

than a month. The 5 with stiffness of the skin all volunteered that the skin was more mobile. The 4 patients with painful ulcers, ischaemic and infected, were relieved of pain within forty-eight hours and the ulcers healed in one to three weeks which was far more rapidly than was to be expected, as for example in Case 3 who had complete loss of the terminal phalanges.

We were surprised at the prolonged effect of the treatment. We cannot explain our findings but wonder whether the increased oxygen supply to the arterial wall resulted in less spasm and whether the capillary oxygen beyond the arterial blocks was sufficiently raised to improve materially the oxygenation of the tissues. The method is well established and safe. The only complication which we encountered was a transient myopia lasting three weeks in one patient. Notwithstanding the success of the treatment we advocate that this time-consuming method which needs special skill and equipment should be kept for acute episodes in the disease.

**Dr G B Dowling:** I find this case interesting as a possible example of the 'systemic sclerosis' type of scleroderma in which the skin change has appeared primarily on the trunk. Otherwise you must assume that Raynaud's phenomenon appearing first well in middle age and scleroderma at about the same time are disconnected events.

**Dr R E Church:** What is the temperature inside the hyperbaric oxygen chamber and has Dr Copeman tried the effect of enclosing patients with scleroderma in the chamber without giving hyperbaric oxygen?

In Sheffield we have a room in which the temperature can be controlled and we have treated a number of patients with scleroderma by keeping them at a constant temperature of about 85°F (29°C). The effects are most striking in the relief of pain in digital ulcers as the pain disappears in about 24 hours and there is an improvement in their skin condition which lasts for several months.

**Dr E J Moynahan:** We have used hyperbaric oxygen for these cases and also for stasis ulcers at Guy's Hospital, but have abandoned it, as we found in the case of the latter that exposure of the ulcer to oxygen in a polythene bag was more effective.

I am a little surprised that your patients are allowed to read in the chamber and I would expect a catastrophe like the recent one at Cape Kennedy, especially as the pressure of oxygen inside the chamber is far higher than that in the Apollo spacecraft.

**Dr P W M Copeman:** Regarding the temperature inside the Vickers Hyperbaric Oxygen Bed, this is maintained at the same level as the surrounding air, and hyperthermia is not part of the treatment. As regards putting patients inside without the oxygen, we thought this would be unjustified until there were a reasonable number of cases who had improved with the oxygen. On the point of reading in the chamber, this is perfectly safe, and the accident at Cape Kennedy was not caused by it.

### Systemic Sclerosis of the Small Intestine

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(for Professor C D Calnan FRCP)  
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Mrs J C, aged 56. Housewife

**History:** In 1961 she was investigated for right-sided abdominal pain and backache. A uterine fibroid was found and her symptoms attributed to this. Soon afterwards she started to get Raynaud's phenomena in all four limbs and noticed that her face, arms and hands were swollen. In February 1962 she had an abdominal hysterectomy. Later in the same year she had joint pains and when seen in October slight but definite sclerosis was present in the skin of her face, forearms and hands. Systemic sclerosis was diagnosed. A group of about twelve shiny, flat-topped, mauve papules, each 0.5 cm in diameter, then evolved on her forearms. These showed the histological changes of scleroderma with a marked cellular infiltration by eosinophils and lymphocytes. In May 1963 her ESR reached 70 mm in 1 hour (Westergren) and her plasma globulin 4.1 g/100 ml. In December 1963 abdominal pain after meals returned and after a few weeks she started to vomit small quantities of bile-stained fluid. Examination of her abdomen showed guarding and rebound tenderness maximal in the right iliac fossa. There was a wide-necked incisional hernia resulting from her hysterectomy. Straight X-ray of the abdomen showed a distended large bowel and a diagnosis of incomplete obstruction was made. She was treated expectantly with intravenous fluids and suction. Barium enema two weeks later showed no abnormality.

In February 1964 water brash was troublesome and in April diarrhoea started three or four times a day, usually after meals. By June she was again getting abdominal cramps in her right side. Barium swallow showed normal oesophageal peristalsis and a hiatus hernia. In October 1964 she had dysphagia for solids. Her incisional hernia was repaired in April 1965. In May diarrhoea returned and she was again passing three or four watery stools a day with some mucus. She vomited occasionally. This lasted for two months. In April 1966 she had a further attack of diarrhoea. Her ESR had now reached 128 mm in 1 hour (Westergren). X-rays of the hands showed no bone absorption or soft tissue calcification. Manometry showed oesophageal aperistalsis. In December 1965 the diarrhoea restarted with mucus and a little fresh blood.

In March 1967, at routine outpatients attendance, she complained of severe abdominal pain; signs of peritonitis were found and she was referred for a surgical opinion.

**On examination:** Apyrexial. Pulse rate 72/min.

Blood pressure 130/90. Besides the features of scleroderma described, the only abnormal findings were generalized abdominal tenderness and guarding, maximal in the central lower abdomen with rebound tenderness. The bowel sounds were decreased. Rectal examination was negative. X-ray of the abdomen was not helpful. In view of the history of rectal bleeding and diarrhoea with passage of mucus, a diagnosis was made of peritonitis from a leaking diverticulum or colonic carcinoma in spite of the normal colon on X-rays in 1963. A laparotomy was carried out. A little free fluid with a fibrinous exudate was present in the peritoneal cavity. The stomach, duodenum, gall-bladder, pancreas and large bowel were normal. The small bowel was normal up to 12 cm from the ileocaecal valve. The terminal ileum was oedematous and dilated; the mesentery was thickened and contained several grossly enlarged lymph nodes. In the mesentery and wall of the ileum were some striking white streaks resembling dilated lymphatics. The appendix was normal and was not removed. A lymph node was removed for histology and has been reported as showing reactive hyperplasia.

The appearance was very similar to Crohn's disease or regional enteritis and, as there was no obstruction present, no resection was performed. Post-operative recovery has been uneventful apart from an unusual degree of fibrosis in the abdominal wall. Her bowel action has returned to normal with one formed motion daily.

#### *Discussion*

Reports of involvement of the gastrointestinal tract with collagen disease below the oesophagus are infrequent. Kraus (1924) first described the small bowel changes. Most of the reports have been at autopsy or were X-ray diagnoses.

Irving Marshall (1956) reported 3 cases in which laparotomy was performed for abdominal pain and peritoneal signs. He described an appearance of the small bowel in which 'the bowel was oedematous, showed diminished peristalsis and increased caliber, associated with enlarged, soft lymph nodes and dilated nodal lymphatics filled with a white substance.' Biopsy was performed in each of these cases and changes of sclerosis of the serosa and atrophy and replacement of muscle coat were reported. The jejunum was affected in each of these cases, with no ileal involvement. In 2 of these cases free gas was present under the diaphragm on X-ray, but no macroscopic perforation could be found.

From these descriptions and others, we were struck with the resemblance of the operative findings, particularly with the white streaks in the mesentery. Changes similar to this are seen in intestinal lipodystrophy (Whipple's disease).

Fisher & Whitman (1954) demonstrated that the 'fatty substance' in the mesentery in this condition is in fact 'ground substance' which constitutes collagen. Other than this feature the changes in this patient were typical of Crohn's disease.

*Postscript:* Barium meal two months after operation showed the whole of the small intestine to be widely dilated, in the terminal ileum there was also loss of the mucosal pattern. The changes were those of systemic sclerosis.

#### REFERENCES

- Fisher E R & Whitman J (1954) *Cleveland Clin. Quart.* 21, 213  
 Kraus E J (1924) *Virchows Arch. path. Anat.* 253, 710  
 Marshall I (1956) *New Engl. J. Med.* 255, 978

**Dr M Feivel:** It is an occasional problem in the collagen diseases, perhaps most often in systemic lupus erythematosus and perhaps in a patient already on corticosteroids, to be presented with the clinical picture of an acute abdomen. In the absence of certainty that the symptoms are the result of an exacerbation of the disease it is probably right to consult a surgeon with a view to considering an exploratory laparotomy.

**Dr R B Fountain:** In a series of 727 patients with systemic sclerosis reported by Tuffanelli & Winkelman (1961, *Arch. Derm.* 84, 359), 15 had small bowel involvement. It is interesting that both the skin and gut lesions in this patient showed a particularly inflammatory type of sclerosis.

The following cases were also shown:

- (1) Pityriasis Lichenoides in Identical Twins
- (2) Sarcoidosis
- (3) Reticulosis
- (4) Civatte's Poikiloderma

Dr H R Vickers

**Whitwell's (Behçet's) Disease?**

Dr P W M Copeman (for Dr J L Franklin)

**Acrocyanosis, Skin Infarcts and**

**?Dysproteinæmia**

Dr P W M Copeman (for Dr P D Samman)

(1) **Sézary Syndrome**

(2) **Necrobiosis Lipoidica**

Dr K R Durrant (for Dr H R Vickers)

**Recurrent Stevens-Johnson Syndrome**

Dr O L S Scott

**Necrotizing Arteriolitis**

Dr J D Cohen (for Dr J L Franklin)

**Erythropoietic Protoporphyrria**

Dr R J Cairns

**Lupus Erythematosus Profundus**

Dr J Morgan and Dr G C Wells

**Cutaneous Amyloidosis (Pigmentary Form)**

Dr J A Young (for Dr R H Champion)