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Gastrointestinal Radiology

Barrett's Esophagus Complicating Scleroderma

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Abstract. Two patients with scleroderma whose esophageal involvement was associated with long-standing reflux esophagitis were found to also have Barrett's esophagus. Since Barrett's esophagus is a premalignant condition, these patients with scleroderma should be considered at high risk for the development of adenocarcinoma of the esophagus.

Key words: Esophagus, progressive systemic sclerosis – Scleroderma – Barrett's esophagus – Esophagus, columnar lined.

Scleroderma is a generalized multisystem disorder of unknown cause characterized by vascular, fibrotic, and inflammatory changes that involve both skin and internal organs. The latter include digital arteries, gastrointestinal tract, lungs, heart, musculoskeletal system, and kidneys. Within the gastrointestinal tract, the esophagus is involved most commonly and 50–80% of patients may have esophageal involvement at the time of diagnosis [1–5]. There is disruption of normal esophageal peristaltic activity, and the distal esophageal high-pressure zone loses its tone and normal response to swallowing. This hypotonia of the lower esophageal sphincter (LES) leads to gastroesophageal reflux, which, coupled with the inability of the dilated lower esophagus to clear refluxed material back into the stomach, creates an ideal setting for development of reflux esophagitis. This, in turn, not infrequently results in stricture formation. Chronic reflux esophagitis may also lead to Barrett's esophagus, or the replacement of the acid-damaged squamous epithelium with columnar epithelium. Although Barrett's esophagus should be a common complication of scleroderma, it has only been recognized recently [1, 2]. Of the 5 cases so far reported 3 had adenocarcinoma of the esophagus complicating Barrett's mucosa.

We describe 2 patients with scleroderma whose long-standing history of reflux esophagitis was associated with the development of Barrett's esophagus. The difficulties of diagnosing Barrett's esophagus in patients with scleroderma are discussed.

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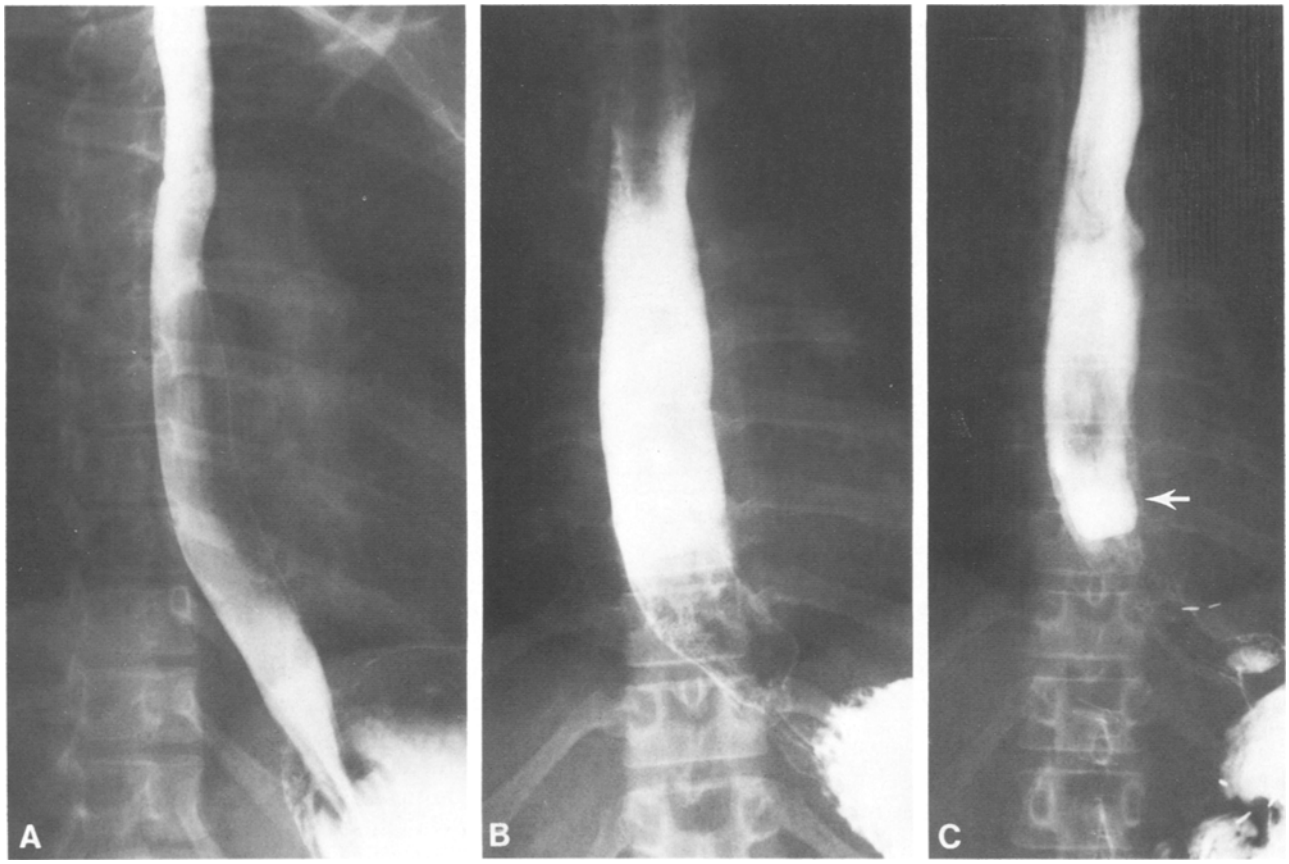
Case Reports

Case 1

The diagnosis of scleroderma was made in a 38-year-old black woman in 1972 based on a 2-year history of Raynaud's phenomenon and lung and esophageal involvement. Her symptoms of recurrent peptic esophagitis necessitated a Collis–Belsey hiatus hernia repair in 1975 and revision to Collis–Nissen fundoplication in 1978. However, she continued to have gastroesophageal reflux with substernal pain, heartburn, and dysphagia. A barium swallow revealed a dilated atonic esophagus with severely disordered motility and free spontaneous gastroesophageal reflux (Fig. 1). Endoscopy and esophageal function tests confirmed the presence of spontaneous gastroesophageal reflux and severe ulcerative reflux esophagitis. A biopsy specimen of the esophagus revealed the changes of advanced reflux esophagitis. Despite medical antireflux treatment regimen and 2 antireflux operations she was disabled by her esophageal difficulties. No specific radiographic, endoscopic, or histopathologic features suggested Barrett's esophagus.

In October 1979 she underwent transhiatal esophagectomy and cervical esophagogastric anastomosis. The histopathologic examination of the resected esophageal specimen revealed changes of reflux esophagitis and Barrett's mucosa (Fig. 1). Her only postoperative complication was paralysis of the left true vocal cord related to traction injury to the recurrent laryngeal nerve during surgery, which required Teflon® injection. Occasionally she has required dilatation of her cervical esophagogastric anastomosis because of mild cervical dysphagia in the 4 years since surgery. Otherwise she is eating a regular diet and her systemic disease is stable.



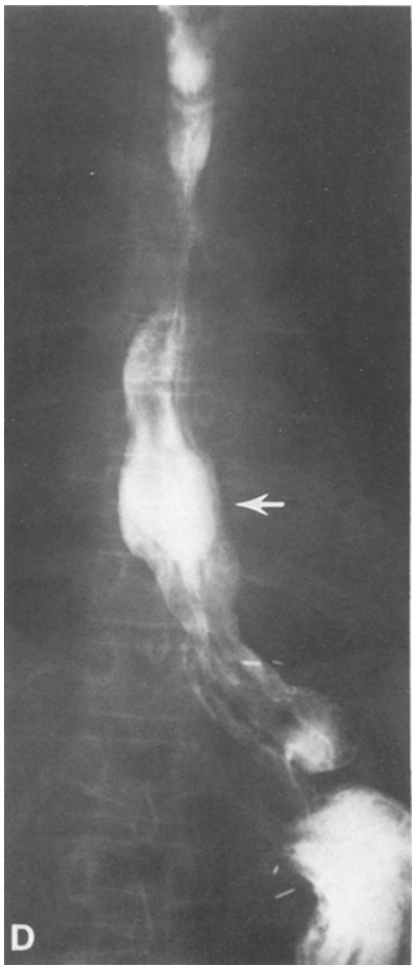
Case 2

The diagnosis in 1968 of scleroderma in this 44-year-old white women was made based on her history of Raynaud's phenomenon and subcutaneous calcinosis which had to be removed frequently from her fingers and knees. In July 1979 she was referred to University Hospital for evaluation of dysphagia and to rule out the possibility of a malignant tumor of the esophagus. Her history at that time included dysphagia with food sticking in the midesophagus, as well as 15–20 years of substernal burning pain with definite postural aggravation. Intermittently during sleep she also noted regurgitation of acid through her nose and mouth. These symptoms had gradually increased over the previous 5 years. She had been treated with antireflux measures but failed to obtain significant relief.

A barium swallow revealed a dilated atonic esophagus, a small hiatal hernia, a stricture in the distal esophagus, and spontaneous gastroesophageal reflux (Fig. 2). Esophageal function tests confirmed an advanced stage of involvement by scleroderma and severe gastroesophageal reflux. Endoscopy revealed severe ulcerative esophagitis. A biopsy specimen of the distal esophagus failed to show malignancy, but there were changes of reflux esophagitis and the appearance of columnar epithelium was consistent with Barrett's mucosa (Fig. 2). In November 1979 she underwent a Collis–Nissen fundoplication operation. Her postoperative course was unremarkable. She has complained on occasion of minor heartburn in the 4 years since surgery, and is being evaluated yearly with endoscopy and barium esophagography.

Discussion

For the past 2 decades attention has been focused on both the physiological and radiographic aspects of the esophageal changes in patients with scleroderma [3–11]. The motor disorder ranges from decreased peristaltic waves to complete aperistalsis. Although lower esophageal sphincter hypotonia is generally present, an occasional patient may manifest normal LES pressure. The abnormality was assumed to be the result of fibrous replacement of muscle. However, a combined manometric–pathologic study showed defects in peristalsis in the absence of fibrosis or muscular atrophy, which suggested a functional abnormality [12]. Pharmacologic testing was consistent with either a neural or a primary myogenic defect as the cause of LES hypotonia [13]. The main radiologic features of esophageal involvement with scleroderma have been well-documented [4–6, 14, 15] and consist of a dilated esophagus with decreased peristaltic activity, most commonly in the lower two-thirds, and a wide gastroesophageal junction with incompetent cardia resulting in free and spontaneous gastro-



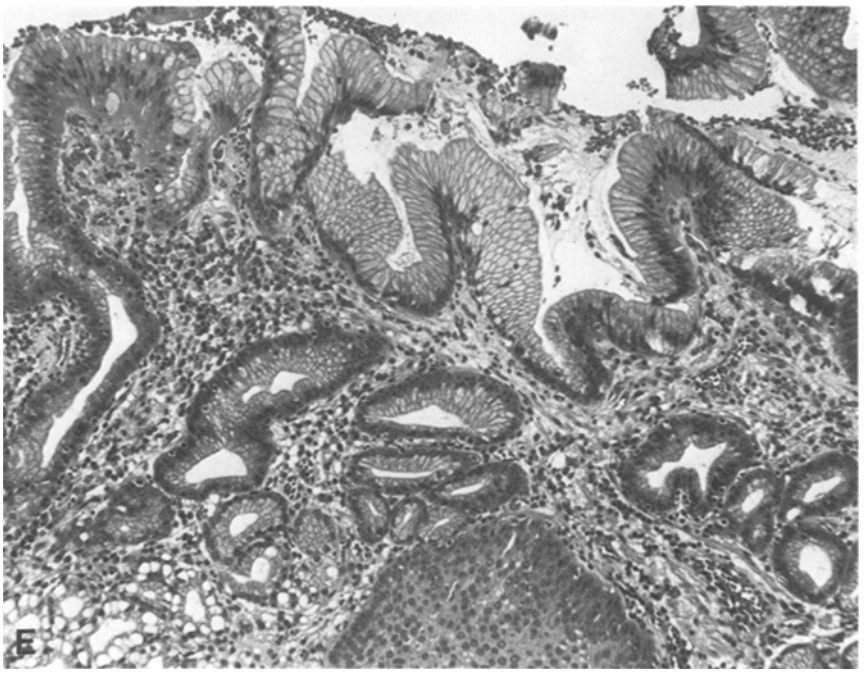


Fig. 1. Double-contrast views from esophagograms performed from 1973 through 1979. **A** (*facing page, left*) Mild esophageal dilatation affecting the lower two-thirds and wide open gastroesophageal junction (1973). **B** (*facing page, center*) Slight progression of esophageal dilatation (1975). **C** (*facing page, right*) and **D** (*left*) Further progression of dysmotility is manifested by flow artifact created by poor mixing of barium (transient intraluminal diverticula) (*arrows*) and a hiatal hernia. Metallic clips are from previous antireflux operations. Barrett's esophagus is not suggested by any of these studies. **E** (*above*) Photomicrograph from distal esophagus shows columnar lined epithelium with intestinal metaplasia and goblet cells consistent with Barrett's type mucosa (hematoxylin and eosin, ×108)

esophageal reflux. This leads to reflux esophagitis and may result in stricture formation [16–19]. Less frequently, chronic reflux esophagitis may lead to the development of Barrett's mucosa. Although gastroesophageal reflux is common in patients with scleroderma, Barrett's esophagus was not reported until 1978. Cameron and Payne [1], in a review of Mayo Clinic patients, reported 2 cases of scleroderma complicated by development of Barrett's esophagus. Recently, Halpert et al. [2] reported adenocarcinoma of the esophagus in 3 patients with scleroderma who also demonstrated adjacent areas of Barrett's mucosa. Malignancy in patients with scleroderma is much less common than in those with dermatomyositis. However, several reports described malignant tumors of lung, breast, and, less commonly, esophagus [20–25].

The actual incidence of Barrett's esophagus in patients with scleroderma is difficult to determine. With recent awareness of Barrett's esophagus as a premalignant condition, this diagnosis is probably made more often. We suspect that many cases of Barrett's esophagus in patients with scleroderma were missed because of lack of awareness of this association. Increased awareness of Barrett's esophagus in patients with scleroderma will lead to greater detection of this entity, provided biopsy specimens of the esophagus are obtained for histologic examination. A search of records at the University of Michigan revealed only 2 cases among 502 registered and treated cases of scleroderma between 1975 and 1984. This does not suggest increased incidence. Perhaps more cases of Barrett's esophagus had been missed in this group of patients and the condition is probably more common than reported.

It is important to recognize this association between scleroderma and Barrett's esophagus because of the high frequency of gastroesophageal reflux and reflux esophagitis in these patients. However, several difficulties arise in diagnosing Barrett's mucosa in patients with scleroderma on

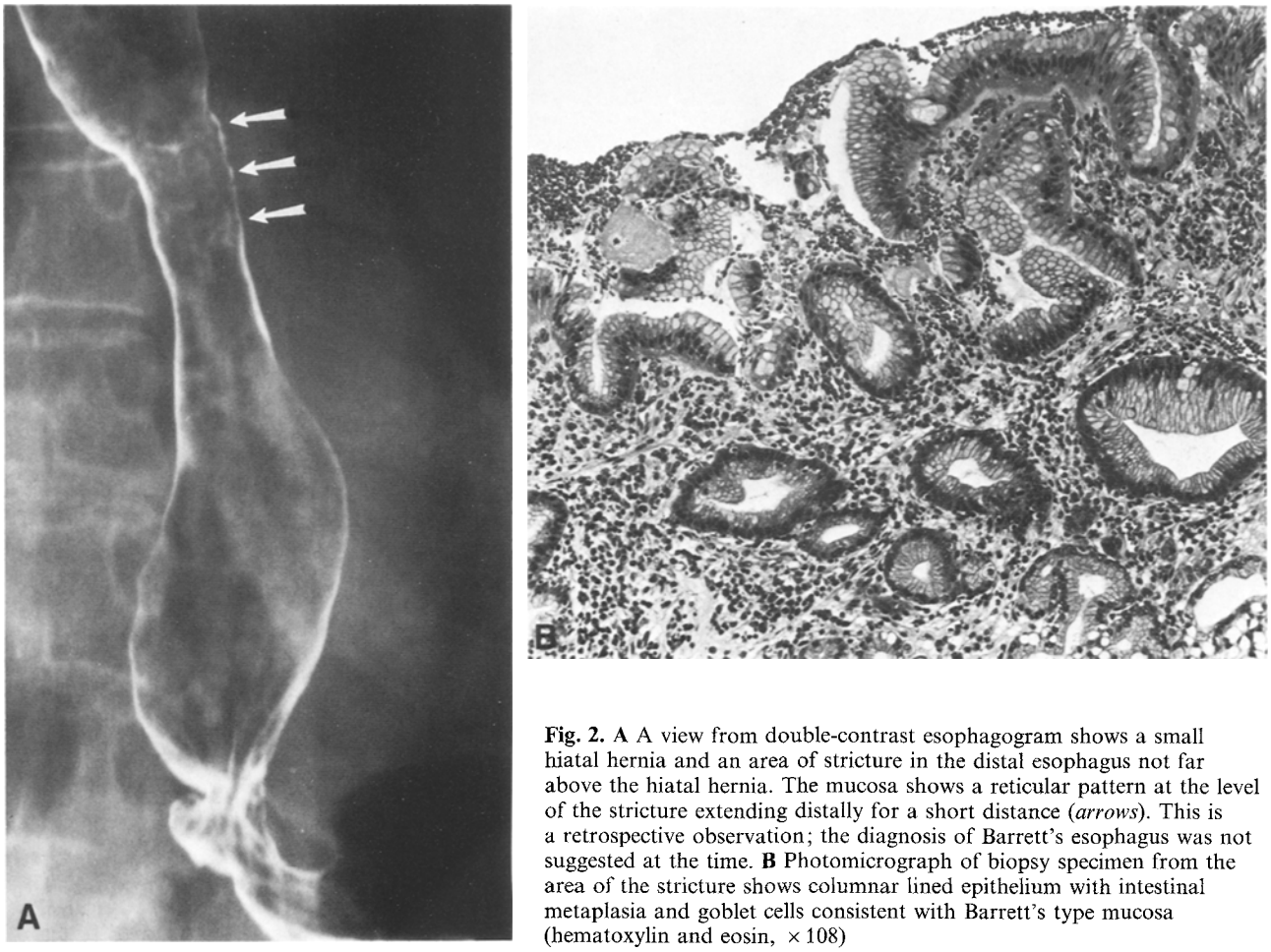


Fig. 2. A A view from double-contrast esophagogram shows a small hiatal hernia and an area of stricture in the distal esophagus not far above the hiatal hernia. The mucosa shows a reticular pattern at the level of the stricture extending distally for a short distance (*arrows*). This is a retrospective observation; the diagnosis of Barrett's esophagus was not suggested at the time. **B** Photomicrograph of biopsy specimen from the area of the stricture shows columnar lined epithelium with intestinal metaplasia and goblet cells consistent with Barrett's type mucosa (hematoxylin and eosin, ×108)

esophagogram. Hiatus hernia, gastroesophageal reflux, dilated esophagus with dysmotility, chronic reflux esophagitis, and strictures are nonspecific features [26]. On double-contrast esophagography, a reticular pattern of the mucosa [27] may be helpful in suggesting the diagnosis of Barrett's mucosa, but it is found in only 12.5–24% of patients [28]. The endoscopist encounters the same difficulties in diagnosing Barrett's mucosa amid changes of chronic reflux esophagitis and severe degrees of dysmotility. We recommend endoscopic biopsy of esophagus in patients with scleroderma who have persistent gastroesophageal reflux and chronic symptomatic reflux esophagitis. As in our case 1, several biopsy specimens did not show Barrett's mucosa and the diagnosis was made at the histologic study of surgical specimen after esophagectomy.

Since Barrett's esophagus is considered a premalignant condition predisposing to the development of adenocarcinoma [29–35], its recognition in patients with scleroderma is important for identification of this high-risk group so that close follow-up can be instituted.

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