Case Report

Gastric Antral Vascular Ectasia (Watermelon Stomach) in a Patient with Sjögren's Syndrome

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ABSTRACT. Gastric antral vascular ectasia (GAVE), a rare yet treatable cause of upper gastrointestinal bleeding, has been described in a variety of autoimmune diseases. We describe a patient who had typical Sjögren's syndrome and iron deficiency anemia requiring blood transfusion. An endoscopy showed characteristic findings of GAVE. After several fulguration therapies with argon-plasma coagulator, the mucosal lesions improved and her hemoglobin levels returned to normal. (J Rheumatol 2003;30:1090–2)

Key Indexing Terms:
SJÖGREN'S SYNDROME GASTRIC ANTRAL VASCULAR ECTASIA
WATERMELON STOMACH

Although occurrence of gastric antral vascular ectasia (GAVE) in sicca syndrome has been described[1], a Medline search using the keywords "Sjögren's syndrome" or "sicca syndrome" and "gastric vascular antral ectasia" or "watermelon stomach" did not reveal positive results. We describe an elderly Caucasian woman previously diagnosed with Sjögren's syndrome (SS) who developed iron deficiency anemia secondary to GAVE.

CASE REPORT

A 74-year-old white woman was seen by her primary care physician in 1996 for a one year history of ocular and oropharyngeal dryness associated with upper esophageal dysfunction, burning sensation of eyes, and bilateral parotid gland swelling. An ENT consultant performed a lower lip biopsy that showed inflammatory cells in minor salivary glands. The antinuclear antibody (ANA) was positive (1:320 nucleolar pattern), rheumatoid factor was positive (23 IU/ml); Sjögren's antibodies (< 0.91) were not detected. The pathology on an excisional biopsy of 6.5 × 3 × 1.5 cm right parotid mass reported marked chronic inflammation, lymphoid nodule hyperplasia, myoepithelial islands, and a benign lymphoepithelial cyst with adjacent acute and chronic inflammatory infiltrate. The patient was treated with artificial tears and frequent oral hydration. Subsequently, in 1998, because of persistent tender swelling of the left parotid gland, an excisional biopsy was performed that also revealed histological changes characteristic of SS.

In 1999, during hospitalization for gradually progressive exertional dyspnea (ongoing for 4 years), her hemoglobin (8.2 g/dl) was notably decreased from a baseline of 13.2 g/dl nine months previously. She also had heme-positive stools. She was given blood transfusions. Benign polyps seen on colonoscopy were removed. Radiographic and pulmonary function studies were characteristic of diffuse interstitial pneumonitis. She was given colchicine 0.6 mg/day and oxygen by nasal canula. She continued to feel weak and dyspneic.

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An upper endoscopy showed multiple angiectasias in a linear radiating pattern (Figure 1), without bleeding, in the gastric body and antrum. Congestion and edema was noted in the duodenal bulb. The esophagus appeared normal.

There was no history of peptic ulcer disease other than a small hiatal hernia seen on a barium study of the upper gastrointestinal (GI) tract in 1995. Her liver function tests were abnormal: SGOT 56 (normal 14–33), SGPT 61 (10–42), alkaline phosphatase 181 (44–128), albumin 3.6. The serum bilirubin was normal. Hepatitis A, B, and C serologies were negative, as were assays of multiple autoantigens including smooth muscle, double-stranded DNA, Smith, RNP, and Scl-70. Multiple tests for anti-SSA (Ro) and anti-SSB (La) were also negative. There was no history for vascular events or pregnancy related morbidity, IgG antiphospholipid assay was negative, IgM (19 MPL) and IgA (23 APL) antiphopholipid tests were mildly elevated.

She underwent argon-plasma fulguration of the gastric mucosa several times over 9 months, and subsequent endoscopic evaluation showed diffuse hemorrhagic mucosa with no evidence of active bleeding. She subsequently developed reflux esophagitis that was successfully managed with omeprazole. The clinical course for more than a year was stable; hemoglobin levels increased to normal range and prominent sicca symptoms persisted. Then manifestations of diffuse pulmonary disease began to progress in spite of high doses of prednisone and azathioprine. She died 20 months after the completion of argon-plasma coagulation therapy. Permission for postmortem examination was not obtained.

DISCUSSION

The most common GI manifestation in SS is dysphagia associated with xerostomia. In addition, chronic atrophic gastritis[2], clinically insignificant mild elevation of liver enzymes[3], primary biliary cirrhosis, sclerosing cholangitis, and subclinical pancreatic disease[4] have been observed with increased frequency in patients with SS. Vasculopathy with SS, commonly manifesting as small vessel cutaneous vasculitis, is rare in other organ systems[5,6].

The clinical spectrum of GAVE was formally recognized first in 1984. The diagnostic hallmark of the disease is based on characteristic endoscopic findings: visible columns of blood vessels traversing the antrum in longitudinal folds and converging in the pylorus. Because the longitudinal red columns collectively resemble the stripes of a watermelon,

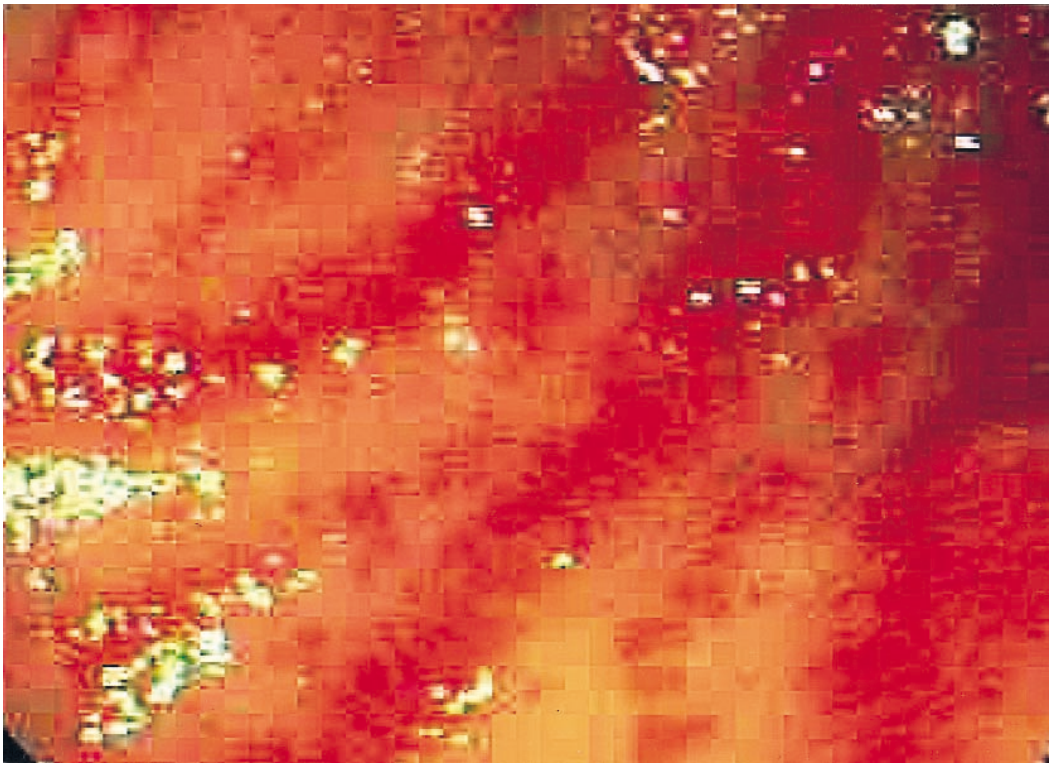

Figure 1. Gastric endoscopy shows multiple angiectasias in a linear radiating pattern.

Jabbari, *et al* coined the term "watermelon stomach" to describe this condition. Histology confirms the vascular basis of the disorder, showing dilated and thrombosed capillaries in the lamina propria, associated with reactive fibromuscular hyperplasia.

GAVE is rare in SS. It is known to occur with various diseases — scleroderma[7], cirrhosis, atrophic gastritis, hypothyroidism, diabetes mellitus, pernicious anemia, and others. The finding of GAVE along with other overlapping features in many of these diseases highlights their autoimmune background. In scleroderma, the vascular changes are typical of those described in GAVE in nonscleroderma patients; in addition, small vessel fibrin deposition has been seen. This patient had a nucleolar pattern ANA, most frequently associated with scleroderma, yet no clinical features of systemic sclerosis[8]. Overlap between SS and systemic sclerosis is a distinct possibility since there was pulmonary fibrosis, which is more frequent in systemic sclerosis than SS.

Several hypotheses have been put forth to explain the pathogenesis of watermelon stomach, which remains unknown. It is believed that a complex interplay between altered histochemical physiology and traumatic mechanical factors may exist. It has been postulated that vascular alterations may be related to excessive production of vasoactive mediators by hyperplastic neuroendocrine cells in the gastric mucosa[9]. However, immunohistochemical studies in gastric tissue failed to confirm this hypothesis[10]. Among traumatic factors, sustained antral contractions and diaphragmatic hernia have been proposed as potential mechanical causes of the disease. Argon-plasma coagulation, a noncontact electrocoagulation technique, is emerging as a safe, tolerable, and effective laser treatment for GAVE[11].

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