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Squamous carcinoma in situ of the oesophagus in a patient with achalasia

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There is a strong aetiological association between achalasia and squamous carcinoma of the oesophagus, particularly of the middle third. Until now all reported cases have been of advanced tumours, 80% of which have been found unsuitable for resection. We report the first case of squamous carcinoma in situ in a patient with achalasia. It is reasonable to suppose that all tumours go through this "early" stage and that prompt detection may lead to higher resection rates and improved survival.

Case report

In 1981 a 42 year old caucasian man presented to another hospital with a 12 year history of excessive belching, vomiting at night, and epigastric and retrosternal pain. Barium swallow showed gross dilatation of the oesophagus and a diagnosis of achalasia was made. An abdominal Heller's myotomy was performed in May 1981.

In March 1983 he returned with further epigastric pain and belching. A repeat barium swallow showed that the calibre of the oesophagus had diminished considerably since the previous examination, but there was no evidence of carcinoma. Fibreoptic oesophagoscopy showed several white plaques in the mid oesophagus and biopsy showed squamous carcinoma. A full blood count, biochemical investigations, and ultrasound examination of the liver all gave results within normal limits and the patient was referred to the regional thoracic surgical unit at East Birmingham Hospital for further management.

In August 1983 a subtotal oesophagectomy was performed via a left thoracolaparotomy and left neck incision and reconstruction was accomplished by oesophagogastric anastomosis in the neck. There was no evidence of tumour spread at operation. The patient made a satisfactory postoperative recovery and 12 months later remains well with no evidence of recurrent tumour.

The resected oesophagus was 8 cm in circumference. The mucosal lesion in the middle third was not of the usual ulcerated nature but was plaque like with a roughened surface (fig 1) and measured 5 × 2 cm. Histological sections from multiple sites of this lesion all showed squamous carcinoma in situ (fig 2). Sharply delineated tongues of neoplastic cells extended for a short distance into the underlying connective tissue, but there was no evidence of invasion. The overall thickness of the affected epithelium varied from 150 to 750 μm.

Discussion

Carcinoma of the oesophagus in association with achalasia was first reported by Fagge in 1872[1] and its incidence has been variably reported as 0.3–20% of patients known to

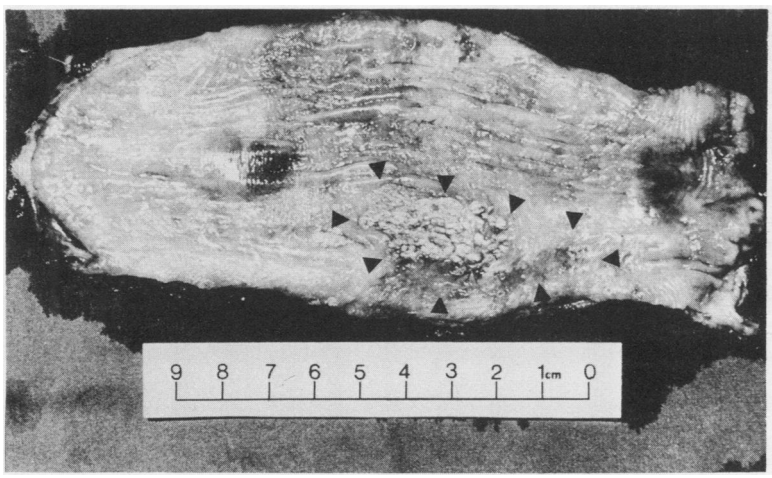

Fig 1 *Subtotal oesophagectomy specimen. The in situ carcinoma is represented by a roughened area of mucosa in the middle third (arrowheads).*

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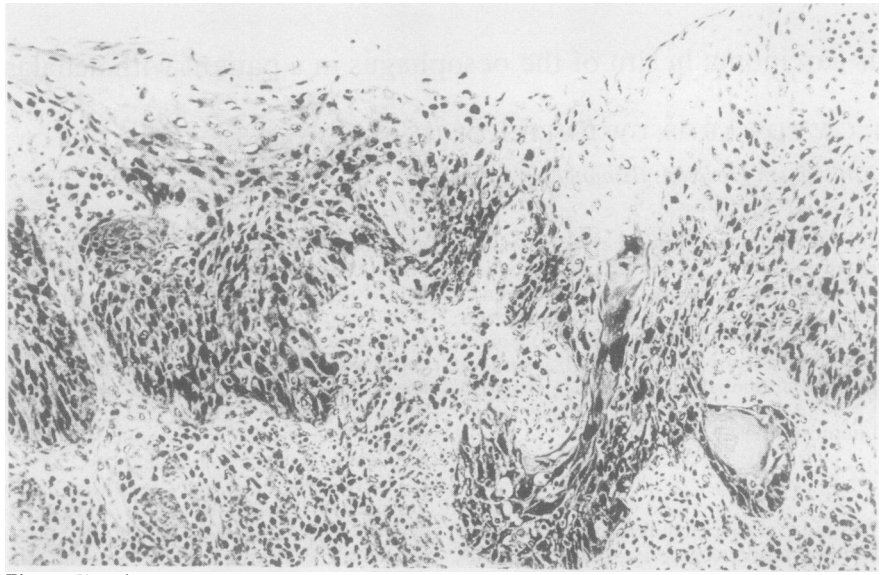

Fig 2 *Histological section of the squamous carcinoma in situ. (Haematoxylin and eosin, × 125.)*

have achalasia,[2-4] Survival after treatment for these tumours is very poor and only one patient has been reported to have survived five years. This poor survival reflects the extent of the tumours at the time of presentation. They develop in a dilated lumen and therefore reach a considerable size before becoming symptomatic; even then, as patients with achalasia are tolerant towards their symptoms, a slight change in them will not immediately result in referral or investigation. This is the first case to be reported of squamous carcinoma in situ of the oesophagus in a patient with achalasia. Although the patient had gastrointestinal symptoms, these are very unlikely to have been caused by the neoplastic area in the oesophagus, which was found incidentally during endoscopy.

Correa[5] has reported a range of preneoplastic changes in the oesophageal mucosa, including chronic inflammation, leucoplakia, atrophy, papillomatosis, dysplasia, and carcinoma in situ. We may reasonably suppose that many tumours are preceded by such lesions, and that their detection might result in the finding of an increased number of patients suitable for resection, with an improvement in subsequent survival. Although screening for malignancy in achalasia is currently recommended it is not performed routinely, and as no patients are reported to have been cured after presymptomatic detection of tumours the recommendation for screening is debatable. This case emphasises that tumours can be detected at a preinvasive stage and that screening for malignancy may be of benefit to patients with achalasia.

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