Atypical GBS as The Presenting Symptom of Celiac Disease: Recognizing Extra-intestinal Symptoms of Celiac Disease

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INTRODUCTION

Celiac disease (CD) is an immune-mediated enteropathy in response to gluten that occurs in 1% of the population.

GI-related symptoms are classic, but extraintestinal symptoms such as cutaneous, asymptomatic liver abnormalities, and more rarely, neurologic symptoms are more common in the adult population.

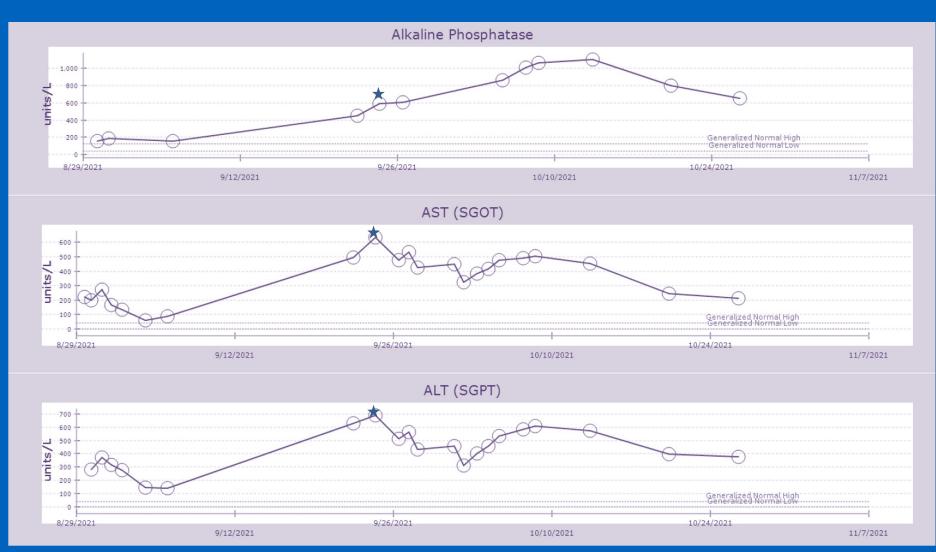
CASE 44-yo M with recent left-chest shingles, presented with right facial paralysis and left leg weakness. **Initially admitted for suspected atypical Guillain Barre** Syndrome (GBS) and treated with prednisone and IVIG. **Readmitted a few weeks later with worsening neurologic** status.

•A thorough infectious workup was unremarkable. He was also noted to have abnormal liver enzymes on prior admission that initially improved, but worsened on readmission (Table 1).

•Extensive liver disease workup was negative, thus subsequently underwent a liver biopsy (Fig.A) with findings concerning for autoimmune vs drug-induced hepatitis.

•Further workup: tissue transglutaminase IgA >250 U/mL and EGD with biopsies consistent with celiac disease. Liver enzymes improved with gluten avoidance, but neurologic impairments persisted.

Atypical extra-intestinal symptoms at presentation of CD can result in diagnostic delay and permanent neurological disability if not treated. Celiac disease has been found in as many as 9% of patients with elevated liver enzymes. It is imperative not only to recognize neurologic presentations of CD but also the spectrum of possible liver impairments. Adherence to a strict gluten-free diet has been shown to improve symptoms of both neuropathy and liver enzyme abnormalities but must be instituted quickly and continued life-long.



DISCUSSION

Table 1: Liver aminotransferases trend between patient's admissions; star indicates readmission.

LIVER DISEASE SEROLOGIC WORKUP

- HAV IgM/IgG

- Ceruloplasmin

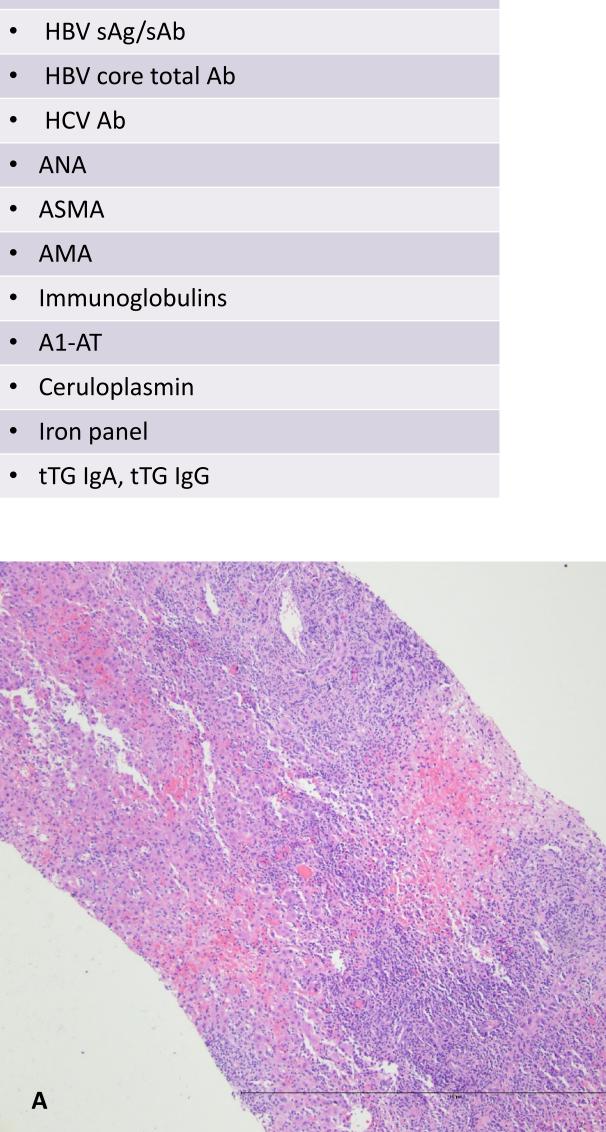


Fig. A: Core liver biopsy showing marked panlobular lymphoplasmacytic inflammation, bile duct epithelial damage, and hepatocytes necrosis in zone 2-3.

CONCLUSION

Early diagnosis can impact prognosis, as early gluten-free diet is the only approved life-long treatment to achieve recovery and prevent long-term complications.