Case Report

A Long-Term Survivor after Aggressive Treatment for Tracheal Adenoid Cystic Carcinoma: A Case Report

Ryuichi Suemitsu, MD, PhD, Tatsuro Okamoto, MD, Riichiroh Maruyama, MD, Hiroshi Wataya, MD, Takashi Seto, MD, and Yukito Ichinose, MD

A 61-year-old male underwent a tracheal resection and reconstruction with omentopexy for the treatment of tracheal adenoid cystic carcinoma. Postoperatively, he received radiotherapy for a microscopic residual tumor of the tracheal margin. It recurred with pulmonary metastasis and para-esophageal lymph nodal metastasis at 7 years and 10 months after the initial operation. A wedge resection and concurrent chemoradiotherapy were carried out to treat the recurrence, followed by consolidation chemotherapy. Eleven months later, he developed a second recurrence with a right hilar lymph nodal metastasis, and thereafter he also suffered from a left hilar lymph nodal metastasis. As a result, he received concurrent chemoradiotherapy twice over a 3-year period. One year and 2 months later, a new pulmonary metastasis appeared, and a wedge resection was carried out. Although the patient had five instances of recurrence over an 11-year period during his treatment course, he is presently doing well as a result of appropriate local treatments using multiple modalities. (Ann Thorac Cardiovasc Surg 2007; 13: 335–337)

Key words: adenoid cystic carcinoma, salvage surgery , tracheal resection, chemoradiotherapy, long-term survivor

Introduction

Pulmonary adenoid cystic carcinoma (ACC) is a relatively rare salivary gland-type malignant neoplasm that is usually located in the central airways.[1)] It appears to be unrelated to smoking, with an equal distribution in males and females.[2)] In general, ACC grows slowly, and it is considered to be a low-grade malignancy, but it tends to metastasize to distant sites and often recurs after a long interval. We herein report a patient with pulmonary ACC who developed repeated recurrences but nevertheless had a long-term survival as a result of appropriate aggressive treatments for recurrent disease.

From Department of Thoracic Oncology, Kyushu Cancer Center, Fukuoka, Japan

Received November 2, 2006; accepted for publication July 2, 2007
Address reprint requests to Ryuichi Suemitsu, MD, PhD: Department of Thoracic Surgery, National Hospital Organization Kyushu Medical Center, 1–8–1 Jigyohama, Chuo-ku, Fukuoka 810–8563, Japan.

Case Report

A 61-year-old male experienced sudden-onset hoarseness, followed by the development of dyspnea on exertion and wheezing in May 1994. The diagnosis was bronchial asthma, and he was treated by his home doctor with high-dose inhaled corticosteroids and bronchodilators. Since the symptoms did not improve after this therapy, he consulted our institution in August 1994. A chest roentgenogram revealed almost normal findings, but a tomogram of the trachea showed a polypoid lesion in the middle trachea (Fig. 1). Fiberoptic bronchoscopy (FOB) revealed a smooth polypoid tumor that occupied approximately 80% of the tracheal lumen, and it was diagnosed to be ACC by an aspiration biopsy. He underwent a tracheal resection and reconstruction in September 1994. During the operation, a frozen section examination revealed a residual tumor remaining in the tracheal surgical margin. We therefore performed an additional resection of the positive margin and then finally resected nine tracheal rings. After reconstruction, the tracheal anastmosis was covered with omentum. Postoperative locoregional radiation around the anastmosis, with a total dose of 50 Gy,

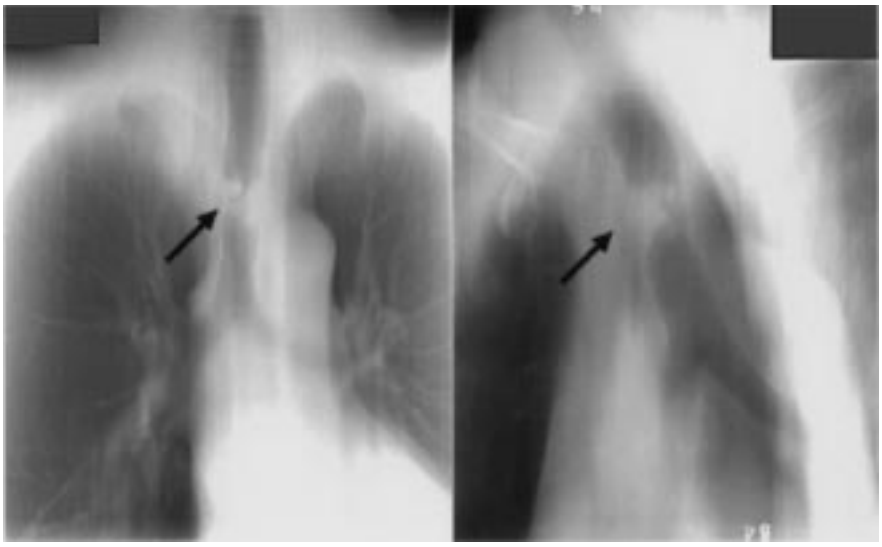

Fig. 1. Tracheal tomogram, showing a polypoid lesion in the middle trachea (arrows).

was delivered in 25 fractions from the 29th postoperative day. After he was discharged from our hospital, he was followed up regularly as an outpatient.

The patient had a relapse in his left upper lobe that was detected as a small nodule by computed tomography (CT) scans in November 2001. The nodule later become enlarged, and the para-esophageal lymph nodes showed a swelling in CT scans in June 2002. An FOB revealed a smooth polypoid tumor pressured on the tracheal wall (Fig. 2A). He underwent a wedge resection in his left upper lobe, but the lymph node metastases were not considered to be resectable during the operation. As a result, he received concurrent chemoradiotherapy (a total of 60 Gy, 30 fractions) for the lymph node metastases, using uracil-tegafur (UFT), 400 mg/m^2 for days 1–14 and 29–42, and cisplatin (CDDP), 80 mg/m^2 for days 8 and 36. Thereafter he received three cycles of monotherapy with docetaxel as consolidation chemotherapy. He demonstrated an objective response from the chemoradiotherapy. After the treatments, the polypoid lesion improved on FOB findings (Fig. 2B).

He had a second relapse as an extra-nodal invasion of the right hilar lymph node metastasis in June 2003. He received the same regimen of concurrent chemoradiotherapy with UFT and CDDP for the relapse site and also demonstrated tumor shrinkage. A third relapse occurred in his left hilar lymph node with extra-nodal invasion of the left main bronchus in November 2004. He again received concurrent chemoradiotherapy with the UFT and CDDP for the left hilar lymph nodes. Thereafter a small pulmonary metastasis also appeared in his left upper lobe, which showed progression. As a result, a wedge resection of the left upper lobe was carried out. He is alive without any further recurrence, and the total survival time is presently 11 years from the first medical assessment.

Discussion

Primary tracheal tumors are rare diseases with an incidence of less than 0.1% of non-small-cell lung cancer (NSCLC). ACC is one of the most common tracheal tumors among them.[3] The prevailing treatment of pulmonary ACC is surgical resection, and, in general, the survival of patients who underwent a complete resection of the tumor has been reported to be long because these tumors tend to grow very slowly. On the other hand, they frequently recur in spite of a complete resection. Therefore the patients need careful and long-term observations to detect a relapse. Prommegger and Salzer reported 16 cases of pulmonary ACC, and pulmonary metastasis was found to occur more frequently (31%) than local recurrence (18%) after the primary operation; pulmonary metastases and local recurrence were found approximately 6.8 and 12 years after the primary operation, respectively.[4] In our case, the patient first developed pulmonary metastasis accompanied by lymph node metastases at 7 years after the primary operation and thereafter suffered from repetitive relapses over the 4-year period following the first recurrence.

ACC has been reported to have sensitivity to radia-

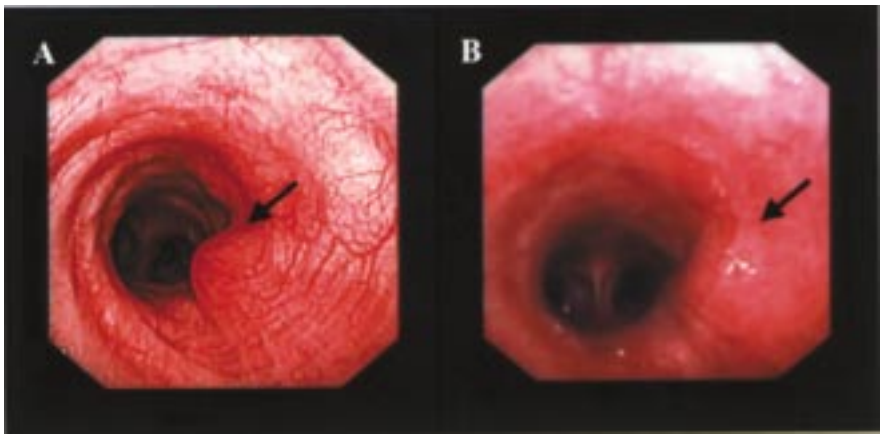

Fig. 2. Fiberoptic bronchoscopic findings, showing tracheal cavity (arrows).
A: Pretreatment of chemoradiotherapy.
B: Posttreatment of chemoradiotherapy.

tion[2]; however, the role of chemotherapy in ACC patients remains controversial, and no effective chemotherapy regimen for pulmonary ACC has yet been established. Recently reported results of randomized trials in NSCLC patients have shown concurrent chemoradiotherapy to be superior to radiotherapy alone, and it is also the standard treatment for NSCLC patients with unresectable stage III disease. The chemotherapy used with this combined treatment was used not only to enhance radiosensitivity, but also to eradicate any occult distant metastases. In our case, we carried out radiotherapy combined with chemotherapy using UFT and CDDP in expectation of enhancing the power of local control. We previously reported the treatment with UFT/CDDP/radiation to be effective for NSCLC patients, with a response rate of 81% and a median survival time of 16.5 months.[5] The present patient demonstrated unresectable recurrences three times and thus received UFT/CDDP/Radiation treatment each time. After receiving this chemoradiotherapy regimen, he has developed no locoregional recurrence, and therefore this regimen may thus be a promising treatment modality for ACC.

The prognosis after surgery for pulmonary metastases of ACC has been reported to be poor, and radiation treatment for metastases tends in some cases to be a more preferable treatment alternative for pulmonary metastasis than an operation.[4] We selected both radiotherapy and surgery based on the patient's condition to control the metastases. If radiotherapy had been selected for the pulmonary metastasis in the present case, the total radiation field would have been too large to avoid severe lung toxicity, though it may be possible to select a stereotactic radiosurgery for small areas of pulmonary metastases.

In conclusion, we herein reported the case of a long-term survivor who developed repeated recurrences that were controlled by chemoradiotherapy and a salvage surgery each time. This case suggests that aggressive local treatments are therefore indicated for patients with metastatic ACC to obtain a long-term survival when the patient is deemed able to tolerate such treatments.

References

1. Moran CA, Suster S, Koss MN. Primary adenoid cystic carcinoma of the lung. A clinicopathologic and immunohistochemical study of 16 cases. *Cancer* 1994; **73**: 1390–77.
2. Grillo HC, Mathisen DJ. Primary tracheal tumors: treatment and results. *Ann Thorac Surg* 1990; **49**: 69–77.
3. Pearson FG, Todd TR, Cooper JD. Experience with primary neoplasms of the trachea and carina. *J Thorac Cardiovasc Surg* 1984; **88**: 511–8.
4. Prommegger R, Salzer GM. Long-term results of surgery for adenoid cystic carcinoma of the trachea and bronchi. *Eur J Surg Oncol* 1998; **24**: 440–4.
5. Ichinose Y, Nakai Y, Kudoh S, et al. Uracil/tegafur plus cisplatin with concurrent radiotherapy for locally advanced non-small-cell lung cancer: a multi-institutional phase II trial. *Clin Cancer Res* 2004; **10**: 4369–73.