Gastric antral vascular ectasia in systemic sclerosis: complete resolution with methylprednisolone and cyclophosphamide

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Abstract
A case of severe, transfusion dependent anaemia in a 72 year old woman, which on endoscopy was found to be due to gastric antral vascular ectasia (GAVE), is reported. Repeated endoscopic sclerotherapy was ineffective. She subsequently developed Raynaud’s phenomenon and on further investigation was found to have classical systemic sclerosis with lung involvement. Treatment with pulses of intravenous methylprednisolone and cyclophosphamide resulted in significant improvement in her pulmonary function tests and skin score. Coincidentally, her haemoglobin stabilised and further endoscopic examinations were normal. This is the first report of cyclophosphamide and methylprednisolone leading to complete and sustained resolution of GAVE in association with systemic sclerosis.

Gastric antral vascular ectasia (GAVE) or “watermelon stomach” has been recognised as a distinct pathological lesion since 1984. GAVE exclusively a gastrointestinal (GI) complication of vascular abnormalities seen in systemic sclerosis. GAVE has been recognised in association with scleroderma and, possibly, the antral lesions may represent part of the spectrum of vascular abnormalities seen in systemic sclerosis.

Case report
A 72 year old Caucasian woman was admitted to the care of the gastroenterologists in June 1996 with an eight week history of lethargy and shortness of breath on exertion. A full blood count showed microcytic hypochromic anaemia (haemoglobin 66 g/l). She denied any GI symptoms. She had a previous history of vitiligo and polymyalgia rheumatica, for which she was taking 5 mg of enterically coated prednisolone.

Abdominal examination was unremarkable, though laboratory tests for faecal occult blood were positive. Endoscopic examination and gastric biopsies supported a diagnosis of chemical gastritis. Iron treatment was started after transfusing three units of blood.

Four weeks later the same patient was re-admitted with symptomatic anaemia. Repeat endoscopy confirmed the previous findings. After a further blood transfusion she was discharged. Four weeks later she again presented with anaemia. On this occasion she was noted to be deficient in vitamin B₁₂ with positive gastric-parietal antibodies and was also found to be hypothyroid. Thyroxine and B₁₂ replacement were started after a further blood transfusion. She had a normal colonoscopy and small bowel meal and follow through. Three months later she was anaemic again. A small bowel enteroscopy ruled out angiodysplasia but noted appearances typical of GAVE or “water melon stomach” (fig 1A), and this diagnosis was confirmed on histology.

Over the following six months she had 19 therapeutic endoscopic examinations with sclerotherapy in an attempt to control her chronic blood loss. Over the same period 42 units of blood were transfused (fig 2).

In April 1997 she developed Raynaud’s phenomenon. Her skin was noted to have become taut and the underlying tissue was oedematous. She had lost 20 kg in weight in the preceding six months and had remained persistently anaemic. At this stage she was referred to the rheumatologists.

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Examination of the skin showed extensive scleroderma affecting the limbs and chest wall. Pulmonary function tests showed a restrictive deficit with a reduced transfer factor. Thoracic high resolution computed tomography showed changes consistent with pulmonary fibrosis. Antinuclear antibodies were weakly positive (titre 1/40), but extractable nuclear antibodies were negative, including antitopoisomerase (Scl-70). She was admitted to hospital and a course of pulsed, intravenous methylprednisolone (10 mg/kg) with cyclophosphamide (10 mg/kg) was started according to a standard protocol, with mesna as a urothelial protective agent.

Over the next three months her scleroderma improved dramatically. It was noted that co-incident with the start of treatment for her scleroderma she had ceased to require blood transfusions. Repeat endoscopy was normal with no evidence of GAVE. After a further six months of treatment she has not required further blood transfusion and remains well with a stable haemoglobin and normal endoscopic appearance (fig 1B).

Discussion

GAVE was first described in 1984. In 1992 Goustout described a series of 45 patients with recurrent iron deficiency anaemia and GAVE. In that series 72% of the subjects were female and their mean age was 73 years. Sixty two per cent were transfusion dependent, requiring a mean of 10 units of blood over one year. Interestingly, they found a remarkably high incidence (62%) of autoimmune and connective tissue disease in these patients. Scleroderma was reported in eight of the 45 patients (18%). Although GAVE had been previously reported in association with systemic sclerosis, this
The treatment of GAVE is usually endoscopic heater probe cautery or laser coagulation. A significant number of patients proceed to partial gastrectomy. It is interesting to speculate that in our patient a combination of autoimmune gastric atrophy and the vascular abnormality peculiar to systemic sclerosis resulted in the florid antral changes of GAVE. There is scope for further study to determine whether scleroderma associated GAVE without pernicious anaemia, or, indeed, non-autoimmune disease associated GAVE, is as sensitive to immunosuppression. Clearly, if our finding could be reproduced in others it might obviate the need for multiple transfusions, repeated endoscopic intervention, and major surgery in this patient group.

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