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Visual Diagnosis Pearl

Gastric Antral Vascular Ectasia (GAVE) and Systemic Sclerosis

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A 76-year-old woman with a history of hypertension, diabetes mellitus type 2, depression, fibromyalgia, and obstructive sleep apnea presented with chronic hand pain and Raynaud's phenomenon. Her work up revealed new-onset iron deficiency anemia. She reported intermittent black stools but no bright red blood per rectum or symptoms of pica. She had chronic nausea but attributed it to her metformin use. Her upper endoscopy revealed gastric antral vascular ectasia (GAVE), characterized by longitudinal erythematous streaks radiating from the pylorus through the antrum, producing the classic "watermelon stomach" appearance (Figure 1). Although the diagnosis of GAVE was established via esophagogastroduodenoscopy (EGD), an underlying connective tissue disease was not identified until several years later. She was referred to rheumatology for persistent joint pain, fatigue, elevated antinuclear antibody (ANA), and ongoing Raynaud's phenomenon. Her physical examination revealed nailfold capillary abnormalities and dactylitis. Laboratory testing demonstrated a positive antinuclear antibody (ANA) of 1:1280 (Normal <1:40 or 1:80), with centromere pattern as well as anti–Scl-70 antibody positivity. In the context of her clinical and serologic findings, she met classification criteria for systemic sclerosis (SSc), formerly scleroderma. GAVE is a recognized vascular manifestation of systemic sclerosis and could have served as an earlier clue for an underlying connective tissue disorder.

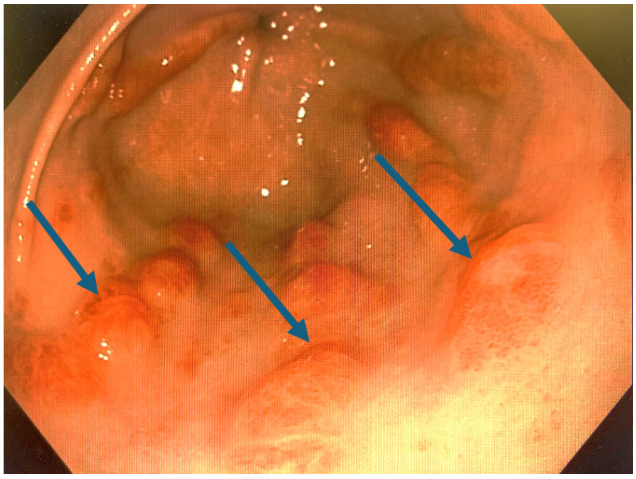

Figure 1. Gastric antral vascular ectasia (GAVE), characterized by longitudinal erythematous streaks radiating from the pylorus through the antrum, producing the classic "watermelon stomach" appearance.

DISCUSSION

GAVE is an uncommon but important cause of upper gastrointestinal bleeding, accounting for up to 4% of non-variceal upper gastrointestinal bleeding cases.[1] Diagnosis is made by endoscopic visualization of longitudinal rows of flat, red stripes radiating from the pylorus to the antrum, resembling a watermelon. On biopsy, findings include vascular ectasia, spindle cell proliferation, and fibrohyalinosis. GAVE is most frequently associated with cirrhosis (around 30%) and end-stage liver disease (ESLD) of whom 1 in 40 have GAVE).[2] The next most common association with GAVE is autoimmune disease. In an observational case series of 45 patients treated for GAVE, 62% had some form of connective tissue disorder. It is also seen disproportionately in patients with metabolic syndrome.[3]

The etiology of GAVE is thought to involve multiple mechanisms. Initially, it was believed to stem from mechanical injury and hormonal changes. The location of the lesions in the antrum may be related to abnormal gastric motility and antral distention, leading to prolapse of the antrum through the pylorus and resulting in repetitive trauma. Elevated levels of Prostaglandin E2, a known vasodilatory hormone-like substance, have been observed in cirrhotic patients with GAVE compared to those without GAVE, potentially linking hormonal changes in liver disease to its pathogenesis. More recently, the concept of microvascular injury leading to neoangiogenesis has emerged. This mechanism is proposed to play a role in metabolic syndrome and possibly in systemic sclerosis.[4]

The clinical presentation of GAVE ranges from chronic iron deficiency anemia secondary to occult gastrointestinal blood loss, as in this case, to severe acute upper gastrointestinal hemorrhage requiring urgent intervention, although the latter is less common. Management primarily consists of endoscopic therapy, most commonly argon plasma coagulation, which can effectively control bleeding. However, recurrence is common, and patients often require repeated treatments. Interestingly, some case reports describe scleroderma patients whose GAVE improved with treatment of other scleroderma-related complications, such as lung disease.[5]

The prevalence of GAVE in SSc patients is estimated to range from 5.6% to 10.7%, depending on the retrospective study and the number of cases examined. The higher end

of this range is likely more accurate, as endoscopy is typically performed only in symptomatic patients, leaving some cases undiagnosed. In SSc patients, GAVE is associated with older age at disease onset, diffuse disease subtype, negative Scl-70, Anti-U1 ribonucleoprotein, and Anti-Polymyositis/Scl antibodies, and positive RNAP III antibodies.[6]

Finally, SSc patients with digital ulcers, delayed gastric motility, and higher nailfold video capillaroscopy scores have higher rates of GAVE. Specifically, nailfold video capillaroscopy scores correlate with risk and may serve as a potential screening tool earlier in the evolution of the disease.

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