An Unusual Case of Abdominal Pain and Weight Loss in a Patient With Leukocytoclastic Vasculitis

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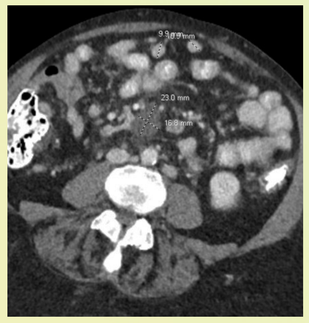
CASE PRESENTATION

- 60-year-old female presenting to the hospital for abdominal pain, nausea, and history of weight loss.
- **Past medical history**
  - Cognitive delay
  - Leukocytoclastic vasculitis dx 2006
  - Asthma
  - Gerd
  - MGUS
  - Secondary parkinsonism
- **Rheumatological history**
  - Hx of lower extremity rash in which biopsy showed evolving leukocytoclastic vasculitis. During initial evaluation, no definitive findings of major organ vasculitis
  - Hx of eosinophilia and low complement levels
  - Most recent occurrence of leukocytoclastic vasculitis was in 2019 and treated with low dose prednisone with good response

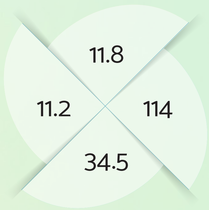
IMAGING PRIOR TO ADMISSION

- CT Abdomen/Pelvis two weeks prior to admission showed multifocal mesenteric and retroperitoneal lymph nodes and mucosal wall thickening of the sigmoid colon and rectum



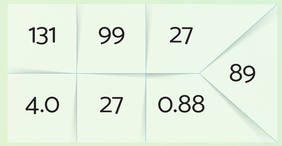
INITIAL WORK UP

- **Vitals**
  BP 112/62
  HR 87
  Temp 98.2
  RR 16, SpO2 93% on RA



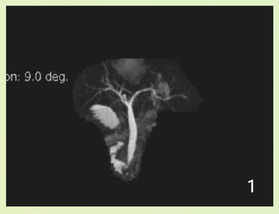
- **Physical exam**
  - Unremarkable except for abdominal tenderness and distension

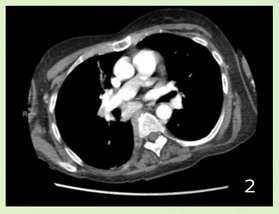
AST 91
ALT 39
Albumin 1.5
Lipase 25



IMAGING FROM HOSPITALIZATION

- MRCP was suspicious for calculus cholecystitis with minimal biliary ductal dilatation but no evidence of choledocholithiasis (figure 1)
- CT Chest showed mediastinal and bronchial adenopathy and interstitial infiltrates and pleural thickening at the lung bases which is new compared to old exams (figure2)





RHEUMATOLOGY WORK UP

- ESR: 90, CRP: 129
- ANA Comprehensive Panel: Negative
- C3: 28, C4: < 6
- Complement CH50: < 13
- ANCAs
  - Anti-Neutrophil Cytoplasmic Antibody: Negative
  - PR3: 29
  - MPO: Negative
- ACE Level: 28
  SPEP: Monoclonal protein (IgG Kappa and Lambda Free Light Chains)
- Immunoglobulins
  - IgE 622
  - IgM 14
  - IgA 53
  - IgG 3917
- Cryoglobulins: Negative
- IGG subclass 4
  - Prior to admission: 2703
  - During admission: 2054
- EGD Biopsy from gastric antrum
  - Submucosal and lamina propria fibrosis with numerous IgG4+ cells suspicious for IgG4-RD
- Patient underwent cholecystectomy by general surgery and OR pathology showed
  - IgG4-associated cholecystitis in the setting of dense lymphoplasmacytic infiltrates admixed with eosinophils, dense fibrosis in a storiform pattern, and increased numbers of IgG+ plasma cells

DIAGNOSIS AND TREATMENT

- Collaboration between rheumatology, gastroenterology, surgery, and hematology teams
- Initial biopsies from EGD showed fibrosis with numerous IgG4+ cells suspicious for IgG4-RD which was confirmed on gallbladder pathology from OR specimen by surgery
  - Patient diagnosed with IgG4-associated cholecystitis
- **Treated with prednisone taper**
  - 40 mg daily x 1 week, 30 mg daily x 2 weeks, 20 mg x 2 weeks, 15 mg x 2 weeks, currently on 10 mg daily
- Significant improvement in IgG4 levels and complements in 1 month
  - IgG4 levels: 2703 → 336
  - C3: 121, C4: 31.8

DISCUSSION

- IgG4-Related Disease (IgG4-RD) is a multi-system immune mediated fibroinflammatory disorder
  - Dense lymphoplasmacytic infiltrations with IgG4-positive plasma cells
  - Fibrosis – classically "storiform" pattern
  - IgG4 levels typically > 135 mg/dL
  - Type 1 Autoimmune pancreatitis (AIP)
  - IgG4-related sclerosing cholangitis
  - Major salivary gland enlargement or sclerosing sialadenitis
  - Orbital disease
  - Retroperitoneal fibrosis
- Epidemiology
  - 1 per 100,000
  - Slight predominance in middle-age/older males
- IgG4-related sclerosing cholangitis typically occurs with Type 1 AIP therefore making the diagnosis of isolated IgG4-related cholecystitis or sclerosing cholangitis a challenge
  - Two Types
    - Diffuse gallbladder wall thickening
    - Localized Mass
- Can often mimic malignancy specifically cholangiocarcinoma
  - Biopsy via EUS can be limited and a cholecystectomy is often needed to make diagnosis
- Our patient presented with a history of abdominal pain, N/V, significant weight loss, and imaging with lymphadenopathy highly concerning for malignancy
  - This case demonstrates the importance of maintaining a broad differential as well as obtaining an adequate biopsy to make the correct diagnosis
- IgG4 levels have been found to be elevated in other rheumatologic conditions specifically ANCA-associated vasculitis
  - Our patient could have an IgG4-RD and vasculitis (with + PR3) overlap syndrome
- Further investigation is needed on isolated IgG4-related cholecystitis as well as IgG4-RD and vasculitis overlap syndromes

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