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Multicentric tracheobronchial and oesophageal granular cell myoblastoma

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O'Connell, D J, MacMahon, H, and De Meester, T R (1978). *Thorax*, **33**, 596–602. **Multicentric tracheobronchial and oesophageal granular cell myoblastoma**. Two patients with multiple intrathoracic granular cell myoblastomas are described. In one case multiple tumours were present in the major airways and oesophagus. The patient presented with recurrent pulmonary infections and stridor due to airway obstruction. In the other case dysphagia caused by multiple oesophageal granular cell myoblastomas was the major symptom. Granular cell myoblastoma is a rare tumour of neurogenic origin with a characteristic histological appearance. The pattern of multiple tracheobronchial and oesophageal tumours is uncommon and forms the basis of this report.

Granular cell myoblastoma (GCM) is a rare tumour of neurogenic origin, most often found in the skin, tongue, or larynx (Vance and Hudson, 1969; Oparah and Subramanian, 1976). Other less common locations include the bile ducts, breast, thyroid, and vagina (Serpe *et al*, 1960; Umansky and Bullock, 1968; Ostermiller *et al*, 1970). Multiple tumours are reported to occur in 7% of these cases (Moscovic and Azar, 1967). There have been few reports of tracheobronchial or oesophageal locations for these tumours, and multiple intrathoracic lesions are exceptionally uncommon. We have recently encountered two patients who presented with symptoms referable to multiple granular cell myoblastomas in the larynx, trachea, bronchus, and oesophagus.

Case reports

CASE 1

A 36-year-old black woman presented in 1971 with a productive cough of three months' duration. A chest radiograph showed patchy consolidation in the left upper lobe, which cleared after a ten-day course of penicillin. Two subsequent left upper lobe infections led to a bronchogram, which showed a smooth, 2 cm submucosal mass at the junction of the left main and lower lobe bronchi (fig 1). Lingular bronchiectasis was also shown. Bronchoscopy confirmed these findings, and biopsy of the endobronchial mass showed histological features characteristic of GCM. The tumour was curetted through the bronchoscope.

The patient was seen again in 1974 after a haematemesis. This was considered to be due to alcoholic gastritis. Results of a barium examination of the oesophagus and stomach, performed at that time, were normal.

In May 1977 she complained of mild dyspnoea, and a chest radiograph showed consolidation in the lingula, which cleared after antibiotic treatment. Two months later, however, she presented again with severe stridor, dyspnoea, and dysphagia. The chest radiograph was normal, but tomograms of the trachea and major airways showed a large, lobulated mass in the upper trachea, narrowing the lumen considerably (fig 2). Bronchoscopy confirmed the tomographic findings, and an additional tumour mass was seen in the larynx. Biopsy of the tracheal mass showed granular cell myoblastoma. An upper gastrointestinal examination showed three separate submucosal masses in the mid and lower oesophagus (figs 3(a) and (b)). At thoracotomy the affected segment of trachea and the solitary laryngeal tumour were resected. Later, the patient developed severe tracheal stenosis at the operation site, but further treatment was refused. She died after a massive aspiration of stomach contents. At

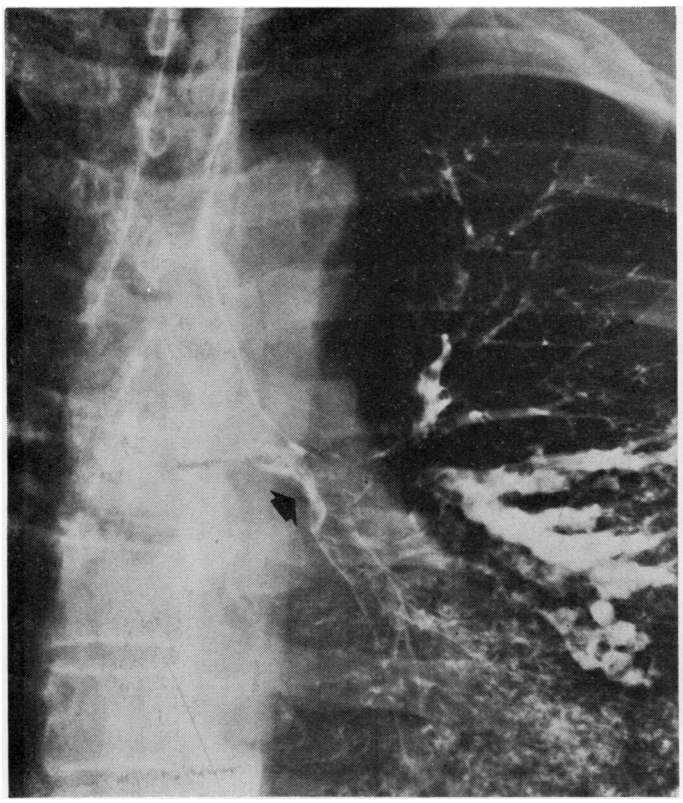


Fig 1 *Case 1. Bronchogram shows submucosal mass (arrow) and lingular bronchiectasis.*

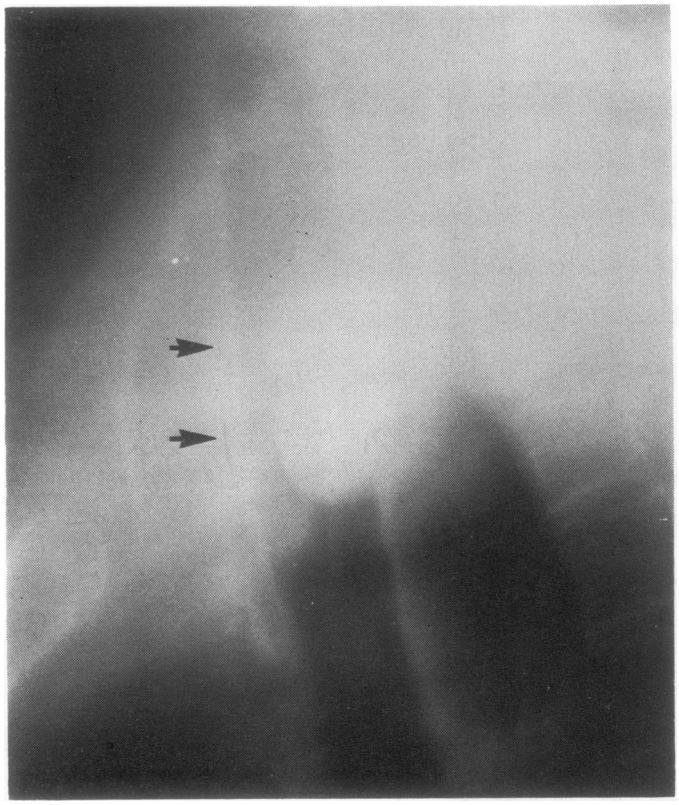
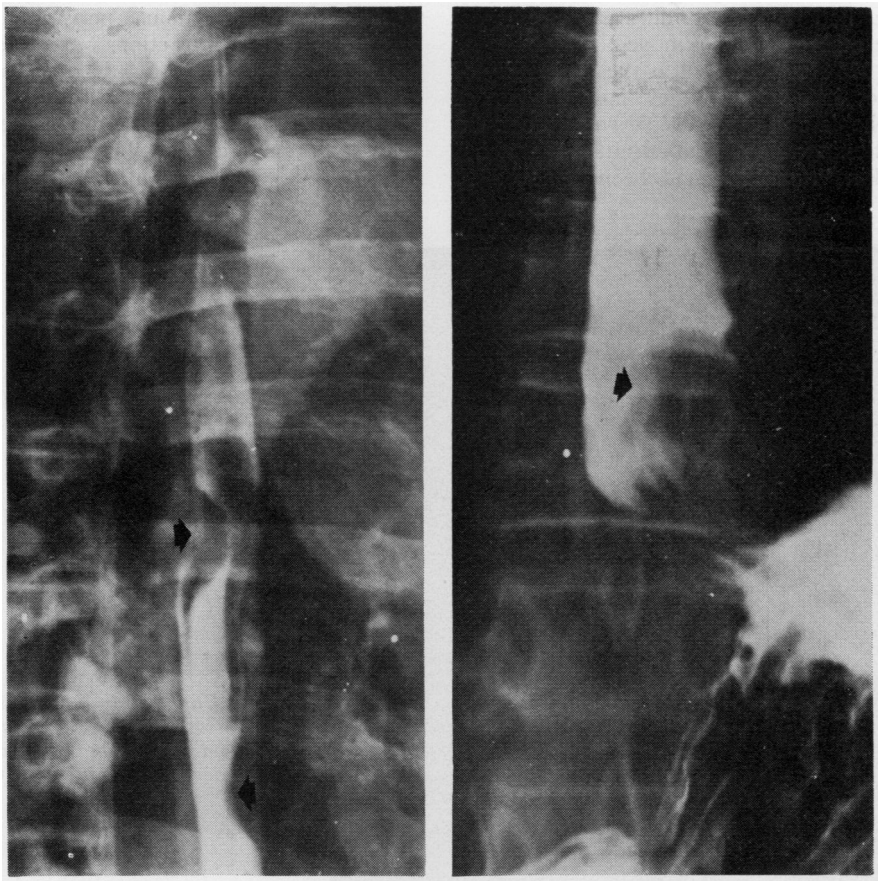


Fig 2 *Case 1. Lateral tomogram shows a lobulated mass attached to posterior wall of trachea (arrows).*



(*a*) (*b*)

Fig 3(a) and (b) ***Case 1. Barium examination shows two submucosal masses (arrows) in mid-oesophagus and another mass distally.***

necropsy, in addition to the tumours found in vivo, there were three small tumours in the stomach and another in the pericardium.

CASE 2

This 56-year-old white woman presented with a two-year history of worsening dysphagia and intermittent substernal chest pain. Investigation at another institution in January 1976 showed a submucosal mid-oesophageal mass. Oesophageal manometry had indicated diffuse oesophageal spasm. The mass was not removed.

In July 1976 she presented at the University of Chicago hospitals and clinics complaining of severe dysphagia and considerable weight loss. On direct questioning, the patient admitted that about 20 small skin "tumours" had been removed 10 years previously. These tumours had been called "myoblastomas." Physical examination was unremarkable apart from obvious recent weight loss. The chest radiograph was normal. Barium examination showed a large, smooth submucosal mass in the mid-oesophagus and another small submucosal mass just above the gastro-oesophageal junction (fig 4). Oesophageal manometry showed a high-pressure zone in the lower oesophagus, which failed to relax during swallowing. There was no effective primary peristaltic wave, and multiple tertiary contractions were noted. This was considered to be diagnostic of achalasia. A Heller's myotomy was performed, and the two submucosal tumours were removed. Histological examination showed granular cell myoblastoma. The tumours were very cellular with pleomorphic, and

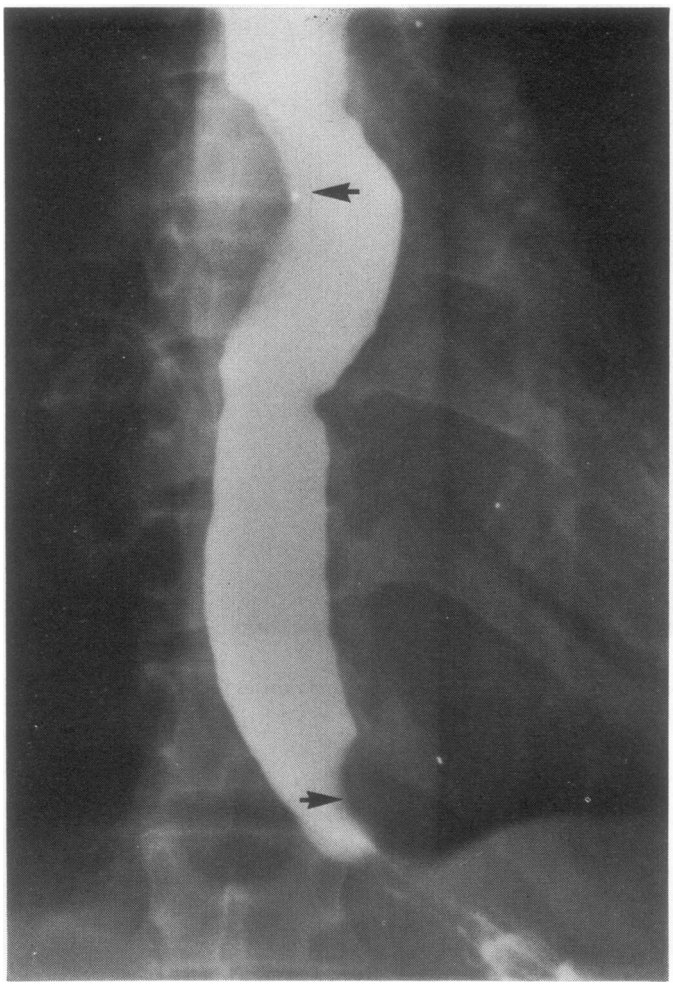


Fig 4 *Case 2. Barium examination shows a large mid-oesophageal submucosal mass and another mass just above gastro-oesophageal junction (arrows).*

occasionally bizarre, vesicular nuclei with prominent nucleoli. These histological features are associated with aggressive tumours and were thought to indicate malignant potential. Six months after operation the patient remains well and has no symptoms.

Discussion

Granular cell myoblastoma of the tracheobronchial tree is rare, representing less than 2% of all myoblastomas (Murphy *et al,* 1949; Umansky and Bullock, 1968; Oparah and Subramanian, 1976). Multiple lesions in the major airways are uncommon. Although lesions from the tongue to the rectum have been described, only 13 cases of GCM located in the oesophagus have been reported (Mansour *et al,* 1977). Multiple oesophageal tumours have not been previously recorded.

Original reports suggested that these tumours arose from embryonic muscle cells (Abrikossoff, 1926), and for many years a myogenic derivation was accepted. At present, however, it is generally accepted that granular cell myoblastoma originates from Schwann cells (Fisher and Wechsler, 1962; Moscovic and Azar, 1967; Mansour *et al,* 1977).

The histological appearance is characteristic. The lesions are very cellular, with sheets or clumps of large polyhedral cells containing abundant granular eosinophilic cytoplasm and small dark vesicular nuclei. The cells tend to be closely packed, have varying shapes, and are arranged in a syncytial fashion (fig 5) (Sobel and Churg, 1964). A characteristic pseudo-epitheliomatous hyperplasia of overlying epithelium is seen in a high proportion of cases and may lead to an erroneous diagnosis of squamous cell carcinoma (Moscovic and Azar, 1967; Booth and Osborn, 1970).

The gross appearance of endobronchial GCM ranges from a plaque-like thickening of the mucosa to a polypoid mass. The lesions range in size from 3 mm to 6·5 cm (Oparah and Subramanian, 1976). Endobronchial GCM is considered a benign tumour, and no case of distant metastasis has been reported. Extrathoracic tumours, however, may occasionally metastasise (Moscovic and Azar, 1967).

Clinically, tracheal and endobronchial GCM presents in a non-specific fashion with dyspnoea, wheeze, and episodes of infection. The chest radiograph may show lobar or segmental atelectasis or consolidation distal to the obstructing lesion. Recurrent infection may cause bronchiectasis as seen in case 1. Bronchotomography may show the endotracheal or endobronchial extension of the mass, although occasionally the submucosal mass will be plaque-like and escape detection on tomography. Bronchoscopy or bronchography will

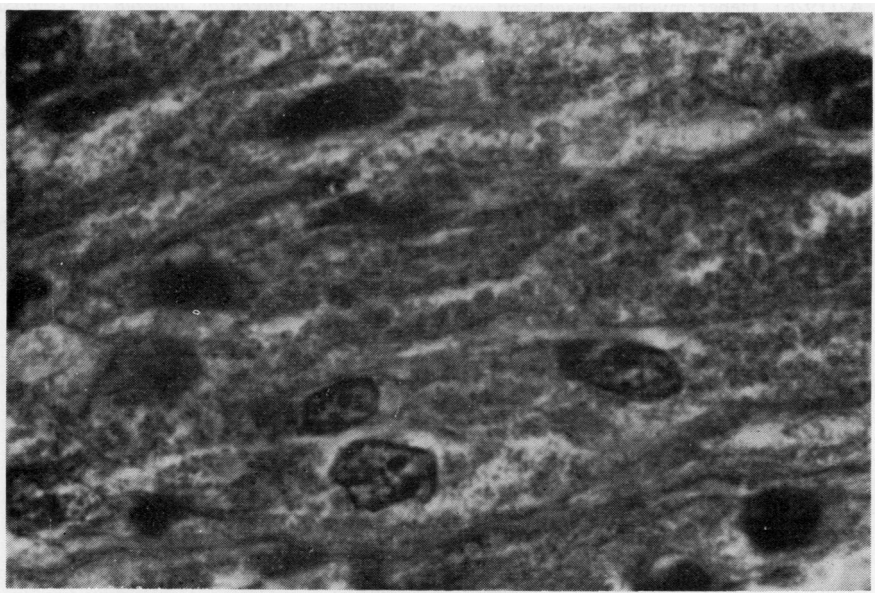


Fig 5 *Case 1. Histological section of lower oesophageal lesion shows characteristic findings of closely packed polyhedral cells containing many eosinophilic granules in cytoplasm and small darkly staining nuclei (H and E ×415).*

usually show the endobronchial extent of the tumour mass, which may be mistaken for a bronchial adenoma. Occasionally the tumour may grow along the bronchial tree to present as a non-specific parenchymal mass lesion (Teplick *et al,* 1975).

Oesophageal granular cell myoblastoma may be asymptomatic or the patient may complain of dysphagia with substernal discomfort. As in case 2, the history of dysphagia may extend over a prolonged period (Crawford and De Bakey, 1953).

The treatment of choice for tracheobronchial or oesophageal GCM is surgical excision. Endoscopic removal of the tumour is not advised as recurrence is likely (Peterson *et al,* 1957). This is because most of the tumour mass is submucosal so that total removal via the bronchoscope or oesophagoscope is difficult. Furthermore, endoscopic biopsy of the submucosal oesophageal mass may be complicated by perforation, infection, and mediastinitis.

The differential diagnosis of multiple tracheobronchial and oesophageal masses includes leiomyomatosis, metastases, amyloidosis, and neurofibromatosis. Despite its rarity, granular cell myoblastoma should also be considered, especially in a patient with an associated skin or tongue mass.

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