CASE REPORT

Primary Adenoid Cystic Carcinoma of the Trachea: A Case Report of a Twelve Year Survivor

Naoyuki Kohno, Hideki Tateno, Masahiro Kawaida and Hiroyuki Fukuda

Department of Otolaryngology, Juntendo University School of Medicine, Tokyo, 1 Saiseikai Kanagawaken Hospital, Yokohama, 2 Ohtuka Metropolitan Hospital and 3 School of Medicine, Keio University, Tokyo, Japan

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Abstract. Malignant tumors arising from trachea are not common. This paper presents an example of primary tracheal adenoid cystic carcinoma treated by surgical resection with good prognosis. A 46-year-old woman presented with a short history of dyspnea. Five months before the onset of dyspnea, the patient had mild wheezing. She had no history of smoking. Physical examination suggested primary tracheal tumor. The patient underwent a V-shaped resection of 3.5 cm of trachea followed by reconstruction with the trough method. Histological examination revealed adenoid cystic carcinoma. Adjuvant chemotherapy was indicated with 50 mg of adriamycin postoperatively. The patient has done well for 12 years with no further treatment. The tumor was an adenoid cystic carcinoma that was slow-growing, infiltration of mucus membrane was few and growth fraction (mitotic index was less than 1%) was low. Those were considered the reason for good prognosis. (Keio J Med 44 (1): 30–32, March 1995)

Key words: adenoid cystic carcinoma, trachea, twelve year survivor

Introduction

Malignant tumors arising from trachea are not common. Although the larynx, trachea and bronchi are one continuous structure, the primary tracheal carcinoma is not the same incidence rate with that of bronchial carcinoma. Tumors of the larynx and lungs are 75 and 180 times more frequent than malignant lesions of the trachea, respectively. The relatively lower incidence of malignant tracheal tumors is thought to be based on the decreased contact of carcinogens owing to the vigorous cough reflex and the effective cleansing action of cilia, which lead to less trapping of mucus.1,2 This paper presents an example of primary tracheal adenoid cystic carcinoma with good prognosis. The literature on carcinoma of the trachea is also reviewed.

Case Report

A 46-year-old woman noted the gradual development of shortness of breath upon exertion and referred to
Fig 1  Cervical CT scan: An approximately 2-cm, broad-based, oval shaped mass projecting from left side of trachea at the level of the second tracheal ring was seen (→). Tumor extended externally to the tracheal ring.

Fig 2  Histopathologic findings: The tumor was characterized by a small glandular with cribriform pattern and had mucus-secreting cells and very little cellular atypia. Histopathology of permanent preparation was diagnosed as adenoid cystic carcinoma.

Fig 3  After the reconstruction of the trachea. Anterior cervical skin is slightly cicatrical. However the quality of life is satisfactory.

Fig 4  Fiberscopic finding of intraluminal trachea. Three fourth of trachea (nine to six o’clock) is white coloured and cicatrical. The tracheal air space is fully kept.

Trachea using anterior neck skin. The first stage, partial resection of the trachea is carried out and a trough (large tracheostoma) is performed. Four or five weeks later, tracheal reconstruction utilizing a skin tube is performed.

During the operation, hard yellowish-white color tumor projecting from inside of the tracheal wall to outside at the level of second tracheal ring (dumbbell shaped) was detected. Tumor extended externally to the tracheal rings. The tumor mainly located on the back side of trachea and recurrent laryngeal nerve was compressed laterally approximately 1 cm. The tumor mass was well capsulated and any adhesion was not detected.

The partial resection of the trachea at the level of first to sixth tracheal cartilage and total resection of dumbbell shaped tumors were carried out. Surgical stump of the trachea, hyperplastic change of mucus membrane was detected. No tumor invasion to surrounding tissues were seen macroscopically. Cricoid cartilage, anterior cervical muscles, recurrent laryngeal nerve and thyroid gland were preserved. The defect portion of trachea was rebuilt by trough method using anterior neck skin. Recurrent laryngeal nerve paralysis was not seen postoperatively.

The tumor was characterized by a small glandular with cribriform pattern and had mucus-secreting cells and very little cellular atypia. Histopathology of permanent preparation was diagnosed as adenoid cystic carcinoma (Fig 2). On this account, additional removal of left thyroid gland, anterior cervical muscles, left recurrent laryngeal nerve, three fourth of first to sixth tracheal cartilage and cricoid cartilage were carried out.
Excised nerve showed perineural invasion of the tumor. Thus 50 mg/body of adriamycin was administered post-operatively. The patient has done well for 12 years with no further treatment (Fig 3,4).

Discussion

Primary tracheal carcinoma is rare disease. Price reviewed the world literature up to 1979 and found 352 cases of tracheal malignancy. This review shows 55% squamous cell carcinoma and 31% adenoid cystic carcinoma. Other tumor include adenocarcinoma, anaplastic carcinoma, small cell carcinoma, lymphoma, chondrosarcoma, carcinosarcoma, mucoepidermoid carcinoma and carcinoid tumor. In spite of this fact, in Japan, adenoid cystic carcinoma is the most common.4,5 Typical presentation of adenoid cystic carcinoma shows exophytic growth. Compression or displacement of structures is common, but metastasis involvement of adjacent nodes is less common than in squamous cell lesions. As indicated of the present case, the propensity for this tumor to extend by perineural invasion is well known. In contrast to squamous cell carcinoma, adenoid cystic carcinoma has a tendency to a more equal sex distribution, a younger population, and frequently a non-smoking history.

The prognosis of tracheal cancers depend upon the cell type and the size of the lesion. Hadju reports the average survival time of this disease to be nine years, as compared to nine months for squamous carcinoma. Those patients with either squamous cell or mucus-secreting adenocarcinomas that are large in size have a poor prognosis.8 Furthermore extra tracheal extension correlates with poor prognosis whereas length of tumors does not. Over all five year survival is around 30%. Patients with extra tracheal extension show 18% five year survival while those with only intraluminal disease show 50% survival.7 Local recurrence is the usual cause of death. Pulmonary metastasis may occur many years after the onset of local lesion and may remain asymptomatic for many years.

The criteria for non-resectability used by Grillo include involvement of greater than 50–60% of tracheal length, extension to the carina, or radiologically determined mediastinal extension.9 The adult trachea is 11 cm long and Parrish and Jones9 state that 30–40% of the trachea can be resected with successful end to end suture. Two centimeters can be obtained with laryngeal release, by dividing the thyrohyoid muscles and membrane and freeing the superior horn of the thyroid cartilage, as described by Bryce.10 Tracheal dissection alone will give another 2 cm.11 Grillo has described extensive mobilization of right hilum of the lung allowing 6 cm of resection and has shown that because the blood supply of trachea is mainly through the inferior thyroid arteries, the upper segment of the trachea can be mobilized and re-anastomosed to a lower segment with its blood supply intact, allowing a safer reconstruction in the neck if necessary.12 Since adenoid cystic carcinoma have a predilection for submucosal spread, which makes the actual extent of the tumor.13 Incomplete resection is directly related to recurrence. The resectability rate of adenoid cystic carcinoma reported in the literature tends to be low. Thus the wide resection with adequate surgical margins should be needed. On this account, additional removal was performed.

In the present case, the tumor was an adenoid cystic carcinoma that was slow-growing, infiltration of mucous membrane was few and growth fraction (mitotic index was less than 1%) was low. Those were considered the reason for good prognosis. However the tumor extended externally to the tracheal rings and contained mucus-secreting cell. Thorough follow up including local recurrence and distant metastasis should be warranted.

References


Kohno N, et al: Primary Tracheal Adenoid Cystic Carcinoma