

BMP	
Na	139 mmol/L
K	4.5 mmol/L
Cl	107 mmol/L
CO2	23 mmol/L
BUN	**43 mg/dL ^**
Cr	0.75 mg/dL
BUN/Cr	**57.3 ratio ^**

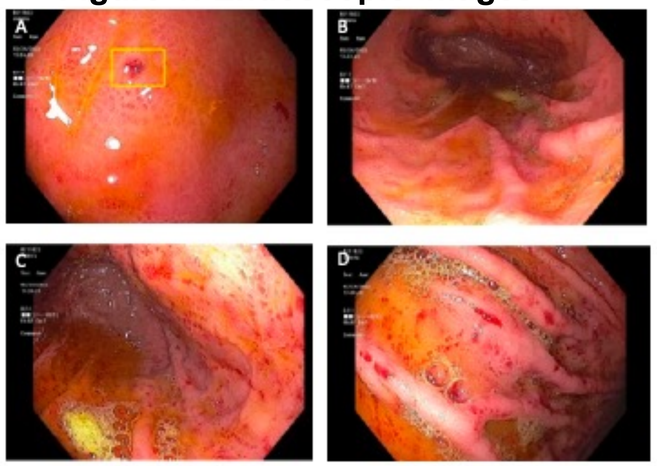
Immunology	
IgG	**2,344 mg/dL ^**
IgA	**637 mg/dL ^**
IgM	**727 mg/dL ^**
SPEP/UPEP	No monoclonal peak
IgG4	**271 mg/dL ^** (2-96 mg/dL)

CBC	
WBC	**2.53 K/uL v**
Hgb	**5.0 g/dL v** (baseline 8)
PLT	**37 K/uL v**

Bone Marrow Biopsy
Appropriately decreased cellularity, no monocytes on flow cytometry

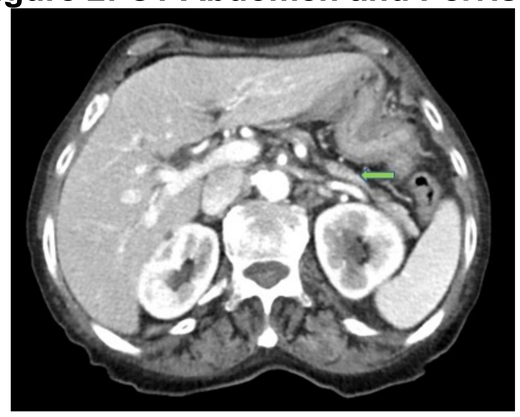
Endoscopy and Imaging Findings

Figure 1: Endoscopic Images



A) Small, high risk protruding red spot, cauterized B) gastric fundus and C, D) gastric body with hemorrhagic gastropathy

Figure 2: CT Abdomen and Pelvis



CT with diffusely atrophic pancreas noted (arrow)

- **Endoscopic findings:** Normal esophagus, no active bleeding or ulcers, small, protruding red spot that was cauterized, hemorrhagic gastropathy in fundus and body (Fig 1), biopsied, normal duodenum
- **Histology:** Gastric mucosa with patchy severe chronic gastritis and predominant plasma cell infiltration (without light chain restriction)
- **CT abdomen/pelvis:** diffusely atrophic pancreas (Fig 2) and infrarenal aneurysmal abdominal aorta (AAA, 4.8 cm)
- Melena resolved with supportive care
- Four months after initial evaluation, patient suffered from a cerebrovascular event and passed away a month later after progressive debilitation

Discussion

- On initial evaluation, patient appeared to present with isolated IgG4 gastropathy, rarely demonstrated in the literature
- However, on further review, given patient's history of salivary gland disease, pancreatic disease, and AAA, high degree of suspicion existed for underlying IgG4-RD connecting previous diagnoses with gastric biopsy and serum IgG4 level findings
- Patient underwent re-evaluation with hematology-oncology, including bone marrow biopsy, which was negative for underlying malignancy. Ruling out alternate diagnoses (i.e., underlying malignancy) is important in IgG4-RD diagnosis
- Given our patient's clinical course and undiagnosed potential IgG4-RD, they had not received glucocorticoids or other immunosuppression to assess response
- Given broad differential diagnosis of various GI tract pathologies, biopsy for further histologic evaluation is important. It is particularly important in GI tract diagnosis, as it may manifest in various ways (gastropathy as in our patient's case, or mass lesions, tumors, or ulcers)[5,6]

Conclusion

This case highlights the rare GI tract presentation of a patient's heretofore unrecognized IgG4 disease. IgG4-RD may present in various ways, and clinicians must remain vigilant for appropriate diagnosis and treatment

References

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