Final diagnosis

Encephalotrigeminal angiomas (Sturge-Weber syndrome).

Keywords: encephalotrigeminal angiomas; Sturge-Weber syndrome; nasal discharge

- 1 Fötter R, Sager WD, Schneider GH, *et al.* Computer tomography in the Sturge-Weber syndrome. *Computertomographie* 1981;1:80-2.
- 2 Marti-Bonmati L, Menor F, Mulas F. The Sturge-Weber syndrome: correlation between the clinical status and radiological CT and MRI findings. *Childs Nerv Syst* 1993;9:107-9.
- 3 Holmes GL. Surgery for intractable seizures in infancy and early children. *Neurology* 1993;43:S28-S37.
- 4 Mackay I. Infective rhinitis and sinusitis. In: *Scott-Brown's Otolaryngology*, 6th edn, vol 3. Butterworths, London, 1997; pp 810-35.
- 5 Stammberger H. Endoscopic endonasal sinus surgery. Concept in treating of recurrent rhinosinusitis. Part I. Anatomic and pathophysiologic considerations. *Otolaryngol Head Neck Surg* 1986;94:143-7.

Sudden-onset watery diarrhoea in a middle-aged woman

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A 55-year-old woman presented with a 2-month history of profuse watery diarrhoea. The onset of diarrhoea was very sudden – she was woken up at 04.00 h one morning with a severe urge to move her bowels. The motions were watery, frequency averaging five times per day. There was no rectal bleeding, abdominal pain or vomiting. She had lost 2 kg in weight but there was no history of iritis, arthritis, rashes or aphthous ulcers. Clinical examination was unremarkable. Laboratory investigations revealed normal full blood count, urea and electrolytes. C-Reactive protein was not raised and there was no growth on stool culture. Colonoscopy showed mild erythema with loss of vascular pattern in the transverse colon, but no ulceration. Multiple biopsies were taken from the region. Histology of the colonic biopsy is shown in the figure.

Questions

- 1 What is the diagnosis?
- 2 What is the pathogenesis of this condition?

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Figure Histology of the colonic biopsy

Answers

QUESTION 1

The diagnosis is collagenous colitis. Histologically, there is a characteristic, thick, subepithelial collagen band. There is a chronic inflammatory cell infiltration of the lamina propria, but the crypt architecture is well maintained.

QUESTION 2

It has been postulated that an initial insult by a luminal agent, which may be an infective agent, gluten, or a drug, eg, a non-steroidal anti-inflammatory, may damage the bowel mucosa leading to an autoimmune response, with ongoing chronic inflammation in susceptible individuals.¹

Discussion

Collagenous colitis was first described by Lindstrom in 1976.² He reported a patient with chronic watery diarrhoea in whom a subepithelial collagen band was seen on rectal biopsy. More recent reports of collagenous colitis have suggested that it is not as rare as initially supposed.³⁻⁶ There is another group of patients with colorectal biopsies revealing changes very similar to collagenous colitis in terms of lymphocytic inflammation, but without a collagen band. To reduce confusion the term 'lymphocytic colitis' has been suggested to describe this entity. 'Microscopic colitis', in turn, is a general term denoting conditions characterised by microscopic colonic inflammation, which includes the subsets of collagenous and lymphocytic colitis.¹

Collagenous colitis has a striking female predominance (10:1) and the mean age is in the sixth decade.¹ There is chronic, watery diarrhoea with no bleeding. The diarrhoea is secretory in nature, with an active colonic secretion of chloride ions associated with a passive secretion of sodium and water. The diarrhoea may be of sudden onset in over 40% of patients.³ Typically, colonoscopy is normal or shows only mild, nonspecific changes. All the other investigations are negative and diagnosis depends on histology. There is an increased prevalence of collagen deposition in the proximal colon. In an initial series, up to 40% of cases were reported to have normal histology of the rectosigmoid colon.⁴ This has been challenged in a recent study which found that left-sided biopsies alone would have been sufficient for diagnosis in 89% of cases.⁵ The collagen band consists of type III collagen rather than the usual type IV. Since type III is usually involved in repair processes, the suggestion is

Features of collagenous colitis

- watery diarrhoea with no bleeding
- female preponderance (F:M, 10:1)
- mean age: sixth decade (rare in children)
- colonoscopy normal or only minor non-specific changes
- histology diagnostic: subepithelial collagen band in colonic biopsy
- treatment largely symptomatic
- chronic relapsing course (spontaneous remission in about 1/3 rd)

that the collagen band is a result of inflammation. This view is supported by the results of sequential biopsy studies showing inflammation preceding the appearance of collagen.⁴ Most evidence suggests that the band is not important in the pathophysiology of the disease. The presence or thickness of the collagen band correlates poorly with the symptoms, whereas the degree of inflammation correlates with diarrhoea and response to treatment.⁴ Certain disease associations have been noted in patients with collagenous colitis. These include ulcerative colitis, Crohn's disease, coeliac disease, rheumatoid arthritis, systemic lupus erythematosus, scleroderma, pernicious anaemia, myasthenia gravis and Hashimoto's disease.³

The natural history of collagenous colitis appears to be benign, following a chronic relapsing course. Often spontaneous or treatment induced, sustained resolution of symptoms may occur. The collagen band can disappear during therapy or occasionally in association with spontaneous remission. Sulfasalazine and other 5-ASA derivatives have the best response rate.¹ Steroids are also very effective, but their response is not sustained after withdrawal and the doses required are often high.³ Symptomatic treatment with anti-diarrhoeals is the only therapy required in most cases. Surgery in the form of ileostomy for faecal diversion may be required in patients with incapacitating, unresponsive disease.⁶ Our patient responded very well to a short course of steroids. Her diarrhoea settled within a few days of starting prednisolone and uncharacteristically she remained in sustained remission months after stopping the steroids.

Final diagnosis

Collagenous colitis.

Keywords: collagenous colitis; microscopic colitis

1 Jawhari A, Talbot IC. Microscopic, lymphocytic and collagenous colitis. *Histopathology* 1996;**29**:101-10.

2 Lindstrom C. Collagenous colitis with watery diarrhoea: a new entity? *Pathol Eur* 1976;**11**:87-9.

3 Bohr J, Tysk C, Eriksson S, *et al.* Collagenous colitis: a retrospective study of clinical presentation and treatment in 163 patients. *Gut* 1996;**39**:846-51.

4 Carpenter HA, Tremaine WJ, Batts KP, *et al.* Sequential histologic evaluations in collagenous colitis. Correlations

with disease behaviour and sampling strategy. *Dig Dis Sci* 1992;**37**:1903-9.

5 Goff JS, Barnett JL, Pelke T, *et al.* Collagenous colitis: histopathology and clinical course. *Am J Gastroenterol* 1997;**92**: 57-60.

6 Jarnerot G, Tysk C, Bohr J, *et al.* Collagenous colitis and fecal stream diversion. *Gastroenterology* 1995;**109**:449-55.

Pancreatitis and chronic abdominal pain in a patient with AIDS

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A 37-year-old white woman was infected with the human immunodeficiency virus (HIV) through a blood transfusion which she received during a complicated labour in the Ivory Coast. Three years later, due to a decline in the CD4⁺ cell counts to 200/ml, zidovudine (AZT) and trimethoprim-cotrimoxazole were administered. A few months later, zalcitabine (ddC) was added, due to the appearance of fever, abdominal pain and weight loss, and a further decline in the CD4⁺ cell counts down to 15/ml. Despite these measures the patient continued to complain of abdominal pain, which was often exacerbated by food intake. Laboratory studies then showed normal serum amylase and liver enzyme levels. Abdominal ultrasonography (US) and gastroscopy were normal. Due to a suspected role of the antiretroviral drug therapy in inducing the abdominal pain, AZT and ddC were discontinued.

Three months later, the patient was admitted because of severe abdominal pain, vomiting and a fever of 38.5°C. On examination, marked epigastric tenderness was noted. Investigation results were as follows: amylase 1120 IU/l, hypocalcaemia (1.52 mmol/l), alkaline phosphate 219 IU/l, and gamma-glutamyl transpeptidase 478 IU/l. US and computed tomography (CT) revealed gall bladder wall thickening, with a dilated common bile duct and no evidence of stones. The pancreas was mildly enlarged and oedematous.

A diagnosis of pancreatitis was made, a nasogastric tube was inserted and fluids were administered with marked improvement. Blood and urine cultures for cytomegalovirus (CMV) were negative, and fundoscopic examination did not reveal the presence of CMV retinitis. Blood cultures were negative for *Mycobacterium avium-intracellulare* (MAI), and *Cryptococcus neformans* antigens were not detected.

A few weeks later, epigastric pain recurred. Amylase levels remained normal. An endoscopic retrograde cholangiopancreatography (ERCP) was performed (figure). Brush cultures from the mucosa and the biliary fluid grew CMV. Ganciclovir was administered at a dose of 10 mg/kg daily for 3 weeks, followed by 5 mg/kg daily, with improvement in her general condition and a gradual decrease of the pain.

Three months later, parenteral nutrition was started due to intractable abdominal pain which was related to food intake. On this therapy the patient developed severe abdominal pain accompanied by deterioration of liver function tests. A repeat ERCP was performed.

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Figure ERCP with contrast material within the common bile duct, showing mild dilatation without any filling defects. There was no dilatation of the pancreatic duct

Questions

- 1 What diagnosis is suggested by the findings on ERCP?
- 2 Which other organism besides CMV is likely to be found in the biliary tract in this patient?
- 3 What is the optimal management of this patient's condition?