Case report

Rapidly progressive scleroderma associated with carcinoma of the oesophagus

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SUMMARY A patient with scleroderma of recent onset was found to have a carcinoma of the oesophagus. The rapid progression of the scleroderma suggested the possibility that it represented a systemic manifestation of malignancy.

There are numerous reports of an association between dermatomyositis and malignancy, and Barnes (1976) has reviewed 258 cases from the literature. There are few reports of scleroderma associated with malignancy, of which only 3 were oesophageal tumours.

Case report

A 70-year-old woman was admitted in July 1977 for investigation of rapidly progressive scleroderma. She had noticed mild Raynaud's disease, with no evidence of skin changes, in her hands and feet for many years, but quite suddenly in May 1977 she developed very severe persistent Raynaud's disease with areas of incipient gangrene at the tips of several fingers. This was followed by pain and swelling of the fingers with skin thickening and flexion contractures. The necrotic lesions healed in 3 weeks, but permanent vascular changes persisted in the hands. She had smoked 20 cigarettes a day for many years, and for 10 years had had symptoms of chronic bronchitis, which had been worse in the last 3 years.

On examination there were characteristic changes of scleroderma in the hands, with tight, red, shiny skin and flexion contractures of the fingers. There were no skin changes elsewhere and no other musculoskeletal abnormalities.

Investigations: haemoglobin 12.4 g/dl; white cell count $6.0 \times 10^9/l$; ESR 32 mm in the first hour; blood urea and liver function tests normal; creatinine clearance 55 ml/min; electrophoresis, normal

pattern; latex test and antinuclear factor negative. Hand x-rays: generalised soft-tissue thickening in the phalanges, with some sclerosis and resorption of bone in the tips of the distal phalanges. Chest x-ray: overinflated lungs and mild diffuse interstitial changes in both mid and lower zones. Barium swallow showed an irregular stricture in the mid-oesophagus extending over a 5 cm segment. The peristalsis in the oesophagus, apart from this segment, was normal. Oesophagoscopy showed a tumour at 30 cm from the oral margin, and histology of biopsy specimens revealed a primary squamous cell carcinoma of the oesophagus, which was anaplastic in places.

The barium swallow had been performed to assess the extent of the scleroderma and not because of dysphagia. On questioning, the patient admitted to occasional mild dysphagia. For various reasons it was decided that no specific treatment of the tumour was indicated, but in view of the severe disability caused by the scleroderma treatment with D-penicillamine was begun. One month later the scleroderma had progressed still further, with gross finger contractures. The patient's general condition deteriorated rapidly, and she died in October 1977, 3 months after the tumour had been discovered. No necropsy was performed.

Discussion

We know of three other reports of oesophageal malignancy associated with scleroderma, but only in 1 case (Kilton and Gottlieb, 1971) was the scleroderma of short duration. Matzner *et al.* (1963) described a patient who had scleroderma for 11 years before severe dysphagia occurred; histology

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showed scleroderma of the oesophagus with co-existent epidermoid carcinoma. Johnson and Monroe (1973) reported a patient with progressive systemic sclerosis with oesophageal involvement who developed carcinoma of the oesophagus 26 years after the first symptom of dysphagia. It was postulated that the carcinoma resulted from long-standing stasis, irritation, and scarring, as may occur in other conditions, for example, stricture and achalasia, producing oesophageal stasis.

Although there are isolated reports of other malignancies in association with scleroderma, the best-documented association is that of alveolar or bronchiolar cell carcinoma of the lung superimposed on sclerodermal lung. This type of lung tumour may be superimposed on any type of inflammatory interstitial pulmonary disease (Twersky *et al.*, 1976).

There have been at least two reports of sclerodermatous changes in the skin of the legs in association with functioning carcinoid tumours (Zarafonitis *et al.*, 1958; Asboe-Hansen, 1959). It was postulated that the excessive amount of serotonin (5-hydroxytryptamine) secreted by the carcinoid tumours might be related to the development of sclerodermatous changes in the skin (and to fibrosis in other tissues). Basten and Bonnin (1966) reported a patient in whom the development of severe scleroderma was thought to be related to the presence of an adenocarcinoma of the stomach. They suggested

that the development of scleroderma in malignant disease of any origin might possibly be related to the elaboration by these neoplasms of serotonin-like substances.

The short history and rapid progression of the scleroderma changes in the hands of our patient suggest that scleroderma, like dermatomyositis, may occasionally represent a systemic manifestation of malignancy.

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