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# Primary tumours of the trachea: analysis of clinical features and treatment results

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**Abstract**

Primary tracheal tumours are extremely rare and present with widely variant clinical and histological features. Treatment methods vary considerably, and few studies have sought to provide adequate guidelines. A retrospective analysis was carried out of all patients treated in our unit between 1965 and 1990. Our experience deals almost exclusively with high tracheal tumours involving the adjacent subglottic region. Squamous carcinoma (SCC) and adenoid cystic carcinoma (ACC) were the commonest subtypes, and presented with dyspnoea and hoarseness as the most frequent symptoms. ACCs occurred commonly in young individuals, presented insidiously, and ran a long, and often, unpredictable course. Endoscopic evaluation revealed the majority of the lesions to be bulky and obstructive in nature. Primary surgery with adjuvant radical radiotherapy, when indicated, appeared to provide optimal results. Debulking surgery followed by radiotherapy provided effective and lasting control in two cases of ACC. Other malignant subtypes behaved aggressively and progressed uncontrolled.

**Key words: Tracheal neoplasms**

## Introduction

Primary malignant tumours of the trachea are extremely rare entities and account for a mere 0.2 per cent of all malignancies of the respiratory tract (U.S. Department of Health and Human Services, 1981) and 0.04 per cent of all malignant neoplasms reported (Manninen, *et al.*, 1991). The largest series, reported by Grillo and Mathisen (1990), very convincingly reflected the application of modern techniques of tracheal resection and reconstruction.

This report presents the experiences of the Professorial unit at the Royal National Throat, Nose and Ear Hospital from 1965 to 1990, a period of 25 years, in the treatment of primary tumours involving the trachea. Being an otolaryngology unit, our referrals were largely restricted to lesions affecting the cervical trachea and the contiguous subglottic region.

Hospital records of all patients were examined in detail, and every effort made to obtain up-to-date follow-up information.

## Materials and methods

*Patient characteristics*

A total of 14 patients were treated for primary tumours of the trachea over the period of the study, of whom 13 had malignant lesions. Five patients had SCCs, five had ACCs, one had a large cell neuroendocrine carcinoma, one had a cribriform adenocarcinoma, and one had a plasmacytoma. The solitary patient with a nonmalignant lesion was a 13-year-old girl with a granular cell myoblastoma.

All patients with SCC had a long history of smoking, while three out of five with ACC were known smokers. The age distribution (Table I) reflects a conspicuous distribution between the fifth and seventh decades of life (10/14 cases). With SCC, the mean age was 61 years and the median age 63 years, while with ACC, the corresponding ages were 60 and 50 years respectively. The youngest patient in the latter group was 29 years old.

Gender distribution revealed a female preponderance (9/14 cases). Twelve of the 14 patients had lesions involving the upper cervical trachea and subglottis, one patient had a long segment disease extending from the subglottis to the carina, and one had an ACC producing a low tracheal mass in the region of the carina. In 10 patients, precise information concerning the duration of symptoms was available. Amongst those with ACC, three out of four (where information was available) experienced symptoms of insidious onset and gradual progression lasting over a year. In one lady, a small tumour was incidentally noticed at thyroidectomy and biopsied from the region of the upper trachea. Since the pathology came back as ACC, she was subjected to irradiation which controlled the disease for 13 years, before a massive relapse.

With SCC, the duration of symptoms was shorter, and did not exceed three months in all except one patient.

The most frequent symptoms were dyspnoea and hoarseness, and were present in eight patients of each type. A

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TABLE I
AGE DISTRIBUTION: PRIMARY TRACHEAL TUMOURS (N = 14)

| Age (years) | SCC | ACC | Others |
|---|---|---|---|
| 0–10 | – | – | – |
| 11–20 | – | – | 1 |
| 21–30 | – | 1 | – |
| 31–40 | 1 | 1 | – |
| 41–50 | – | – | – |
| 51–60 | 1 | 1 | 1 |
| 61–70 | 1 | 2 | 1 |
| 71–80 | 2 | – | 1 |

notable symptom mentioned by some was a 'frog-in-the-throat' sensation. Other manifestations included haemoptysis, pneumonia, and dysphagia. One patient of ACC presented with a woody, hard midline neck swelling which was thought to originate in the thyroid, while another presented with total dysphagia with aspiration due to tracheal overspill.

Amongst radiological evaluation methods, tomography was the most frequently employed, and provided adequate information about the vertical extent and luminal involvement. CT scanning, which has been in use since 1978, provides additional information about tracheal wall thickness and adjacent soft tissue involvement. All patients were evaluated endoscopically, and the most frequently used technique was microlaryngotracheoscopy. In the earlier period of the study, rigid bronchoscopy was used, but has now been replaced by safer methods. Initial evaluation today always comprises flexible, fibre optic nasendoscopy performed under topical anaesthesia.

Nine out of 11 patients in whom detailed morphological descriptions were available had bulky, obstructive lesions. With ACC, one patient had an invasive, submucous, circumferential pattern of involvement and another had a carpet-like pattern of spread down to the level of the carina.

*Treatment*

The major treatment modalities employed were surgery and radiotherapy, alone and in combination. Table II outlines the essential treatment profile for the group.

In all cases, except one, surgery involved total laryngectomy, with manubrial resection in five cases to obtain adequate margins. Two patients required circumferential laryngopharyngectomy, one of whom was reconstructed by gastric transposition. The thyroid gland was managed according to the needs of each individual case. In one patient of ACC treated in 1967, primary surgery consisted of complete endoscopic debulking of all gross tumour, since it was situated just above the carina and considered unresectable then. Radiotherapy was administered thereafter.

TABLE II
TRACHEAL TUMOURS: TREATMENT PROFILE (N = 14)

| Mode | SCC | ACC | Others |
|---|---|---|---|
| S alone | 1 | 1 | 1 |
| S + RT | 1 | 2 | 1 |
| RRSS | 2 | 1 | 1 |
| Laser | 1 | 1 | 1 |

S: primary surgery; RT: radiotherapy; RRSS: Radical radiation with surgical salvage.

Radiotherapy was delivered by cobalt 60 machines in the earlier part of the study and by Linear accelerators in recent years. All cases received radical radiotherapy averaging 60 Gy over 30 fractions.

Laser debulking was performed in one patient with an ACC involving almost the entire length of the trachea, and followed by radical radiotherapy. Amongst the other two patients treated by laser, one elderly lady had an apparently pedunculated SCC of the upper trachea, for which no other treatment could be administered. On three occasions spaced two months apart each time, she was brought to the hospital with inspiratory stridor and sent back after palliative laser therapy to open up the airway. On the fourth episode she died, presumably of asphyxia. The other patient to be managed only by laser was a young girl with a granular cell myoblastoma, who needed two resections in all.

*Complications*

One patient developed a pharyngeal leak following laryngectomy and was managed conservatively with success. Stomal stenosis developed in two out of five patients who had manubrial resections, and needed corrective stoma plasties. Hypothyroidism and hypoparathyroidism occurred in two patients, and one patient developed a major and fatal vascular catastrophe following relapse of an operated tracheal adenocarcinoma. The patient with a benign lesion developed subglottic and tracheal stenosis which demanded division and a silastic keel.

**Results of treatment** (see Table III)

Adequate follow-up information was available in all except one case, where salvage surgery was performed for a recurrent SCC in a 37-year-old man from a foreign country.

All patients of SCC as well as ACC who received primary surgery remained alive without disease at the last follow-up. Two of them required prosthetic speech rehabilitation, and were successfully fitted with Blom-Singer valves, in one case after a stoma plasty and myotomy.

The two patients with ACC who had primary debulking and radiotherapy also remained controlled. The only patient with a benign lesion remained disease-free as well.

Amongst the two patients of SCC who had salvage surgery, one died at home following massive haemoptysis, presumably due to a local relapse. The other patient was lost to follow-up.

The only patient of ACC who received salvage surgery following a relapse at 14 years post-radiotherapy went on

TABLE III
TRACHEAL TUMOURS: TREATMENT RESULTS (N = 14)

| | Status at last follow-up | | |
|---|---|---|---|
| Mode | SCC | ACC | Others |
| S alone | 1 NED | 1 NED | 1 DOD |
| S + RT | 1 NED | 2 NED | 1 DOD |
| RRSS | 2 DOD, LFU | 1 DOD | 1 AWD |
| Laser | 1 DOD | 1 NED (+RT) | 1 NED |

NED = no evidence of disease; DOD = dead of disease; DWD = dead with disease; LFU = lost to follow-up; AWD = alive with disease.

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to develop lung metastases, and succumbed at 16 months post-surgery.

All patients with malignant subtypes other than SCC and ACC failed treatment. Two were dead and one alive with distant metastases of a plasmacytoma at the last follow-up.

## Discussion

Our rather small experience of 14 cases presents a valuable historical perspective which, along with other reported studies, should help improve our understanding of this rare group of tumours.

The histological distribution in our series, namely five SCC (36 per cent), five ACC (36 per cent), and four others (28 per cent) was nearly identical to that of the largest series reported by Grillo and Mathisen (1990). However, a nationwide epidemiological survey from Finland (Manninen *et al.*, 1991) reported a high preponderance of SCC (72 per cent). As for age distribution, all studies seem to agree that ACC generally afflicts younger individuals than does SCC.

While our small study reflects a female preponderance when considering gender distribution, others reflect interesting differences. For SCC, all reports mention a distinct male pattern. With ACC, while the American study reflects an even sex distribution (male/female = 41:39), the Finnish study reflects a strong female pattern (male/female = 1:5).

Concerning symptomatology, we found dyspnoea and hoarseness to be the commonest symptom. The series reported by Li *et al.* (1990), on the other hand, mentioned haemoptysis as the commonest symptom, followed by dyspnoea and hoarseness.

Radiological assessment in our series was largely by conventional tomography. Other studies by Li *et al.* (1990) and Aberle *et al.* (1991) speak of the distinct superiority of CT scanning, particularly thin section scanning with intravenous contrast. Clearly the most vital consideration of all is the plan of treatment and its anticipated success. One has to choose between surgery alone, radiation alone with surgery for salvage, and surgery followed by planned adjuvant radiotherapy. Our study as well as the other two reported, one from Grillo and Mathisen (1990) and the other from a radiotherapy group (Fields *et al.*, 1989) suggest convincingly that the last of the three options is most likely to work.

Two patients in our study remained controlled after debulking surgery and radiotherapy for ACCs, surviving six and 14 years respectively without disease. Since no comparable data are available, no clear recommendations can be made. However, the approach deserves consideration in cases of ACC where definitive surgery cannot be offered as primary treatment. With the availability of the laser, debulking can be accomplished with considerable precision.

Considering the technical aspects of surgery, there is no doubt that the larynx must be spared whenever possible, and the series by Grillo and Mathisen (1990) amply illustrates this. Unfortunately, none of our patients were suitable candidates, on account of the high incidence of subglottic involvement. Resection of the manubrium considerably aids obtaining adequate margins following laryngo-tracheal resection.

The biological behaviour of different histological types was rather distinct, as seen in our study and other studies (Nomori *et al.*, 1988), and provides useful insights. While ACCs present insidiously, have a long natural history and behave capriciously, SCCs generally present sooner and demonstrate a predictable, early pattern of relapse.

All other malignant subtypes behaved aggressively and failed treatment in our experience. The numbers are too small but the addition of chemotherapy may benefit these rare histological types.

## Conclusions

It must be stated that malignant tracheal tumours, though uncommon and fatal if untreated, are amenable to effective control in a significant number of patients.

Definitive primary surgery, either in the form of limited tracheal resection with primary closure, or laryngo-tracheal resection when necessary, should be regarded as the cornerstone of management. Adjuvant radiation is indicated in all patients treated by tracheal resection alone and in those cases of laryngo-tracheal resection with unfavourable histological characteristics such as positive nodes, close margins, extra-tracheal spread, etc.

Precise and thorough debulking by laser may be satisfactorily employed prior to radiotherapy in inoperable cases of ACC. Laser resection alone must be reserved only for selected benign lesions or for palliation of airway obstruction in advanced cases.

The role of primary radiation with surgery for salvage is limited.

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