Collagenous Sprue and POEMS Syndrome: Association or Coincidence?

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Introduction

- Collagenous sprue (CS) is a rare small bowel enteropathy of unknown etiology that is hypothesized to be immune-mediated.[1]
- CS typically presents similarly to celiac disease (CD) with chronic diarrhea, weight loss, and malnutrition, and if untreated can lead to substantial mortality.
- CS is associated with autoimmune disorders, with it most frequently linked with CD. It is also associated with angiotensin receptor blocker, NSAID, statin, and proton pump inhibitor use.
- The presence of a subepithelial collagen band on histology differentiates CS from CD, as both have villous atrophy (Table 1).
- CS was once believed to be a refractory version of CD but is now considered to be a distinct disease.[2]
- It is important to make the correct diagnosis as treatment for CS not only requires adherence to a gluten-free diet but also often immunosuppressive therapy.
- Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, and Skin changes (POEMS) syndrome is a multisystemic disorder that also has an unknown etiology, but seldomly involves the GI system.[3]
- This is a case of a patient with CS with comorbid CD, who not long after the onset of sprue symptoms was also diagnosed with POEMS syndrome.

Initial Presentation

- A 75-year-old male with a history of coronary artery disease and hypertension presented with 6 months of intermittent nonbloody diarrhea, abdominal bloating, 20lbs weight loss, and generalized weakness.
- Surgical History: appendectomy
- Family History: none relevant
- Social History: active tobacco use (20 pack-years) and occasional alcohol use
- Medications: aspirin, losartan, simvastatin, metoprolol succinate
- Physical exam:
  - Vital signs within normal limits
  - Thin male with generalized muscle wasting and a benign abdomen
- Labs:
  - Hemoglobin 13.8 g/dL
  - Anti-tissue transglutaminase (TTG) IgA 158U, gliadin IgA 201U, and endomysial antibody 1:40
- The diagnosis of CD was confirmed by an EGD biopsy of the duodenal bulb and second portion which revealed marked villous atrophy and intraepithelial lymphocytosis (Figures A and B).
- Patient was started on a gluten-free diet and referred to nutrition.
- He was then lost to follow up.

Return to Care

- One year following his CD diagnosis, the patient was diagnosed with POEMS syndrome after being referred for peripheral neuropathy and an elevated IgM kappa monoclonal protein.
- Remission of POEMS syndrome was subsequently achieved with melphalan and dexamethasone.
- Four years after CD diagnosis, he had continued weight loss (12lbs) and was referred for GI evaluation.
- Patient noted difficulty adhering to a gluten-free diet.
- Labs:
  - Hemoglobin 13.2 g/dL
  - TTG IgA >250U and gliadin IgA ≥250U
- A repeat proximal duodenal biopsy showed a subepithelial collagen band of 32.2μm along with previous villous atrophy, consistent with CS (Figures C and D).
- On review, a subepithelial collagen band was present from the initial duodenal biopsy but to a lesser degree (28.6μm) (Figure B).

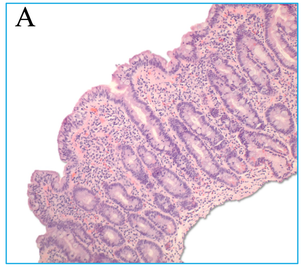
Discussion

- POEMS syndrome rarely results in GI manifestations with no previous cases of small bowel involvement.
- Thus, it is unclear if the diagnosis of both CS and POEMS syndrome within a short period of time was a coincidence or if there is a connection given their proposed similar immune-mediated pathogenesis.
- Due to the proinflammatory environment, it is plausible that CS and/or CD may have triggered the production of POEMS syndrome's monoclonal protein.
- Research investigating CS's pathogenesis and additional cases of CS with POEMS syndrome is needed to further support this hypothesis.

	Collagenous Sprue	Celiac Disease
Villous atrophy	+*	+
Intraepithelial lymphocytosis	+	+
Subepithelial collagen band	+*	-

Table 1. Comparison of the histologic features of collagenous sprue and celiac disease on biopsy of the proximal small bowel.

*Required for diagnosis of collagenous sprue.



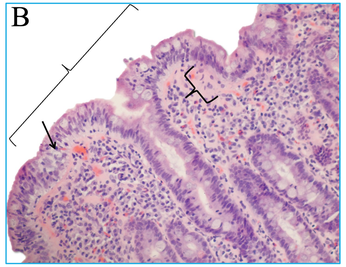
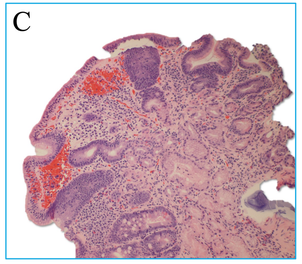


Figure A. Initial proximal duodenal biopsy under low-power view demonstrating villous atrophy.

Figure B. Initial proximal duodenal biopsy under high-power view demonstrating villous atrophy (large outer bracket), intraepithelial lymphocytosis (arrow), and 28.6μm subepithelial collagen band (small inner bracket).



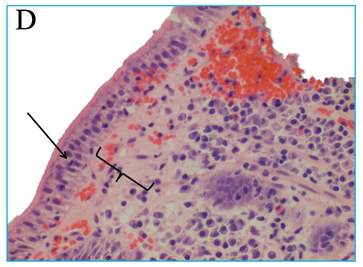


Figure C. Repeat proximal duodenal biopsy under low-power view demonstrating villous atrophy.

Figure D. Repeat proximal duodenal biopsy under high-power view demonstrating intraepithelial lymphocytosis (arrow) and 32.2 μm subepithelial collagen band (bracket).

References

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