

Session 6: Difficult Cases

CASE 4: Recurrent Anaemia

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&

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Miss Joan Marple, a 64 y.o. female

- Referred to centre for the first time by GP
b/o increasing dyspnoea
- Longstanding limited SSc (Δ 1992)
- Raynaud's since age 29
- Recurrent "finger sores"
- "High blood pressure"
- Long-standing "kidney problem"
- Replacement of right hip joint 3 months ago
- Last referral to a rheumatologist >6 months ago

QuickTime™ and a
decompressor
are needed to see this picture.

Presenting complaint:

- Dyspnea at rest

Further symptoms:

- palpitations
- occasional “dizzy spells”
- difficulty walking
- fatigue
- feeling generally unwell
- despite improved RP and healed ulcers

- General:

64 y.o. female, chronically ill, no acute distress

- Vitals:

b.p. 150/90 mmHg, pulse 88/min and regular,
resp. rate 24/min., temperature 36.9° C

- Skin:

no pallor, no cyanosis, no jaundice,
multiple telangiectasias,
healed digital ulcers, calcinosis cutis,
no palpable lymph nodes

- Cardiopulmonary:

normal breath sounds,
no wheezing/crackles,
no murmurs, mild ankle oedema,
6MWT: 400 m



- Abdomen:
soft; mild epigastric tenderness; no masses;
normal peristalsis
- Musculoskeletal:
Heberden nodules, tenderness of the knees
- Neurological:
no sensory/motor deficits,
normal reflexes

Pulmonary Function Test:

- FVC: 2.67 L (112%)
- DLCO SB: 6.17 (92%)

Echocardiography:

- Mildly dilated left ventricle
- Normal valves
- PAP: 45 mm Hg

QuickTime™ and a
decompressor
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FBC and differential:

Erythrocytes: 3.2/pl

Leukocytes: 7.2/nl

Thrombocytes: 325/nl

Reticulocytes: 5‰

Haemoglobin: 6 mg/dl

Haematocrit: 19%

MCV: 65 fl

MCH: 21 pg

Chemistry:

Sodium/potassium/calcium: within normal range

Creatinine: 2.1 mg/dl

Transaminases: within normal range; LDH 280 U/l

Iron: 12 µg/dl; ferritin: 11 ng/ml

TSH and glucose within normal range

ESR: 53 mm/h; CRP: 12 mg/l

ACA: positive

DD of anaemia

- Vascular:

Vasc. malformations, oesophageal varices, PHG

- Infectious:

Blind loop syndrome, chronic infections

- Traumatic/Toxic:

Lead/arsenic poisoning, drug-induced hemolysis, GIT surgery, menstrual bleeding

- Autoimmune:

Pernicious anaemia, connective tissue disease, IBD, hypersplenic syndrome, hemolytic anaemia

QuickTime™ and a
TIFF (Uncompressed) decompressor
are needed to see this picture.

DD of anaemia:

- **Metabolic/hereditary:**

A-/hypochlorhydria, pancreatic insufficiency,
Chronic renal disease, thalassaemia,
Sickle cell disease

- **Inflammatory:**

Ulcerative gastritis

- **Neoplastic:**

Hematologic malignancies, GIT tumors,
Solid tumours

- **Degenerative/dietary:**

Vegetarians/vegans, alcohol abuse, folic acid deficiency,
malabsorption

QuickTime™ and a
TIFF (Uncompressed) decompressor
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DD of anaemia in SSc patients:

- Malabsorption
- Bleeding:
 - > ulcers
 - > diverticula
 - > hiatal hernia
 - > neoplasms
 - > watermelon stomach

Rationale:

- Microcytic, hypochromic anaemia
- Low normal reticulocytes
- Normal haptoglobin and LDH
- Low ferritin in a 64 yo woman

--> **Occult blood loss**

Test for occult blood in the stools:
positive

Colonoscopy:

Hemorrhoids grade II, non-inflamed diverticuli

Gastroscopy:

Test for occult blood in the stools:
positive

Colonoscopy:

Hemorrhoids grade II, non-inflamed diverticuli

Gastroscopy:

QuickTime™ and a
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Diagnosis

Watermelon stomach
or
Gastric antral vascular ectasia (GAVE)

Introduction

History:

- 1953: Ryder et al. report first case
- 1984: Jabbari et al. describe histological characteristics

Epidemiology:

- 76% women; mean age 69.1 years (range 42-89 y.)
- up to 4% of all non-variceal upper GI bleedings
- associated with autoimmune disease (62%) and other conditions:

Raynaud's syndrome	autoimmune liver disease
SSc	primary biliary cirrhosis
atrophic gastritis	liver cirrhosis/portal hypertension
Addison's disease	chronic renal failure
hypothyroidism	cardiovascular disease
diabetes mellitus	familial Mediterranean fever

Introduction

Clinical presentation:

- **Iron deficiency anaemia** 88%
median Hb 6,7 g/dl (range 4,1-11,6)
- Heme positivity 42%
- Melaena 15%
- Haematemesis 3%
- Haematochezia 1%

Diagnosis

- **Esophago-gastroduodenal endoscopy**
- Barium meal, CT, MRT, angiography NOT USEFUL
- Endoscopic U/S may prove useful

Recurrent anaemia

Fecal occult blood positive



EGD

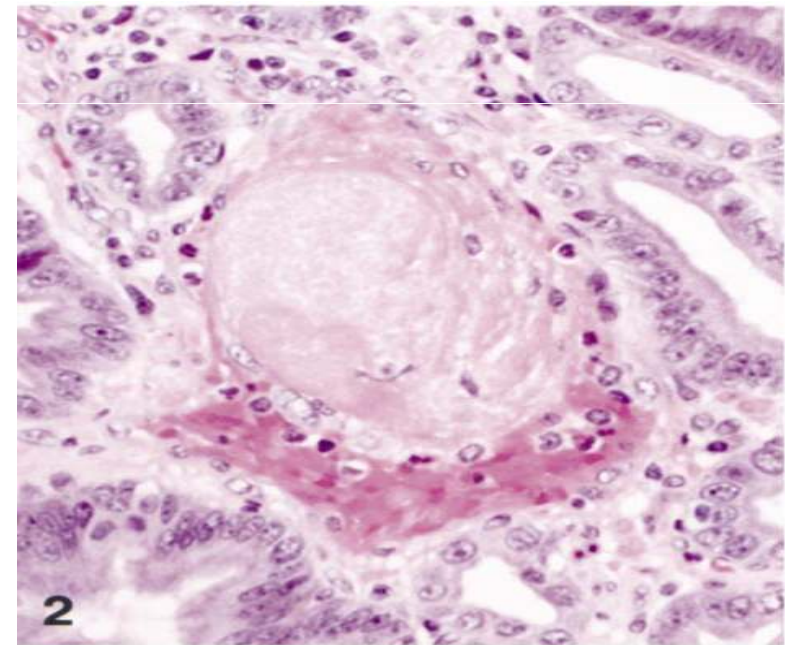
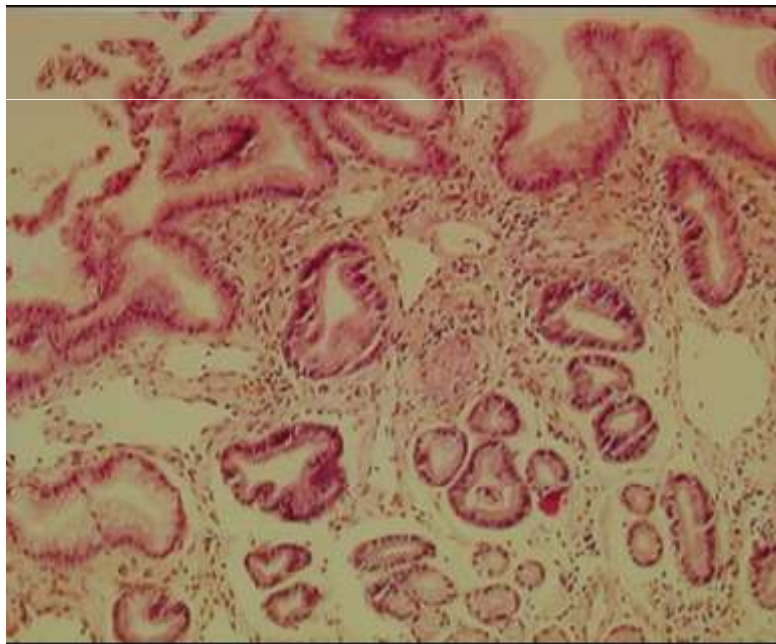
Endoscopic appearance

- Erythematous longitudinal stripes
- Radiating from the antrum to the pylorus



Histology

- Capillary ectasia
- Focal intravascular fibrin thrombi
- Fibromuscular hyperplasia in the lamina propria



Pathogenesis

- **Unknown**

- Two hypotheses:

1. Antral mucosal prolapse through the pylorus

+

Disordered gastric muscular motility

⇒ Submucosal ischemia

⇒ Elongation/ dilatation of mucosal vessels

2. GAVE as part of SSc-associated diffuse cutaneous teleangiectatic lesions

Treatment Options

- Conservative
- Endoscopic
- Surgical

Conservative Treatment

General measures in GI bleeding:

- Transfusion of packed red blood cells for acute GI bleeding and/or poorly tolerated anemia
- PPI (20-40 mg/day)

Specific measures for GAVE:

- Hormonal therapy (estrogen and progesterone)
- Octreotide
- Steroids
- (Tranexamic acid)

Endoscopic Treatment

- Nd:YAG laser
- Argon plasma coagulation
- Heater probe
- Bipolar electrocoagulation
- Injection sclerotherapy

Surgery

- Antrectomy
- Gastrectomy

Second line after failed endoscopic treatment

GAVE and SSc

- Association with SSc:
57% lSSc, 43% dSSc
dSSc with anti-Scl-70 --> 9,4% (negative predictor?)
lSSc with ACA --> 50%
- Usually more often diagnosed in patients with confirmed SSc
- May be an early manifestation of SSc:
<5 years after SSc diagnosis

Thank you for your attention!



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