## CLINICAL VIGNETTE

# Autoimmune Pancreatitis

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### *Introduction*

Autoimmune pancreatitis (AIP) is an uncommon immunologically-mediated subtype of chronic pancreatitis associated with extensive lymphocyte infiltration. Although more specific diagnostic methods have led to increased diagnosis, true prevalence remains estimated at only 5-6% of all patients with chronic pancreatitis[1]. AIP can present as an illness limited to the pancreas itself or with widespread systemic manifestations. Treatment with corticosteroids leads to significant improvement of mass lesions, strictures and symptoms.

### *Case Report*

A 49-year-old female with a history of Graves disease in remission as well as ulcerative proctitis presented to family medicine clinic complaining of ten days of gradually progressive "aching" epigastric pain radiating to the back, worse with eating and leaning forward, improved with warm baths, and associated with bloating and significant stress at work. She denied diarrhea, constipation, nausea, vomiting, fevers or alcohol use. She had been experiencing bright red blood per rectum for six months and was diagnosed with ulcerative proctitis on colonoscopy two months prior. At that time she declined all medical interventions. Physical examination revealed temperature 98.5°F, blood pressure 128/75 mm Hg, pulse 98, and oxygen saturation 99% on room air. She was fatigued but in no apparent distress; exam was notable for mild epigastric tenderness to palpation without rebound or guarding and resolving completely after initial palpation. Presumptive diagnosis at that time was gastroesophageal reflux disease; labs were ordered and the patient was given information on dietary modifications. She declined a trial of antacid therapy.

Labs were notable for elevated amylase 246 and lipase 844, with normal complete blood count, comprehensive metabolic panel, and fasting lipid panel. The patient reported ongoing abdominal pain and was referred to gastroenterology. Autoimmune pancreatitis versus a posterior penetrating duodenal ulcer was considered. Abdominal CT, pancreatic protocol as well as repeat labs were ordered. She failed to return for GI follow-up, but when seen in primary care two months later had been started on prednisone by an outside gastroenterologist "for some sort of autoimmune disorder," with improvement in abdominal pain.

### *Discussion*

AIP is now classified into two subtypes: type 1, in which pancreatitis is one part of a systemic IgG4-mediated autoimmune illness, and type 2, a non-IgG4-mediated disease confined to the pancreas itself, but often associated with inflammatory bowel disease. The most common presentation is obstructive jaundice[2]; other clinical manifestations vary between the subtypes but may include abdominal pain, nausea, vomiting, jaundice, elevated amylase and lipase as well as disproportionately high serum alkaline phosphatase. Systemic components of type 1 AIP are marked by IgG4 positive plasma cells found in a variety of tissues[3] and may manifest as mediastinal or retroperitoneal fibrosis, inflammatory bowel disease, or salivary gland disorders. Imaging, generally in the form of CT or MRI, typically reveals a pancreatic enlargement, mass or stricture in the majority of patients[4], making it difficult to distinguish from pancreatic carcinoma based on imaging alone.

Diagnosis depends upon the clinical subtype: type 2 requires histologic duct-centric pancreatitis without IgG4-positive cells or systemic disease. Conversely, type 1 AIP uses diagnostic guidelines developed by the Mayo Clinic, known as the "HISORt" criteria[5]:

- Histology suggestive of autoimmune pancreatitis per pancreatic biopsy.
- Imaging findings on CT or MRI typical of AIP such as diffuse pancreatic enlargement
- Serum IgG4 (greater than or equal to two times the upper limit of normal)

- Other organ involvement (salivary glands, kidneys, inflammatory bowel disease, mediastinal lymphadenopathy, biliary strictures, retroperitoneal fibrosis)
- Response to glucocorticoids.

AIP has an excellent response to corticosteroids, such as prednisone 40mg/day for one month followed by a taper of 5mg/week for a total of three months[2]. This duration is based on observational data, as no randomized controlled trials exist. Response may take up to four months and is generally monitored with serial serum IgG4 levels and imaging as needed. Relapse rate is significant in type 1 AIP[1], with approximately 25% of patients requiring a second course of glucocorticoids. Refractory cases have shown benefit from further therapy such as azathioprine and, less commonly, 6-mercaptopurine, rituximab, cyclosporine, and cyclophosphamide.

### *Conclusion*

Although our patient was lost to follow-up, her absence of risk factors for traditional pancreatitis as well as her past history of Graves and inflammatory bowel disease and response to steroid therapy make type 2 AIP a likely cause of her abdominal pain and elevated lipase. Further studies are needed to better understand the pathophysiology and treatment of this rare but serious illness.

## REFERENCES

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