Carcinoid tumours of the larynx: Report of two cases

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Abstract

Carcinoid tumour of the larynx is extremely rare. The increasing number of these lesions reported is due to the application of ultrastructural and immunological methods in the diagnosis. Two new cases of this malignant neoplasm are presented and discussed with particular reference to the difficulty in distinguishing carcinoids from other tumours of the larynx. The accurate pathological identification of the tumour is essential for treatment and prognosis.

Introduction

Carcinoid belongs to the group of 'Apudomas', histogenetically deriving from the so-called APUD-system. The apud cell, defined by Pearse (1974) has a high intracellular concentration of amines and a capacity for endocrine activity. In addition, these cells contain intracellular granules; because of the similarity of these granules to those found in neurons, apud cells are thought to originate from neural ectoderm. The notion that neuroendocrine cells are of neural crest derivation is questionable. Alternatively carcinoids may be derived from primitive cells capable of multidirectional differentiation (De Lellis et al., 1984; Mills and Johns 1984).

Williams and Sandler (1963) classified these tumours into foregut, midgut and hindgut carcinoids. Foregut tumours are usually argentaffin, while those of the midgut exhibit a positive argentaffin reaction due to the presence of biogenic amines with endocrine activity. Carcinoid tumours, which typically arise in the appendix and small intestine (midgut) without showing an aggressive course, have also been noted in the respiratory tract (foregut) and more rarely in the larynx.

Bronchial and tracheal carcinoids tend to be locally invasive, while laryngeal carcinoids are frankly malignant and potentially metastasizing lesions (Godwin, 1975; Briselli et al., 1978; McDougall et al., 1980).

Carcinoid tumour is a rare neoplasm of the larynx and 47 cases, including atypical carcinoids, are reviewed and listed by Falito et al. (1988) (five cases up to 1980 and the remainder since). A recent publication of the entire 80-years experience with neural and neuroendocrine tumours of the larynx at the Mayo Clinic, included five carcinoids (Stanley et al., 1987).

We report two cases of this uncommon tumour and emphasize the importance of their recognition and distinction from other epithelial neoplasms because some carcinoids display an aggressive clinical behaviour and may require complete laryngectomy with neck dissection.

Case report

Case 1

A 59-year-old man, was admitted complaining of hoarseness and slight dysphagia. He had a past history of recurrent laryngitis and had an angiomatous-like lesion of the right arytenoid for many years. The patient had smoked 15 cigarettes daily and consumed small quantities of alcohol. He had had pulmonary tuberculosis some years earlier, and two months prior to admission a painful nodule of the scalp had appeared, which was later diagnosed as a metastatic lesion from the laryngeal tumour.

Indirect laryngoscopy showed a small roundish swelling covered by normal mucous membrane on the right aryepiglottic fold and an ulcerated lesion of the right ventricular fold which extended to the ipsilateral arytenoid; the right vocal cord was fixed. There were no enlarged cervical nodes present and the remainder of the examination was normal.

Biopsy of the laryngeal lesion was interpreted as showing an undifferentiated carcinoma.

A right hemilaryngectomy was performed and the frozen section diagnosis was infiltrating adenocarcinoma (December, 1981). Histological examination of the laryngectomy specimen led to the diagnosis of a carcinoid tumour. No signs suggestive of carcinoid syndrome were noticed and the urine assay for 5-HIAA showed normal values.

One year later, indirect laryngoscopy revealed a recurrence in the right hemilarynx. Surgical re-operation showed that this lesion extended through the preceding thyrotomy infiltrating the prelaryngeal muscles; a total laryngectomy with right modified neck dissection and hemithyroidectomy was performed.

Since then the patient developed peristomal and parapharyngeal...
Carcinoid tumour: argyrophil-positive granules within tumour cells (Grimelius x40).

Ryngeal recurrences associated with many painful metastatic nodules of the skin. These were surgically removed during further hospitalization, palliative radiotherapy, followed by chemotherapy, were carried out for the local spread.

However, the disease showed no signs of remission and the patient died due to carcinomatosis five years after the diagnosis of the laryngeal tumour.

Case 2

A 36-year-old female was examined because of intermittent hoarseness. Indirect laryngoscopy demonstrated a small swelling of the left ventricular fold with no signs of erosion. Laryngeal motility was normal and there was no cervical lymphadenopathy. Direct laryngoscopy confirmed these observations. A biopsy was taken from the swelling which was interpreted as a poorly differentiated carcinoma. A supraglottic laryngectomy was performed and histopathological examination of the specimen revealed a carcinoid.

The patient is still free of disease six years after the operation.

Discussion

Carcinoid tumours of the larynx are very rare, although a review of the literature reveals an increase of cases reported in recent years (Ferlito et al., 1988). The analysis of accumulated experiences allows us to define the nature of this unusual neoplasm and its management.

Carcinoids have arisen from the supraglottic larynx in all cases with few exceptions; the growth rate is lower than that of carcinomas. Carcinoid syndrome in the larynx has never been reported, not even in the two cases which showed elevated levels of 5-HIAA (Baugh et al., 1987).

The tumour shows a submucosal origin with a behaviour tendency to high local invasion (cartilage, lymphatics, blood vessels or perineural spaces). Cervical lymphadenopathy may be present at the time of operation.

Regional recurrences and distant metastases (especially skin-involvement) may occur frequently even after long periods of remission, and the survival rate is poor (Blok et al., 1985; Guerrieri et al., 1985).

It may be difficult to distinguish carcinoids from other tumours of the larynx. In many cases, ours included, the tumour was misdiagnosed on the first histological examination, most commonly as an anaplastic or undifferentiated carcinoma, more rarely as an adenocarcinoma (Paladugu et al., 1982; Weighill et al., 1986). The correct histological diagnosis of carcinoid has usually been obtained only after laryngeal surgery. Silver staining and ultrastructural studies have led to a correct
diagnosis of carcinoid. Microscopically, the neoplasm is arranged in nests of uniform cells with granular eosinophilic cytoplasm (Fig. 1); Grimelius silver stain reveals the granules to be strongly positive (Fig. 2) and electron microscopy is of the greatest importance to confirm the diagnosis (Fig. 3).

Carcinoids are not radiosensitive (although many patients have received this treatment) and no response to chemotherapy has been demonstrated (Moertel et al., 1986). Conversely, some forms of poorly differentiated adenocarcinoma, most poorly differentiated squamous carcinomas and small cell carcinomas of the larynx show responsivity to radiation and multidrug chemotherapy.

The treatment of choice is extensive surgical extirpation of the lesion (when possible by conservative surgery), with an associated modified neck dissection if there is clinical evidence of cervical lymphadenopathy. Long-term follow-up is necessary to recognise the development of metastatic spread.

References

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