Watermelon stomach in a patient with primary Sjögren's syndrome

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Abstract

**Introduction.** Watermelon stomach (WS) or gastric antral vascular ectasia (GAVE) is a rare cause of upper gastrointestinal bleeding described in a variety of autoimmune disorders. Association of watermelon stomach with Sjögren's syndrome is extremely rare. **Case report.** We presented a 67-year old female with primary Sjögren's syndrome (SS) who had developed a persistent severe iron-deficiency anemia. An upper gastric endoscopy revealed the presence of gastric antral vascular ectasia (GAVE) as a cause of occult gastrointestinal bleeding. The treatment with argon-plasma coagulation was postponed as the conservative therapy with iron substitution and proton pump inhibitor led to improvement of anemia and hemoglobin levels normalization. **Conclusion.** This is the first report of WS in a patient with primary SS without the presence of coexisting autoimmune disorder. Recognition of this rare, but clinically important, cause of gastrointestinal bleeding may decrease comorbidity in patients with autoimmune disorders including primary Sjögren's syndrome.

Key words: sjogren's syndrome; gastric antral vascular ectasia; anemia, iron-deficiency; drug therapy; treatment outcome.

Introduction

Gastric antral vascular ectasia (GAVE) or watermelon stomach (WS), is a rare cause of upper gastrointestinal bleeding. This entity was first described in 1953, while Jabbari et al. in 1984 introduced nowadays widely used term “watermelon stomach” for the endoscopic presentation of characteristic linear, radially distributed vascular ectasia of the gastric antrum that resembled the stripes on a watermelon. WS has been described in a variety of autoimmune diseases and is typically presented with iron-deficiency anemia and positive fecal occult blood test, as well as melena and hematemesis.

Sjögren's syndrome (SS) may occur as a primary disorder, or in association with other connective tissue diseases, as a secondary SS. Angiodysplastic lesions in SS are frequent and rare occurrence of WS has been reported in sicca syndrome.

In this paper we presented for the first time a female patient with primary Sjögren’s syndrome without coexisting...
autoimmune disorder who developed severe iron-deficiency anemia secondary to GAVE.

Case report

In January 2005, a 65-year-old Caucasian female was referred with a six-month history of fatigue, arthralgia and dryness of the mouth and eyes. Her medical history was unremarkable. Physical exam revealed no abnormalities. Laboratory investigation disclosed a high erythrocyte sedimentation rate (ESR 110 mm/h), normocytic anemia with hemoglobin (Hb) levels of 110 g/L and mean corpuscular volume (MCV) of 84 fL, leucopenia (3.2 × 10³/µL), lymphopenia (1.0 × 10³/µL), hypergammaglobulinemia with an IgG of 19.6 g/L (normal: 7.0–16.0 g/L), positive IgM rheumatoid factor (RF) (95 U/L, normal < 25 U/L), positive antinuclear antibodies (ANA titre 1:160, speckled pattern) and a high level of anti-Ro/SS-A antibodies (> 200 U/mL). Antibodies against centromere (ACA), La/SS-B, RNP, Sm, Scl-70, dsDNA, cyclic citrullinated peptide, cardiolipin and ANCA were all negative. Cryoglobulins were not detected, and C3 and C4 levels were normal. Other laboratory findings, including renal function tests, thyroid hormones and autoantibodies were normal, and routine microbiological tests including HBs antigenemia, anti-HIV and anti-HCV antibodies were negative. Bilateral keratoconjunctivitis was diagnosed along with positive Shimer’s and Rose-Bengal tests. Scintigraphy of the salivary glands revealed impairment of accumulation, delayed uptake and excretion of the tracer. Diagnosis of primary SS was established according to the proposed criteria 7, and the treatment with oral prednisolone (30 mg/day, and than tapered off) along with artificial tears and frequent oral hydration started. The administered therapy led to gradual improvement of the patient’s general condition. In August 2007 the patient complained of a severe fatigue, non-swollen, tender radiocarpal and proximal interphalangeal joints and occasional dull, non-specific epigastric pain. The laboratory examination showed moderately elevated ESR (38 mm/h) and microcytic hypochromic anemia with Hb levels of 111 g/L and MCV of 68 fL, with normal white blood cell and platelet counts. Serum iron (5.5 µmol/L) and ferritin (5.0 µg/L) levels were low. Immunological tests revealed high levels of anti-Ro/SS-A and anti-La/SS-B antibodies. In September 2007, laboratory investigation disclosed elevated ESR (46 mm/h) and progression of iron-deficiency anemia with Hb 95 g/L, MCV 62 fL, serum iron concentration of 3 µmol/L and ferritin of 3 µg/L. Fecal occult blood test was found positive. The thorough examination of the digestive tract was undertaken and upper endoscopy revealed typical features of gastric antral vascular ectasia or watermelon stomach (Figure 1). Transabdominal ultrasonography, colonoscopy and capsule video endoscopy findings were all normal.

Additional examinations were performed to exclude coexisting systemic sclerosis (SSc). Nailfold capillaroscopy gave no evidence of microangiopathy or capillary loops enlargement. Chest X-ray was normal. Respiratory function tests showed mildly lowered transfer factor for carbon monoxide and total lung capacity measured with helium dilution, whereas diffusion coefficients for carbon monoxide were normal (DLco 82%, DLco/VA 98%, TLC 86%). Chest multislice computerized tomography examination showed no signs of interstitial fibrosis, infiltration and consolidation of lung parenchyma, pleural effusion or mediastinal lymphadenopathy.

The patient received conservative therapy with iron supplementation (Fe²⁺-gluconate, 100 mg/day) for three months and proton pump inhibitor ( pantoprazole, 40 mg/day) for one month. The treatment with argon-plasma coagulation was postponed. In the eight-month follow-up, the patient did not develop any symptoms consistent with SSc, including Raynaud phenomenon. Hematological parameters returned to normal (Hb 131 g/L, MCV 82 fL, serum iron concentration 10.75 µmol/L, ferritin 15.0 µg/L) and up to date the patient remains free of gastrointestinal symptoms. During the follow-up period, the patient received the therapy with hydroxychloroquine (200 mg/day) and her SS showed no aggravation.

Discussion

The presented elderly female patient, previously diagnosed with primary Sjögren’s syndrome, had developed a severe iron-deficiency anemia secondary to GAVE or watermelon stomach.

Watermelon stomach is a rare cause of chronic occult or potentially fatal gastrointestinal bleeding and is typically presented as angiodyplastic lesions, with characteristic radially distributed, dilated and thrombosed capillaries in the antral part of the stomach. The etiology of WS remains unknown. WS has been related to mechanical causes and to delayed hypergastrinemia, prostaglandin E2, 5-hydroxytryptamine and vasoactive intestinal polypeptide production by neuroendocrine cells in the gastric mucosa.
GAVE is often associated with systemic illnesses, such as autoimmune connective tissue disorders including systemic sclerosis, bone marrow transplantation, pernicious anemia, atrophic gastritis, diabetes mellitus and cirrhosis of the liver. Angiectasia is very rarely observed in primary SS. The infrequent vascular involvement in SS is most commonly presented as small cutaneous vessel vasculitis with cryoglobulinemia and is rare in other organ systems. The association of watermelon stomach with primary Sjögren’s syndrome has not been reported. Goustout et al. described a patient with sicca syndrome and WS while Goel et al. described WS in a female patient with Sjögren’s syndrome who subsequently developed fatal interstitial pulmonary fibrosis due to the coexisting systemic sclerosis (SSc). Although watermelon stomach is a rare syndrome, it is recognized as a cause of persistent bleeding in patients with SSc and according to published reports, WS may precede SSc by several months or years. The overlap between SS and systemic sclerosis in our patient was excluded since no evidence of Raynaud phenomenon, signs of pulmonary fibrosis nor positive immunological markers (nucleolar ANA pattern, ACA, ScI-70) were recorded. Apart from ocular and oral symptoms, our patient did not present with any significant systemic manifestations, except occasional arthralgia. Anaemia is not uncommon in patients with SS, but it is usually normocytic and normochromic. The presence and progression of iron-deficiency anaemia is not a predicted feature of impairment of the underlying disease, with regard to corticosteroid and antimalaric treatment. In addition, a detailed examination of the gastrointestinal tract were undertaken to exclude other conditions that might incite the development of WS.

Therapeutic options for WS are variable and depend on the severity and progression rate of iron-deficiency anaemia. Encouraging effects of corticosteroid treatment leading to cessation of bleeding and improvement of anaemia has been repeatedly reported. Recent studies suggest efficacy of endoscopic ablation procedures in management of bleeding lesions including argon-plasma coagulation and coagulation with Nd: YAG laser. In mild forms, as described in our patient, conservative treatment with iron substitution and proton pump inhibitor led to a long-term normalization of anaemia and improvement of her mucosal lesions.

**Conclusion**

Although rare, WS should be considered as a potential cause of iron-deficiency anaemia secondary to gastrointestinal bleeding in patients with autoimmune conditions including primary Sjögren’s syndrome.

**References**

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The paper received on March 3, 2009. Accepted on April 23, 2009.