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Original Article

Lower Respiratory Tract Adenoid Cystic Carcinoma: Its Management in the Past Decades

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Abstract

Aims: Adenoid cystic carcinoma of the lower respiratory tract is a rare indolent neoplasm with prolonged survival, propensity for recurrences and metastasis years after initial therapy. We aim to study a 1,700-bed single tertiary academic hospital's long-term experience with ACC of the lower respiratory tract from the larynx to the lungs and review published literature on this subject.
Materials and methods: We analysed the clinicopathology, treatment options and outcome in 33 patients and reviewed the published literature over the last five decades.
Results: The tumour has no gender predilection, a peak incidence in the fifth decade and is not related to smoking. Insidious symptoms are often treated as benign obstructive airway disease and infection; negative signs and normal chest X-rays delayed diagnosis. The tumour was distributed most commonly in the trachea followed by main bronchi, lobar bronchi and larynx. About 22% of patients required emergent bronchoscopic intervention to secure airway patency before definitive therapy with surgery or/and radiotherapy. A high proportion of resected specimens had positive margins. Overall survival and disease-free survival rates at 5 years were 81 and 62%, respectively, and at 10 years 70 and 54%, respectively. Prolonged good palliation was achieved for patients with unresectable lesions with radiation and wide armamentarium of endoscopic therapy.
Conclusions: In time, many patients eventually succumb to this disease. However, advances in medical skill and technology have prolonged survival while maintaining a good quality of life. Adenoid cystic carcinoma of the respiratory tract is a chronic life-long disease that may require interval intensive therapy. The challenge is to find the best therapeutic regimen aiming for a 'true' cure. Further study on the mutational landscape of adenoid cystic carcinoma may provide potential avenues for novel treatments to address a chemoresistant cancer.

Key words: Bronchoscopic intervention; radiotherapy; salivary gland tumour; surgery; systemic therapy

Introduction

Primary tracheal tumours comprise about 0.2% of all respiratory tract cancer, with 10–20% being adenoid cystic carcinoma (ACC) [1–4]. Because of its rarity, almost all published data come from case reports and short series. Much of what is known has been from studies of this tumour in salivary glands of the head and neck [5–7]. We report a hospital's long-term experience with ACC of the lower respiratory tract and a review of the published literature.

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Materials and Methods

With approval from the Ethics Committee and Hospital Institutional Review Board (number 2013/861/C) and waivers of consent from patients, a retrospective review of patients with ACC of the lower respiratory tract between January 1991 and October 2013 was conducted. The search encompassed inpatient discharge diagnoses and procedures, including surgical and pathology databases. Hospital records including clinical, radiological and pathological charts of patients with ACC from the larynx to the lungs were studied. The clinico-radiological data available for review were evaluated and analysed from the time of primary site diagnosis of ACC to the time of death or the end of the study. Patients with pulmonary metastasis from ACC of the salivary

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glands in the head and neck were excluded from the treatment and outcome analyses. All categorical variables are reported as counts and percentages and continuous variables are reported as means and ranges. Overall survival from the time of treatment was calculated using the Kaplan–Meier method.

Results (Table 1)

Clinical Presentation

There were 33 patients with a mean age of 48.2 (range 23–67) years at the time of diagnosis. Twenty-nine patients

Table 1
Clinical, radiology, bronchoscopy, pathology and treatment data of 33 patients with adenoid cystic carcinoma of the lower respiratory tract

Study period 1983–2013	Total, $n = 33$
Age	48.2 (range 23–67) years
Gender ratio	Male:female = 15:18 = 1:1.2
Smoking	Smoker:never smoker = 13:15 = 1:1.2
Symptoms	Evaluated = 29 (%)
Cough	21 (72.4)
Dyspnoea	20 (69.0)
Haemoptysis	14 (48.3)
Wheeze	11 (38.0)
Chest discomfort	5 (17.2)
Loss of weight	4 (13.8)
Signs	
Wheeze/stridor	12 (41.4)
Collapse/consolidation	6 (20.7)
Tachypnoea	2 (6.9)
Nil	16 (55.2)
Imaging	Evaluated = 22 (%)
Chest X-rays	
Mass	7 (31.8)
Collapse/consolidation	5 (22.7)
Normal	9 (40.9)
Computed tomogram	
Airway mass	14 (63.6)
Lung mass	4 (18.1)
Lung collapse	5 (22.7)
Lung or pleural nodules	2 (9.1)
Bronchoscopy	Evaluated = 19 (%)
Stenosis	15 (78.9)
Mass	14 (73.7)
Mucosal infiltration	3 (15.8)
Normal	2 (10.4)
Diagnostic yield*	27 (93.1)
Pathology – Primary sites	Total, $n = 33$
Trachea	12 (36.4)
Main bronchi	8 (24.2)
Lobar bronchi	6 (18.1)
Larynx	1 (3.0)
Salivary glands (all lung lesions)†	6 (18.1)
Resected specimens	Evaluated = 21 (%)
Margins positive	15 (71.4)
Perineural invasion	15 (71.4)
Lymphatic invasion	6 (28.6)
Treatment	
Primary adenoid cystic carcinoma of the respiratory tract	Evaluated = 27 (%)
Surgery	10 (37.0)
Surgery, radiotherapy	8 (29.6)
Bronchoscopic intervention, surgery	2 (7.4)
Bronchoscopic intervention, surgery, radiotherapy	1 (3.7)
Bronchoscopic intervention, radiotherapy	3 (11.1)
Radiotherapy	3 (11.1)

* Of 29 patients who underwent bronchoscopy. Of the two patients with a negative bronchoscopic yield, one had the histological diagnosis made during surgery and the other on transthoracic needle biopsy of the lung lesion.

† All lung lesions were solitary peripheral lung metastases of salivary glands in the head and neck region and these patients were excluded from treatment and outcome analysis.