

Case Report

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Extensive Tracheal Adenoid Cystic Carcinoma – Case Report

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Abstract

Primary tracheal cancer is considered to be relatively rare. Adenoid Cystic Carcinoma (ACC) is a low-grade malignancy which is the second most common tracheal malignancy after squamous cell carcinoma. It has been proved that the majority of primary tracheal tumors, which were locally advanced and unresectable at the time of diagnosis, were squamous cell carcinoma.

We are presenting a case of ACC that encase whole trachea to the main right bronchus at the presentation found in a woman just after she gave birth. Tumor was treated with conformal radiation therapy and showed good regression. Three years after the diagnosis, the patient is alive with stable disease.

Keywords: Adenoid cystic carcinoma; Trachea; Primary tracheal cancer

Introduction

Primary tracheal cancer is considered to be relatively rare [1]. Adenoid Cystic Carcinoma (ACC) is a low-grade malignancy that is the second most common tracheal malignancy after squamous cell carcinoma [2]. ACC has a striking tendency toward submucosal extension, manifesting as a sessile, polypoid, annular, or diffusely infiltrative nodule with a heaped-up margin. Pathologically, the tumor narrows the circumference of the main airway, spreads longitudinally from the main mass, and penetrates through the bronchial wall to invade the surrounding tissues [3]. Longitudinal thickening of the tracheal wall extending superiorly and inferiorly from the main mass is a typical manifestation on MSCT or MR images. These tumors are potentially resectable with 5-year survival rates of 66% to 73% and 10-year survival rates of 53% to 57% even for patients with microscopically positive resection margins [4]. Local recurrence is common. Besides the surgical resection, radiation therapy could be applied. Although ACC of trachea is not highly aggressive malignant tumor more than 50% of patients with tracheal ACC have hematogenous metastases. Pulmonary metastases are the most common and can remain asymptomatic for many years [5]. The best therapy option is surgical resection if possible, with adjuvant chemotherapy, even in patients with pulmonary metastases [6].

Case

A 35-year-old woman presented with progressive cough and dyspnea that lasted 2 months before paying a visit to a physician. No other but respiratory symptoms were present. The onset of symptoms was just after she gave birth to her first child. Routine laboratory tests were normal. Spirometry showed restriction of ventilation of a low degree and obstruction of ventilation of a middle degree. Abnormally narrow trachea was noted on chest radiograph. Narrowed trachea was observed on bronchoscopy (Figure 1), as well as on MSCT and MR images (Figure 1) with almost uniformly thickened wall in the whole length of the trachea without a detectable main mass. Luminal width was 5 to 10 mm. Slight homogenous contrast opacification of the tracheal wall was noted on contrast enhanced MSCT images. Tumor extended from cervical trachea to carina including membranous part of trachea with slight thickening of the right main bronchus on the origin. Membranous part was involved in a lesser degree. Calcifications were not detected. The tumor showed isointens signal on T1 weighted MR images and higher signal intensity on T2 weighted images with a slight

contrast opacification. Thick tracheal wall, up to 17 mm, was visible with Ultrasound (US) in jugular cavity. During the US examination, FNAB of the proximal trachea was performed. The results did not disclose diagnosis. Endotracheal, bronchoscopy guided, FNAB was performed. The diagnosis of ACC was established based on cytological finding. Neither local nor distant tumor spread was detected at the moment of diagnosis.

Conformal radiation therapy with 3 conformal photon beams in energy of 18 and 5 MV was applied in 33 fractions to a final dose of 59.4 Gy. No chemotherapy was applied.

After the treatment the first PET/CT scan was done and the highest SUV in thickened tracheal wall was 2.2. Control bronchoscopy (Figure 1), MSCT and MR (Figure 2) were performed and showed good tumor regression with maximal tumor thickness of 10 mm. The tumor regression was proportional in the whole length. Two years follow up MR showed further tumor regression with maximal thickness of 4 mm. Almost unchanged, the highest SUV of 2.4 was noted on PET/CT scan. No local invasion neither distant metastases were noted. After 3 years patient is alive with stable disease according to the chest MR (Figure 2).

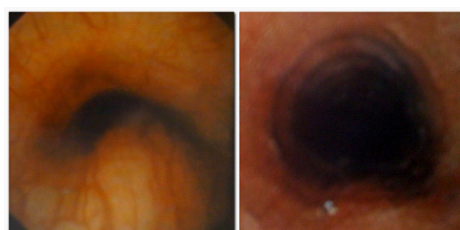


Figure 1: Bronchoscopy before and after treatment.

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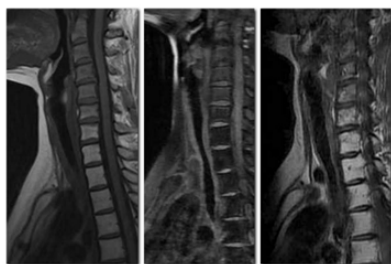


Figure 2: MRI images before and after treatment.

Discussion

Diffuse tracheal narrowing is extremely rare condition. It is almost exclusively nonneoplastic in origin. Searching the literature, authors found no reports of diffuse tracheal involvement by ACC on presentation. Some studies have showed that the majority of primary tracheal tumors, which were locally advanced and unresectable at the time of diagnosis, were squamous cell carcinoma and managed by nonsurgical procedures [7].

Thus, due to tumor extent, primary differential diagnoses were nonmalignant (relapsing polychondritis, Wegener granulomatosis, amyloidosis or inflammatory bowel disease, rhinoscleritis). Some of

the mentioned diseases could show nodularity of the thickened tracheal wall, but it was not obvious in this case.

This case shows that even a diffuse tracheal disorder could be of malignant origin.

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