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# Ulcerative Colitis and Scleroderma

## A Coincidental Relationship?

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Speculation continues that ulcerative colitis is an autoimmune disorder that is frequently associated with other diseases with a similar underlying pathogenic mechanism. In 1965 we reported a patient with ulcerative colitis and scleroderma in support of this hypothesis. Now we supply a follow-up of over 30 years to describe how each disease acted independently, evidence, we believe, that the association was primarily fortuitous.

**Key Words:** Ulcerative colitis—Scleroderma—Autoimmune mechanisms.

There is very little evidence in the literature to support the hypothesis of a relationship between ulcerative colitis and autoimmune diseases except for the demonstration that these two entities often coexist (1–4). In 1965, we described a patient with ulcerative colitis and scleroderma, to support this relationship (1). Since that time we have followed this patient whose disease has persisted for over 30 years and found that the two diseases did not follow in parallel but rather that the ulcerative colitis healed and the scleroderma became progressively worse. The fact that these two entities acted independently is strong evidence that the association was primarily fortuitous. Except for a case report by Bix in 1958 (2), we are unaware of any recent publications describing these two entities in one patient.

## CASE REPORT

### The Clinical Course of the Scleroderma

In 1951, at age 21 years, the patient first developed Raynaud's phenomenon for which, in 1953 a dorsal sympathectomy was performed. The patient noted a progressive tightening of his fingertips with ulcerations, and in 1956 a skin biopsy was consistent with scleroderma. He developed dyspeptic symptoms; in 1964 a barium study of the upper GI tract was normal, but by 1970, the esophagus had become dilated and there was slow emptying of the barium, revealing the first evidence of scleroderma involvement of the GI tract. Esophagoscopy at this time was normal, but biopsies revealed the presence of acute and chronic esophagitis. The patient was treated with antacids and an antireflux regimen. In June, 1977, esophagoscopy revealed glandular epithelium consistent with Barrett's epithelium, extending from the gastroesophageal junction at 33 cm extending up to 29 cm. Subsequently, surveillance panendoscopy and cytology exams approximately every 1–2 years until 1991, showed progressive extension of Barrett's epithelium up to 25 cm. Biopsies showed mild dysplasia. An esophageal motility study on June 1, 1979, revealed no reflux, a negative Bernstein test, and a motility disorder consistent with scleroderma. The patient has been maintained on H2 blockers and an antireflux regimen, is relatively free of GI symptoms, and has no incapacity from the scleroderma. He has no evidence of involvement of his lungs, heart, or kidneys.

### Clinical Course of Patient's Ulcerative Colitis

The patient first noted transient episodes of bloody diarrhea in 1956 and again in 1958. In 1959, an exacerbation of bloody diarrhea led to sigmoidoscopy, which revealed diffuse friability, edema, and minute ulcerations from the anus and extending up to 18 cm. A diagnosis of ulcerative colitis was confirmed by histology, which showed mucosal infiltration with plasma cells, eosinophils, and crypt abscesses. A barium enema was normal. The patient continued to have exacerbations of bloody diarrhea in 1962, 1963, 1971, 1977, and 1983. Follow-up examinations during asymptomatic intervals in 1970, 1987, and 1990, the colon appeared endoscopically normal. Histologic examination of the mucosa revealed only increased lymphoplasmacytic cells in the distal 10 cm of the colon, as well as evidence of focal atrophic changes. The patient was treated with steroids during exacerbations of bloody diarrhea, but received no medication during periods of remission.

### X-Ray Investigations of the GI Tract

Barium enema examinations were performed with the following results: 1958, normal; 1964 air contrast and

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barium study, normal; 1977 sigmoid diverticulosis; 1990 muscular ridging of the entire colon worse in the left colon, with demonstration of pseudodiverticulum along the antimesenteric border. A series of barium studies of the upper GI tract revealed the following: 1962—normal except for duodenal deformity; 1964—upper GI including small bowel study, normal; 1970—slight dilatation of the esophagus with diminution of peristalsis consistent with scleroderma; 1977—more extensive esophageal dilatation; 1990—marked dilatation of the esophagus with atrophy and retention of barium; a small bowel series revealed minimal thickening of the mucosal folds of the ileum with slow bowel transit.

*Lab Data*

In our original report the following laboratory studies were normal: cryoglobulins, protein electrophoresis, colon antibodies by fluorescent antiglobulin technique, antinuclear factors, thyroglobulin antibody, and nondetectable rheumatoid factors. Subsequently routine chemical profiles have remained normal. A hydrogen breath test was consistent with lactose intolerance.

*Family History*

The patient's brother died in his 20's with alpha-1-antitrypsin deficiency, and a second brother died in his 30's of cancer of the neck of unknown etiology. The patient's son died at age 36, with renal failure secondary to diabetes mellitus. The patient's sister had deforming arthritis.

## DISCUSSION

Our patient, closely followed for over 30 years, has shown progressive involvement of his upper GI tract with changes associated with scleroderma. The esophagus is the most involved with dilatation, loss of motility, and ultimately the development of Barrett's epithelium. Barium studies of the small bowel has shown early mucosal ridging and slow transit time, and the colon revealed wide mouth diverticula along the antimesentery border, typical of changes in scleroderma (5). The course of the patient's colon was typical of distal ulcerative colitis (6–7). The colon involvement started with bloody diarrhea in 1956 and subsequently the patient had eight exacerbations that were followed by remissions. The last episode was a mild exacerbation in 1983. The fact that the involvement of the GI tract with scleroderma progressed and that the distal ulcerative colitis had almost completely disappeared, supports the idea that the occurrence of these two diseases of unknown etiology in one patient is probably a coincidence. Although an autoimmune basis is not proven for ulcerative colitis, there appears to be a truer association of autoimmune disorders with ulcerative colitis than with Crohn's disease (8). A more current hypothesis is that ulcerative colitis, in addition to its familial predisposition, may be a defect in mucosal immune regulation with abnormal immune responses to environmental agents (9,10).

## CONCLUSION

A 33-year clinical course of a patient with distal ulcerative colitis associated with scleroderma is reported. The fact that scleroderma progressed and the distal ulcerative colitis totally healed, and the fact that this is the only second known case report that we can find of ulcerative colitis associated with scleroderma, is evidence, we believe, of a coincidental relationship and against the possibility of a common, underlying, predisposing mechanism in these two diseases.

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