

Pancoast syndrome: an unusual presentation of adenoid cystic carcinoma

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ABSTRACT: We report on a patient with primary pulmonary adenoid cystic carcinoma presenting with Pancoast syndrome. Pancoast syndrome has not previously been described with this tumour. Other unusual features of this case include the peripheral origin and mediastinal involvement, with lack of proximal endobronchial spread.

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Pulmonary adenoid cystic carcinoma accounts for 0.2% of primary lung cancers [1]. It is thought to arise from bronchial submucosal glands. The majority are proximal, occurring in the trachea and main bronchi [2]. We report a case of Pancoast syndrome caused by a primary adenoid cystic carcinoma.

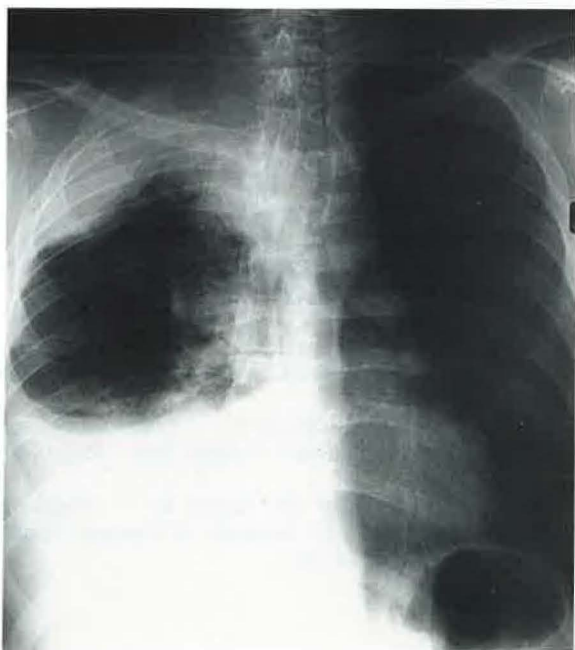


Fig. 1. — Posteroanterior (PA) chest radiograph showing right apical mass with associated tracheal deviation.

Case report

In June 1990, a 43 yr old man presented with pain around the right scapula. During the following months, the pain increased in severity, and radiated down the right arm. In addition, he noticed a right-sided neck swelling and drooping right eyelid. There was a 2 kg weight loss during this period, but he had no respiratory symptoms despite having smoked 75 g of tobacco per week for the last 25 yrs.

On examination he had an ill-defined, soft tissue mass in the right posterior triangle, with right cervical chain lymph node enlargement. The small muscles of the right hand were wasted, with loss of sensation in the T1 distribution, and right Horner's syndrome. Chest signs were left tracheal deviation and diminished breath sounds, with late inspiratory crackles anteriorly over the right upper chest.

The chest radiograph showed a large, lobulated, right apical mass causing the trachea to deviate to the left (fig. 1). Blood investigations included an alkaline phosphatase of 20 U·100 ml⁻¹ (reference range 3-13 U·100 ml⁻¹). At fiberoptic bronchoscopy, external compression reduced the tracheal lumen by 70%; no endobronchial lesion was seen. Computerized axial tomography confirmed the mass, with evidence of direct invasion into the neck, local bone destruction, and widespread mediastinal lymphadenopathy (fig. 2). Biopsy of the mass demonstrated neoplastic cystic spaces lined by glandular columnar and myoepithelial cells, consistent with unequivocal adenoid cystic carcinoma (fig. 3).



Fig. 2. — Computerized axial tomographic scans at the levels of T3-4 showing the tumour mass, direct mediastinal invasion, tracheal deviation and local bone destruction.

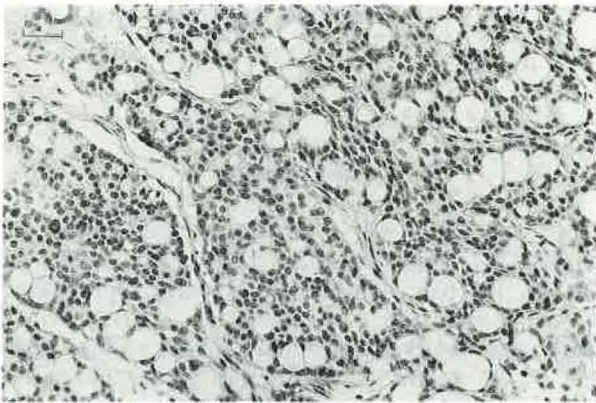


Fig. 3. — Photomicrograph of the biopsy specimen, stained with haematoxylin and eosin (internal marker = 125 μ m).

Discussion

Primary adenoid cystic carcinoma is a rare pulmonary tumour, occurring in the trachea and central bronchi; but 10% have been estimated to arise peripherally in the smaller bronchi [2]. It is a slowly growing tumour, which infiltrates locally into the airway lumen and beneath the bronchial epithelium [3, 4]. Distant spread to lymph nodes and other metastatic sites is unusual; one review of 23 patients reported regional lymph node involvement in 2%, with 26% having more distant metastases [5].

Pancoast syndrome has been described with a variety of malignant and non-malignant conditions, bronchial carcinoma being the commonest cause [6-8].

We believe that this is the first reported case of primary bronchial adenoid cystic carcinoma presenting as Pancoast syndrome. The tumour's peripheral origin and evidence of mediastinal lymphadenopathy in the absence of proximal endobronchial extension are additional unusual features.

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