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Multiple primary laryngotracheal adenoid cystic and left thyroid follicular carcinoma with right vocal cord paralysis: the crucial role of surgery

S. CAFAROTTI, A. CESARIO*, V. PORZIELLA, S. MARGARITORA, G. CARNASSALE, P. GRANONE

General Thoracic Surgery Unit, School of Medicine, Catholic University of the Sacred Heart, Rome, Italy
*IRCCS San Raffaele Pisana, Rome, Italy

Abstract. – BACKGROUND: Primary tracheal tumors are rare, accounting for only 0.2% of all thoracic cancers. Adenoid cystic carcinoma (ACC) diagnosed in the trachea is very uncommon and its coexistence with second histologically distinct malignant tumors of he neck region has never been reported.

SUMMARY: We now report a case of multiple primary laryngotracheal ACC and thyroid follicular carcinoma surgically successful treated with an incidental 8 years follow-up.

CONCLUSIONS: The laringotracheal resection with *en-bloc* thyroidectomy can be adopted for treating multiple primary tumor of tracheal and thyroid carcinoma with good long-term prognosis.

Key Words:
Tracheal tumor, Thyroid tumor, Multiple primary tumor, Collision tumor.

Introduction

Adenoid cystic carcinoma (ACC) is a malignant neoplasm originating in the salivary glands, accounts for less than 0.2% of all lung tumors. Primary ACC diagnosed in the trachea or in the bronchial tree, likely originating from submucosal seromucinous glands, is very uncommon. The natural history and response to therapy differ from that of other malignant tracheo-bronchial tumors. Inspite of the relatively benign evolution, this disease has a high risk of incomplete resection because its extension is commonly beyond the visible gross tumor. Even though unresected cases can be controlled successfully for many years, ACC is considered relatively resistant to treatment, and can metastatize late in the course of disease. The purpose of the present case is to report, for the first time in literature, a multiple primary tracheal ACC with right recurrent laryngeal nerve paralysis and left thyroid follicular carcinoma surgically successful treated with an incidental 8 years follow-up.

Case-Report

A 40-year-old non-smoker woman with no allergies and without cellular and humoral immunity deficit was referred for a sublaryngeal tracheal stenosis after examination with computed tomography (CT) because of persistent wheezing and dysphonia. The CT showed trachea stenosis and left lobe thyroid mass (Figure 1). The flexible bronchoscopy showed right vocal cord paralysis and confirmed the stenosis (Figure 2).

A biopsy sample showed citology of thyroid left lobe carcinoma and tracheal ACC. Normally, when preoperative unilateral vocal cord paralysis is present, we preserve the contralateral thyroid lobe. However, in this situation, total thyroidectomy and laringotracheal resection extending up to right laryngeal nerve and down to four tracheal rings without permanent tracheostomy were performed (Figure 3).

During resection the patients were no kept ventilated by means of a distal intubation tube (Figure 4).

Concerning laryngo-tracheal end-to-end anastomosis we used for the posterior wall a continuous 4.0 polydioxanone suture between the cartilaginous-membranous angles and interrupted sutures for anterior wall. There were described a number of releasing maneuvers to have a tension-free anastomosis, but in this case we used only cervical flexion. No airway obstruction as a result of glottic edema, anastomotic dehiscence, permanent hypoparathyroidism. pneumonia or tracheoesophageal fistula were observed in postoperative period.

Corresponding Author: Stefano Cafarotti, MD; e-mail: cafarottistefano@gmail.com

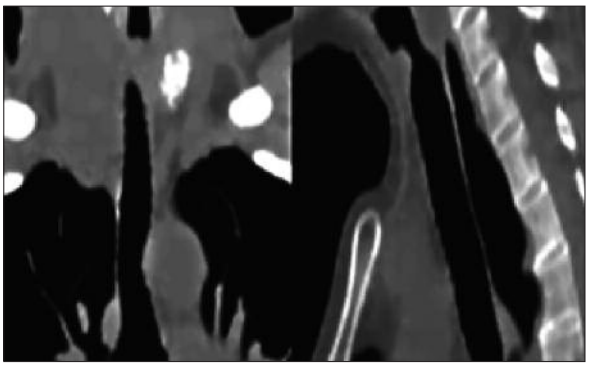

Figure 1. CT- scan.

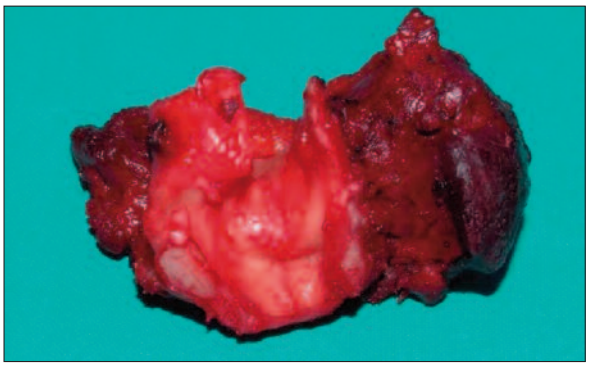

Figure 3. Thyroid end laryngotracheal resection.

The histopathological diagnosis after complete resection was tracheal ACC with right laryngeal nerve infiltration and left lobe thyroid follicular carcinoma (Figure 5).

Irradiation of the treated site with the photon beams of a 6 MV linear accelerator was started 8 weeks later and after bronchoscopy. The clinical target volume (CTV) was considered the site of the resected tumors with a margin of 2.5 cms and the planning target volume (PTV) was obtained adding a further 0.5 cm margin. A total dose of 64 Gy in 30 daily fractions has been administered. The patient has had no recurrence after a 8-year follow-up period.

Discussion

Adenoid cystic carcinoma is a rare primary tumor of the upper airway, which is frequently amenable to resection and reconstruction by primary anastomosis. At present, such surgery can be accomplished with a reasonably low operative mortality rate and remarkably good long-term survival, particularly if survival is compared with that observed with the more common primary lung cancers. ACC are infiltrative tumours that cause irregular narrowing of the airways. The level of invasion seen microscopically is nearly always greater than what is grossly apparent and hence complete resection may be quite difficult to achieve. They usually spread to adjacent structures with perineural spread being common although the infiltration of recurrent laryngeal nerve is not frequent. Rarely, metastases occur to lymph nodes, bone, kidney, liver, lung, and brain. The mainstay or treatment is complete resection of the tumour, which can lead to good long term survival[1]. A clear difference in survival is reported between resected and unresected patients (Table I).

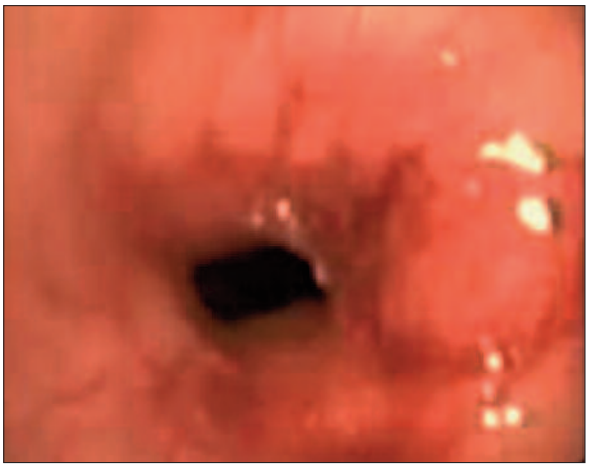

Figure 2. Pre-operative broncoscopy.

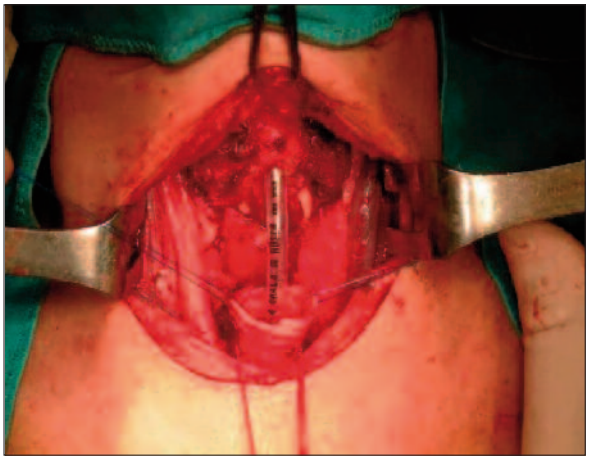

Figure 4. Technique of intubation.

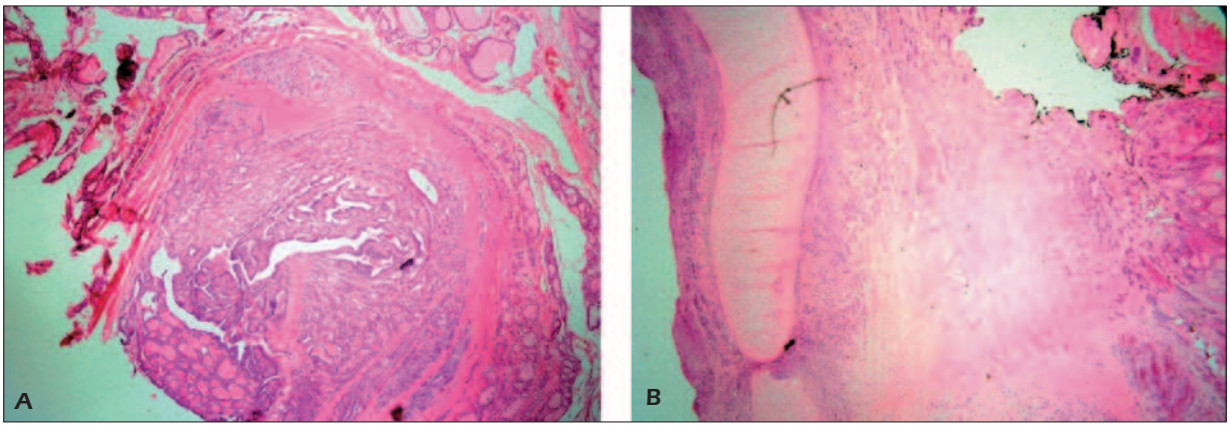

Figure 5. *A*, Thyroid histology (hematoxylin-eosin, magnification 4×). *B*, Laringotracheal thyroid histology (hematoxylin-eosin, magnification 10×).

Multiple primary tumors of neck region are especially uncommon. Only one similar case has been reported in the literature, by Jacobson et al[2], in which papillary thyroid carcinoma "colliding" with laryngeal squamous cell carcinoma. The term "collision tumor" refers to the coexistence of two histologically distinct malignant tumors within the same mass. In our case, probably, we discovered the mass before collision due to the early diagnosis guaranteed by recurrent laryngeal nerve paralysis (the two distinct tumors were at 1 cm of distance).

As some Authors[3-5] reported the double tumors phenomenon is attributed to cellular and humoral immunity deficit and has been described in different processes, such as chronic lymphocytic leukemia or the immunosuppression induced in a transplant, where a notable increase in certain neoplasms has been reported: melanoma, soft-tissue sarcoma and bronchopulmonary carcinoma. Our patient had no history of immunosuppression. After the pathological report, hematologic and serologic studies were carried out, with normal results. Another carcinogenic theory of the double tumor would be the "mechanical" stress or history of smoking[6] but our patient was non smoker. Another hypothesis is that the presence of the first tumor alters the microenvironment, making the development of the second adjacent tumor more likely. Interestingly, certain pairs of collision tumors occur relatively more frequently: hepatocellular carcinoma with cholangiocarcinoma[7] and gastric adeno-

Table I. Summary of the most relevant 25 years series of ACC of the tracheo-bronchial tree.

Author	Year	Cases	Therapy	5-y surv.	10-y surv.
Maziak et al[10]	1996	32	Surgery + XRT	79%	51%
Regnard et al[11]	1996	67	Surgery	73%	57%
Perelman et al[12]	1996	66	Surgery	35.9%	27.1%
Prommegger and Salzer[13]	1998	16	Surgery	79%	57%
Kanematsu et al[14]	2002	16	Surgery	Res. 90% Unres. 40%	Res. 76% Unres. 0%
Bhattacharyya[15]	2004	19	Surgery	78.3%	–
Gaissert et al[16]	2004	135	Surgery + XRT	Res. 52.4% Unres. 33.3%	Res. 29% Unres. 10%
Clough and Clarke[17]	2006	13	Surgery	Better mean survival in resected pts (66 months) vs unresectable pts. months)	
Webb et al[18]	2006	19	Surgery ± XRT	42%	26%
Molina et al[19]	2007	39	Surgery ± XRT	Surgical pts. 57%, non surgical pts. 53%	Surgical pts. 45%, non surgical pts. 31%

XRT: radiotherapy, Res.: resecable, Unres.: unresectable; vs: versus, pts.: patients.

carcinoma with lymphoma[8]. In the thyroid, combinations of papillary with medullary thyroid carcinoma or follicular with papillary with medullary carcinoma have been described[9]. These tumors have arisen either as a coincidental ''meeting'' phenomenon or from a common stem cell. In this reported case, the mechanism is not clear and involved two different organs.

Conclusions

Despite their generally slow and indolent growth in other locations, ACC of the tracheobronchial tree may be aggressive tumors. The laringotracheal resection with en-bloc thyroidectomy is technically feasible and can be adopted for treating multiple primary tumor of tracheal and thyroid carcinoma with good long-term prognosis

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