

Primary mucoid adenocarcinoma of the larynx

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Abstract

Mucous gland carcinomas of the larynx are rare, with most being represented by 'non-specific' adenocarcinoma and adenoid cystic carcinoma. Here we report a unique case of mucoid adenocarcinoma of the larynx occurring in a 46-year-old woman. Despite the presence of regional lymph node metastasis, she remained well four years after surgery.

Introduction

Mucoid (colloid) adenocarcinoma is a well recognized histological subtype of adenocarcinoma which can occur in various organs, such as the breast, gastrointestinal tract, skin, thyroid, pancreas, prostate, and sinonasal tract (Hyams *et al.*, 1988; Rosai, 1989). Histologically, it is characterized by abundant extracellular mucin in which clusters and papillae of carcinoma cells, usually of a bland appearance, float. Clinically it is associated with a better (such as breast, stomach) or worse (such as rectum) prognosis compared with the conventional types of adenocarcinoma in the same organ. To our knowledge,

primary mucoid adenocarcinoma of the larynx has not been previously reported in the English language literature (Cady *et al.*, 1968; Fechner, 1975; Spiro *et al.*, 1976; Cohen *et al.*, 1985; Gadomski *et al.*, 1986; Bloom *et al.*, 1987; Michaels 1987; Hyams *et al.*, 1988). We describe one such case in this report.

Case report

A 46-year-old housewife presented in June 1986 with a two-year history of progressive hoarseness. Direct laryngoscopy under general anaesthesia showed a sessile nodular tumour

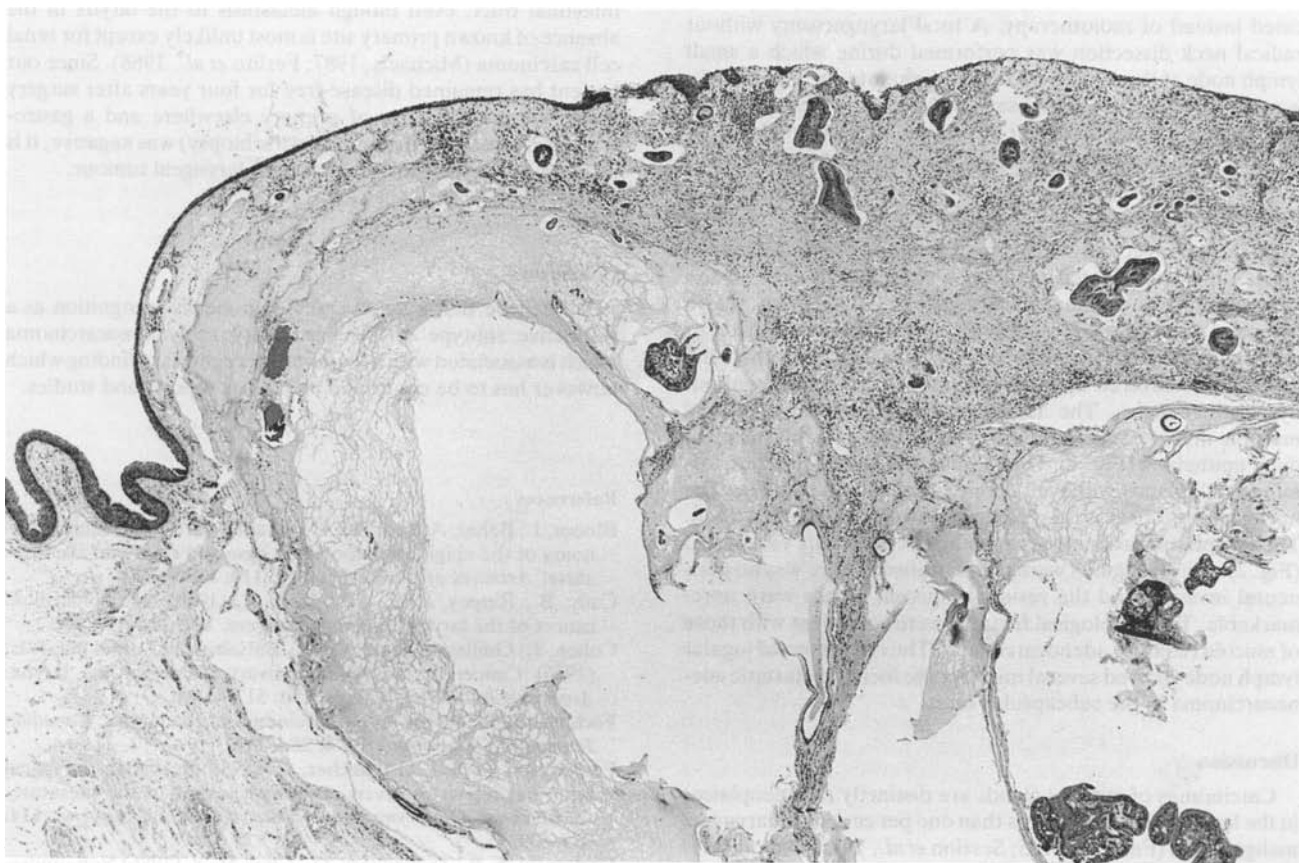


FIG. 1

Beneath the intact covering epithelium are abundant mucin pools containing scattered neoplastic papillae and glands. H&E, $\times 32$.

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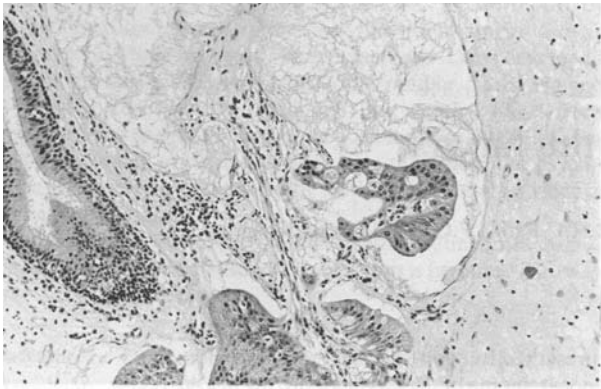


FIG. 2

Tumour papillae are formed by columnar cells possessing mildly atypical nuclei and occasional cytoplasmic vacuoles. H&E, $\times 120$.

involving the right lower half of the laryngeal surface of the epiglottis, right pharyngoepiglottic fold, right ventricular band, and right vocal cord which was fixed. There was no subglottic involvement. A biopsy was interpreted as mucoid adenocarcinoma with some concern that this could represent metastatic tumour. A Barium swallow did not reveal subglottic extension or hypopharyngeal involvement and a chest X-ray did not show any evidence of pulmonary metastasis. There were no palpable cervical lymph nodes. Endoscopic examination of the stomach revealed no tumour.

In view of the extensive local invasion (UICC stage T_4) and the uncertain radiosensitivity of the tumour, surgery was advocated instead of radiotherapy. A total laryngectomy without radical neck dissection was performed during which a small lymph node at the middle part of the right internal jugular vein was also resected. She subsequently acquired oesophageal speech and attended regular follow-up without evidence of loco-regional recurrence or distant metastasis at four years.

Pathological findings

In the laryngectomy specimen, a tumour with gelatinous appearance was located beneath a stretched mucosa in the right supraglottic region, measuring $2.5 \times 2.0 \times 2.0$ cm. It also extended anteriorly and superiorly to form a bulge in front of the epiglottis. Histological examination showed that the mucosa was covered by intact respiratory epithelium without dysplastic change. The lamina propria was expanded by multiple mucin pools containing fragments of neoplastic glandular epithelium (Fig. 1). The tumour cells were columnar, forming small glands with well defined lumina and papillae. The nuclei showed only mild pleomorphism and distinct nucleoli. The cytoplasm was finely granular and occasionally vacuolated (Fig. 2). Mitotic figures were not identified. There was no perineural invasion and the residual mucosal glands were unremarkable. The histological features were consistent with those of mucoid (colloid) adenocarcinoma. The right internal jugular lymph node showed several microscopic foci of metastatic adenocarcinoma in the subcapsular sinus.

Discussion

Carcinomas of mucous glands are distinctly rare neoplasms in the larynx, constituting less than one per cent of all laryngeal malignancies (Fechner, 1975; Session *et al.*, 1975; Gadomski *et al.*, 1986; Bloom *et al.*, 1987; Hyams *et al.*, 1988). Adenocarcinoma of 'non-specific type', adenoid cystic carcinoma and mucoepidermoid carcinoma account for most of these tumours. Their distribution in the larynx corresponds with that of the normal mucosal glands, which are most frequent on the false cords and the subglottic surface of the anterior commissure (Bloom *et al.*, 1987).

Laryngeal adenocarcinomas of 'non-specific' type usually occur in men in their sixth and seventh decades, and are located mostly in the supraglottic region. Both regional lymph node and distant metastasis occur with a high frequency (Cady *et al.*, 1968; Fechner, 1975; Bloom *et al.*, 1987). Most studies have indicated that adenocarcinoma of the larynx is a rapidly lethal neoplasm (Toomey 1967; Cady *et al.*, 1968; Spiro *et al.*, 1976) with Toomey reporting a five-year survival rate of only 12.5 per cent. Wide local excision and radical neck dissection with or without irradiation is the usual recommended mode of treatment. On the other hand, the experience from the Armed Forces Institute of Pathology differs significantly; ten out of 16 patients treated with local or radical surgery were alive and well on follow-up of two to six years (Hyams *et al.*, 1988). Furthermore, Cohen *et al.* (1985) reported a five-year survival of 42.8 per cent, but with most deaths occurring within two years. Notwithstanding these discrepant findings, it appears from the behaviour of our case that mucoid carcinoma may represent a low-grade variant of laryngeal adenocarcinoma. Despite the occurrence of regional lymph node metastasis at presentation, the patient has remained disease-free for four years after surgery. Thus, there may be a need in future studies of laryngeal adenocarcinomas to subdivide them into low and high grade categories similar to those adopted for sinonasal adenocarcinomas (Michaels, 1987; Hyams *et al.*, 1988), which may help to explain the differences in survival from the different series.

The occurrence of mucoid adenocarcinoma in the larynx, an extremely rare tumour which has hitherto not been reported, should prompt a consideration of metastasis from other sites, particularly the kidney, lung, prostate, breast and gastrointestinal tract, even though metastasis to the larynx in the absence of known primary site is most unlikely except for renal cell carcinoma (Michaels, 1987; Ferlito *et al.*, 1988). Since our patient has remained disease-free for four years after surgery alone without evidence of primary elsewhere and a gastroscopic examination (including gastric biopsy) was negative, it is justified to consider this as a primary laryngeal tumour.

Conclusion

We believe that mucoid carcinoma merits recognition as a distinctive subtype of low grade laryngeal adenocarcinoma which is associated with a favourable prognosis, a finding which however has to be confirmed by further reports and studies.

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