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Internal Medicine

GASTRIC ANTRAL VASCULAR ECTASIA IN SINE SCLERODERMA: A CASE REPORT AND REVIEW OF LITERATURE"

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ABSTRACT Gastric Antral Vascular Ectasia (GAVE) is a rare clinical entity with varied clinical presentation ranging from chronic asymptomatic phase to acute or chronic gastrointestinal hemorrhages and iron deficiency anemia. GAVE has been described in association with many medical conditions like hepatic cirrhosis, chronic kidney disease, chronic pulmonary disease, hypertension, ischemic heart disease, post-bone marrow transplantation, diabetes and many autoimmune diseases. We report a case of seventy five year old female who presented to us with dyspnea on exertion and transfusion dependent refractory anemia. She was investigated and was found to have Systemic sclerosis sine scleroderma with GAVE

KEYWORDS : GAVE, systemic sclerosis sine scleroderma, refractory anemia, autoimmune diseases.

BACKGROUND:

Gastric Antral Vascular Ectasia (GAVE) is a rare clinical entity with varied clinical presentation ranging from chronic asymptomatic phase to acute or chronic gastrointestinal hemorrhage and iron deficiency anemia. Rare presentations of GAVE include shortness of breath and right upper quadrant pain with vomiting. It was first described in 1953 by Rider et al [(1)] and has been described as a distinct pathological lesion by Jabbari et al. in 1984 [(2)]. GAVE is responsible for about 4% of non-variceal upper gastrointestinal hemorrhage[(3)].

GAVE has been described in association with many medical conditions like hepatic cirrhosis, chronic kidney disease, chronic pulmonary disease, hypertension, ischemic heart disease, post-bone marrow transplantation and diabetes. It has also been associated with autoimmune diseases, including Raynaud's phenomena, rheumatoid arthritis, polymyalgia rheumatica, primary biliary cirrhosis, and systemic sclerosis (SSc) [(4)(5)].

SSc is a connective tissue disorder of unknown aetiology characterized by hallmark skin thickening which distinguishes it from other connective tissue disorders. Based on skin involvement it is broadly classified into diffuse and limited variety. "Systemic sclerosis sine scleroderma" is a rare form of limited systemic sclerosis. These patients are without skin involvement, but have the similar clinical or laboratory features and prognosis compared to classical systemic sclerosis.

In the absence of cutaneous signs, its diagnosis is delayed leading to significant morbidity and mortality. We report a case of seventy five year old female who presented to us with dyspnea on exertion and transfusion dependent anemia. She was investigated and was found to have Systemic sclerosis sine scleroderma with GAVE.

Case report:

A 75 year old female who is known to have Systemic Hypertension, Type 2 Diabetes mellitus, and Hypothyroidism presented to us with generalized weakness, fatigue and dyspnea on exertion for past 6 months. She denied having any other complaints. Initially she was evaluated in a local hospital and was diagnosed to have severe anemia for which she received multiple blood transfusions.

Her hemoglobin at the time of admission was 6.9gm/dl with normal white blood cell count and platelet count. Other significant labs were low hematocrit (30%), high ESR (105mm/hr), Red cell distribution width of 19.7%, and reticulocyte count of 11%. Peripheral blood smear revealed normocytic normochromic red blood cells. Renal parameters and liver function tests were normal. Anemia work-up revealed iron deficient state (transferrin saturation 7.6% and serum iron - 31 µg/dL) and normal vitamin B12 levels (613 ng/L). Her serum Lactate dehydrogenase (LDH) was 155 U/L and Thyroid stimulating hormone (TSH) was 2.35 µIU/mL. Direct antiglobulin test was negative. Stool for occult blood was negative.

The patient was transfused with packed red blood cells. Oesophago gastroduodenoscopy (OGD) done previously revealed GAVE and endoscopic band ligation was done twice earlier. OGD was repeated here in view of recent drop in hemoglobin and worsening of exertional dyspnea and it revealed GAVE and argon plasma photocoagulation **(Figure: 1, 2 and 3)** was done for achieving haemostasis. Subsequently to rule out small bowel sources of bleeding capsule endoscopy was done and it was normal.

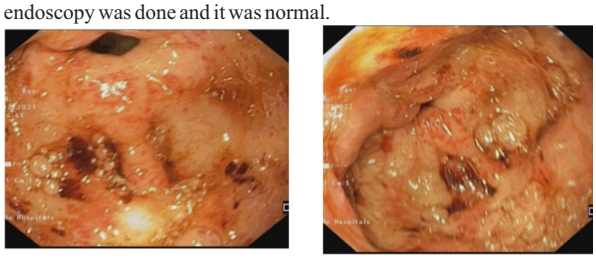

Figure1&2: OGDscopy of patient revealing gastric antral vascular ectasia with mild ooze

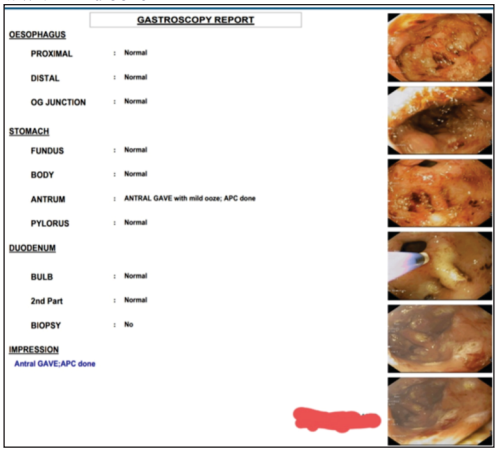

Figure: 3 showing Antral GAVE; APC done

In view of recurrent transfusion dependent anemia in an elderly women with background of GAVE requiring endoscopic band ligation/ argon plasma photocoagulation, autoimmune work-up was done which revealed ANA positivity with punctuate nucleolar pattern. Antigens associated with this pattern are RNA polymerase I and hUBF/NOR-90. Diseases usually associated with this pattern are Systemic Sclerosis and Sjogrens Syndrome. Rest of the autoimmune work-up was negative (ANCA, Extractable nuclear antigens (ENA) panel, Anti dsDNA). Her complement levels (C3, C4) were within normal limits.

Echocardiogram was done to rule out pulmonary hypertension, and it did not reveal any significant abnormality. Computed tomography (CT) scan chest was done to look for features of interstitial lung disease and it revealed moderate cardiomegaly, minimal subpleural reticulations in both lower lobes. Rest of the lung showed normal lung parenchymal pattern with even distribution of the bronchial tree. The pulmonary arteries appeared normal.

Here we report a patient of systemic sclerosis sine scleroderma with severe transfusion dependent anemia requiring multiple transfusions and specific serology for scleroderma in the absence of cutaneous manifestations.

DISCUSSION:

Systemic sclerosis typically affects skin and internal organs by widespread micro vascular damage and excessive collagen deposition [6]. There are scarce data in the literature for systemic sclerosis sine scleroderma. In a study from Brazil in 2013 out of 947 patients with systemic sclerosis, 79 (8.3%) were classified as having systemic sclerosis sine scleroderma [7].

Gastrointestinal complications are well recognized in systemic sclerosis. In general, these complications are sequale of gut dysmotility due to increased collagen deposition. GAVE is characterized by red stripes or multiple longitudinal folds from the pylorus through the antrum, defined as watermelon stomach, or arranged in a diffused-way, called as honeycomb stomach. Alternatively multiple red dots may be seen. The prevalence of GAVE in association with SSc is highly variable and it ranges between 1-76% in different studies [8,9,10].

The exact cause of GAVE is not clearly known. Several authors suggest that GAVE is a vascular manifestation of systemic sclerosis. This is supported by the finding that 60% of GAVE patients will develop vascular abnormalities like skin telangiectasias and renal artery stenosis. Additionally, they have histopathologic similarities in skin biopsies of patients with systemic sclerosis and in gastric mucosal biopsies from GAVE patients in terms of capillary dilation, fibrin deposits, and platelet thrombosis[11]. Also the events that are commonly associated with SSc-vasculopathy are also associated with GAVE, such as systemic hypertension, renal crisis and reduced diffusing capacity of the lung divided by alveolar volume (DLCO/AV) [10].

Some authors propose that a loose connection between the distal gastric mucosa and the adjacent muscularis externa can cause prolapse of the antral mucosa in the pylorus and result in development of GAVE [12,13]. Few authors have linked GAVE with an autoimmune process due to the fact that GAVE has been associated with other autoimmune diseases in population studies. Several autoantibodies have been detected in patients with GAVE: antinuclear antibodies (ANAs), anti-Centromere and anti-RNA helicase II, especially in patients diagnosed with systemic sclerosis and GAVE. SSc-associated GAVE is usually negative for SCL-70. In one study among 49 SSc-associated GAVE, only one was found to be positive for Scl-70; similar to this finding our patient was found to be negative of Scl-70[10]. Scleroderma with GAVE have female preponderance and is usually diagnosed in elderly patients both of these findings are in concordance with our patient findings.

The primary physician must suspect GAVE when the anemia is refractory to regular treatment, and patient should be screened with endoscopy for GAVE and in case of confirmation of endoscopic GAVE; the patient must be evaluated for underlying cause and treated accordingly. Endoscopic laser therapy is effective in the treatment and prevention of GI bleeding related to GAVE. The technique most commonly used is argon plasma coagulation (APC) and is considered one of the best endoscopic therapeutic options which showed significant improvement in our patient. She is under primary physician and rheumatology follow-up for the last 6months and has not developed any skin thickening or other features of scleroderma.

CONCLUSION:

GAVE in patients with systemic sclerosis is a rare and poorly understood condition, but can be properly diagnosed and successfully treated. Early diagnosis is crucial in the management of GAVE because it makes symptomatic therapies and endoscopic approaches feasible. The first step is to have a high index of suspicion for changes in hemoglobin levels or clinical features of gastrointestinal bleeding. There are reported cases of GAVE as the presenting and only manifestation of systemic sclerosis. Studies have shown that the majority of patients with GAVE, around 60%, have telangiectasias of the skin. But in our patient, detailed physical examination did not show any cutaneous changes, which would result in delay of diagnosis if the treating physician is not aware of this association. Scleroderma should be considered in the aetiology of GAVE even in the absence of skin thickening.

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- **Competing interests** -- The authors declare that they have no competing interests.
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