inheritance must be autosomal, either dominant or multifactorial.

Norrgård[4] has studied familial aggregations of aneurysms sending questionnaires to patients following aneurysmectomy. Eighty-nine survivors had 204 brothers and sisters and 3.4% of these had died of ruptured aneurysms in a population with a 0.4% mortality from ruptured aneurysms in the over 50 years age group.

Further evidence that aortic aneurysms may not be solely due to atheroma is derived from comparing patients undergoing aortic surgery for dilating or occlusive disease[5]. The mean age of patients with aneurysmal disease is greater than that for occlusive disease, 67 versus 56 years. Also the sex distribution differs, 12% of aneurysmal patients were female, but 56% of occlusive patients were female.

Serum triglyceride and cholesterol levels have been studied with regard to atherogenesis and in patients with aortic aneurysms[6,7]. Greenhalgh has shown significant differences in the fasting serum lipid levels and lipoprotein patterns between patients with dilating or occlusive disease.

A collagen or elastin deficiency has also been proposed as a possible aetiological factor in the formation of abdominal aortic aneurysms. Most of the strength of the aortic wall is derived from the collagen in the media, the collagen occurring in two forms. Type I collagen is more abundant, but Type III collagen exhibits greater elasticity and tensile strength. When the media is damaged Type III collagen is replaced by a Type I collagen scar[8].

The ratios of collagen types in the skin closely resembles that of the aorta, and thus skin biopsies can be studied to give an indication of aortic collagen. The normal range for Type III collagen from forearm skin biopsies is 17-27% of total skin collagen. Skin biopsies have been taken from both our patients and they have been shown to be Type III collagen deficient, the younger brother having 13.4% Type III collagen and the elder only 10.5% Type III collagen. Absence or a more severe deficiency of Type III collagen is found in Ehlers-Danlos Type IIII syndrome which is characterized by spontaneous vessel rupture with or without previous aneurysm formation. Abdominal aortic aneurysms have not been described in association with Ehlers-Danlos Type IIII Syndrome.

Acknowledgement: We thank Mr K P Robinson for permission to study his patient.

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Epidermolysis bullosa acquisita and total ulcerative colitis

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Keywords: epidermolsis; bullosa; ulcerative colitis; suction blister time

Epidermolysis bullosa acquisita (EBA) is a condition characterized by blistering and skin fragility. Roenizk *et al.* developed the following clinical criteria for diagnosis: (1) The presence of clinical lesions of epidermolysis bullosa dystrophica; (2) Adult onset; (3) Negative family history of epidermolysis bullosa; (4) Exclusion of other bullous disease. More recently histological criteria have been included in the diagnosis[2].

Case report

Case presented to Section of Dermatology, 15 May 1987

A 43-year-old teacher presented to the Dermatology Department at The University Hospital of Wales in March 1985 with blisters precipitated by minor trauma. She gave a history of painless diarrhoea of one year's duration. This had been diagnosed as irritable bowel syndrome and treated with mebeverine, Fybogel and lorazepam. Investigation of the blisters at that time, including histological examination and immunofluorescence were unhelpful. Three months later she presented to the Gastroenterology Outpatient's with weight loss of one and a half stone, abdominal pain and bloody diarrhoea up to 6 times a day. She developed a further crop of blisters at the onset of the illness. At that time she had multiple discrete flaccid blisters on her shins and abdomen. There were areas of depigmentation and mild scarring and milia at the site of old blisters. Blistering was also present in the mouth. Nail dystrophy was absent.

Histology showed a subepidermal blister with a mild chronic inflammatory cell infiltrate (Figure 1).

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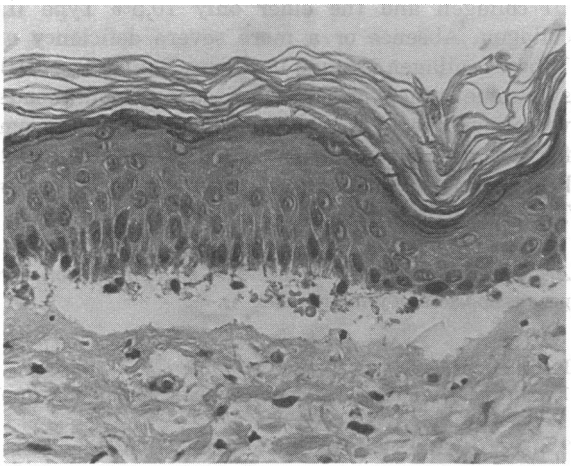

Figure 1. Subepidermal blister with sparce inflammatory cell infiltrate

Porphyrins and indirect immunofluorescence were negative as was direct immunofluorescence to IgG, IgM, IgA, fibrin and complement.

Colonoscopy was performed and showed marked friability of the colonic mucosa with loss of vascular pattern. The changes were mild in the rectum and sigmoid accounting for previous negative sigmoidoscopies. Multiple biopsies throughout the colon showed a severe chronic inflammatory cell infiltrate with goblet cell depletion and surface pus, consistent with total ulcerative colitis.

As there were no fresh blisters at the time of admission, we felt it would be appropriate to obtain information from suction blisters as described by Kustala[3]. The time for blisters to appear on normal skin on the thigh is 90 min. This was performed on the thigh using a pressure of 200 mmHg. The suction blister time in our patient was 4 min. Suction was applied to a new area of adjacent skin for 2 min and biopsy taken to obtain information about early changes. The level of cleavage on paraffin section was subepidermal and on electron microscopy occurred just above the lamina lucida (Figure 2).

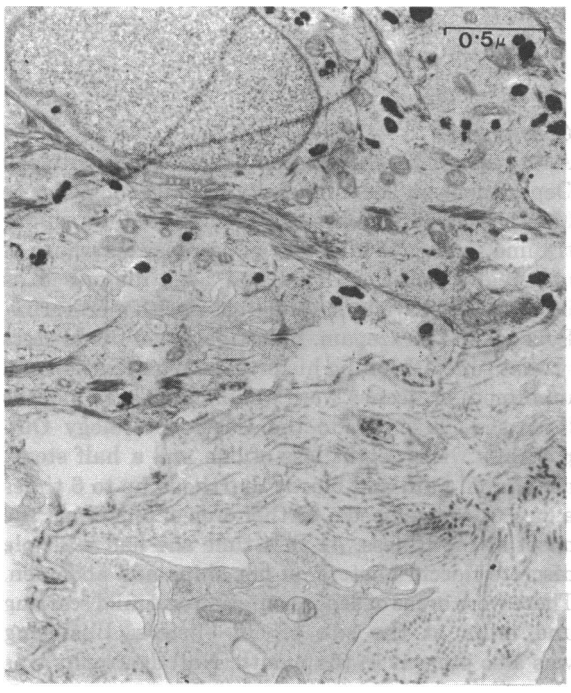

Figure 2. Electron micrograph showing cleavage just above the lamina lucida following suction blister formation

The minimal trauma dose to produce erythema was performed with an instrument developed in the Department of Dermatology, producing a rotary force which gives rise to erythema and later rupture of the epidermis. This was used as an index of ease with which injury could be induced in the skin. The value obtained was 20 g/cm^2 (normal 50 g/cm^2).

She was treated with oral prednisolone 40 mg daily and sulphasalazine 1 g four times a day with rapid resolution of bowel symptoms. There were no further new blisters. One week after treatment had been started, both the suction blister time (6 min) and the minimal trauma dose (50 g/cm^2) were reverting to normal, although the blistering times were still grossly abnormal, in agreement with the clinical picture of persistent skin fragility.

The dose of prednisolone was gradually tailed off and, apart from a brief relapse (of diarrhoea and blistering) in September 1985, which responded to systemic steroids, she has been maintained on sulphasalazine alone. She still notices that her skin is fragile and occasionally blisters, but her weight has returned to normal and she feels well.

Discussion

We feel that this case most closely fits the criteria for EBA, despite immunofluorescence being negative on two occasions. There are now two reports in the literature of EBA associated with negative immunofluorescence[4,5].

We know of no reports of suction blisters being performed in EBA. In suction blisters in all forms of hereditary epidermolysis bullosa, the split occurs in the lamina lucida despite natural blisters occuring at a deeper level in some groups[6]. Although the significance of this is not known, this difference in levels in our case and the abnormal blistering time suggest there may be multiple abnormalities of adhesion.

Intestinal disorders have been noted in association with 30% of EBA[7] but the majority of these are Crohn's disease. We know if only 4 previous cases of EBA in association with ulcerative colitis, reported in the literature[1,8]. In none of these cases was confirmatory evidence provided for the diagnosis of ulcerative colitis by barium enema or colonic biopsies and only in one case[9] was information on treatment and subsequent progress given. No previous association has been described between the activity of the colitis and EBA.

In summary, we report the rare association of ulcerative colitis and EBA. In our patient there was an association between activity of the two conditions, both responding to treatment. We also report an objective method of monitoring response of the skin to treatment.

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Disappearance of an Anglechik prosthesis

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Keywords: Anglechik prosthesis; complication; gastro-oesophageal reflux

Case presented to joint meeting of Sections of Colo-Proctology and Surgery, 26 January 1987

Since 1979 when Anglechik and Cohen[1] reported their experience with the silicone gel prosthesis, over 15 000 devices have been implanted.[2] With more widespread use, reports of serious complications have appeared. One case of complete oesophageal transection, oesophago-gastric fistula and disappearance of the prosthesis is reported from Colchester General Hospital. This seems to be the only case in the literature.

Case report

Mr AW, a 32-year-old man had an Anglechik prothesis inserted on 11 June 1985 for refractory gastro-oesophageal reflux. The oesophagus was easily mobilized. A snug fit was ensured and the knot on the tape was secured with nylon. His postoperative course was uneventful and he was discharged on the 10th postoperative day.

He was re-admitted on 29 July 1985 with severe epigastric pain and dysphagia. A barium meal was reported normal with the prosthesis in place. The barium meal, repeated on 18 September 1985, because of progressive dysphagia, no longer visualized the prothesis. Chest and abdominal X-ray also failed to show it. Laparatomy showed complete transection of the oesophagus with a fistulous communication between the oesophagus and the lesser curve of the stomach.

The abdomen was searched thoroughly but the prosthesis could not be found. An oesophago-gastric anastomosis with a stapler was carried out; postoperatively the patient did well.

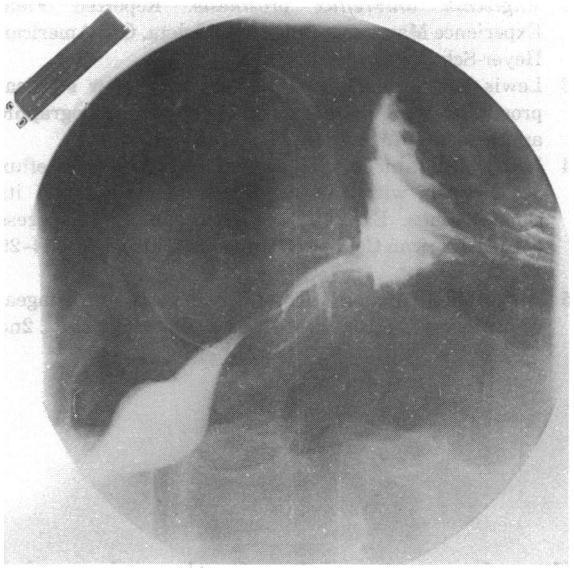

Figure 1. Barium swallow at 6 weeks

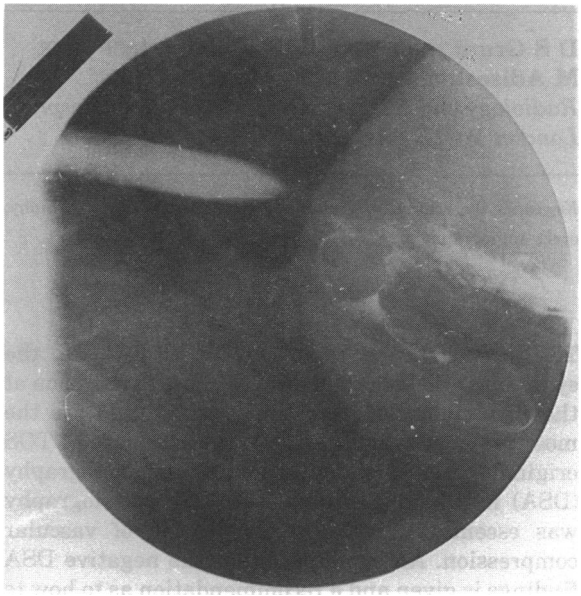

Figure 2. Barium swallow at 6 weeks, lateral film

Discussion

Retrospective review of the barium swallow showed that the prosthesis was not in place and that the tape was cutting into the gastro-oesophageal junction (Figure 1). This probably happened because the tape was too loose around the oesophagus enabling the prosthesis to rotate anteriorly and downwards and act as a deadweight.

This is apparent on the lateral film in the barium swallow (Figure 2). A lateral film is strongly advised in patients with dysphagia following this procedure.

The complete disappearance of the prosthesis can only be explained by its fistulation into the stomach and passage per rectum.

To date there have been no reports of deaths caused by the prosthesis, but there are about 150 serious

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