Adenoid Cystic Carcinoma

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Abstract

Adenoid cystic carcinoma (cylindroma) is a rare neoplasm which usually arises from the salivary, lacrimal or other exocrine glands. This report is based on a retrospective analysis of 4 patients with adenoid cystic carcinoma seen between 1995 and 2000 in our center.

Turkish Respiratory Journal, 2003;4:(2):85-87

Keywords: *adenoid cystic carcinoma, cylindroma, exocrine glands*

Introduction

Adenoid cystic carcinoma (cylindroma) is a rare neoplasm which usually arises from the salivary, lacrimal or other exocrine glands. It may also arise from the lung, breast, vulva, eosofagus, cervix or the external auditory canal tissues (1,2).

Second to squamous cell carcinoma, adenoid cystic carcinoma is the most frequent type of neoplasm encountered in primary tracheal tumors. Primary tracheal tumors are reported to have an incidence of 0.2 persons per year (3).

We report four cases of patients with adenoid cystic carcinoma seen between 1995 and 2000 in our center.

Case 1

The patient was a 52 year old male who presented with coughing, dyspnea and bloody sputum complaints. He had worked in a coal mine for 20 years. He had a smoking history of 120 pack/years, and started coughing 7-8 months ago. Dyspnea initially occurred with effort. He only had noticed minimal blood in his sputum 4-5 times. The patient was using a bronchodilator at present. His blood pressure and heart rates were normal. An increased expirium/inspirium ratio was the only noteworthy finding of the physical examination. Bilateral hyperlucency was the only abnormality in his chest X- ray. A CT scan revealed a small mass with irregular contours at the right hilar region around the right main bronchus.

Routine blood laboratory results were normal. Fiberoptic bronchoscopy was performed under local anesthesia. A tumor in the trachea infiltrating the distal portion of the trachea and obstructing 75% of the left main bronchus was detected. Pathological

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diagnosis of the punch biopsy specimens were reported as adenoid cystic carcinoma. Palliative radiotherapy was planned.

Case 2

The patient was a 41 year old male carpenter working in furniture construction. He presented with complaints of dyspnea, malaise, fever, weight loss and coughing. His complaints had begun as a nonproductive cough 2-3 months ago. He had a smoking history of 18 pack/years. His blood pressure and heart rates were normal. Bronchial pulmonary sounds were present at auscultation. Physical examination was otherwise normal. His routine blood laboratory results also were within normal ranges. A fiberoptic bronchoscopy was performed under local anesthesia and a tracheal tumor was detected obstructing the lumen by 70%. Pathological diagnosis of the biopsy specimens reported adenoid cystic carcinoma. Palliative radiotherapy was started. A 30% regression in the stenosis was observed in the fiberoptic bronchoscopy repeated following this treatment. The patient's complaints also disappeared. However the patient, eighteen months later was readmitted to hospital with stridor. At his time, the fiberoptic bronchoscopy revealed that the stenosis had progressed to 80%. Radiotherapy was repeated. After this second radiotherapy period the patient was clinically stable for a year. The CT scan at this time revealed paramediastinal fibrotic lesions which were interpreted as the sequelae of the radiotherapy. Fiberoptic bronchoscopy was performed. The tumour was observed to obstruct 50% of the lumen at a location 5 mm below the vocal cords. A surgical intervention was considered and the patient was referred to a center where an ear, nose and throat surgery team also was available.

Case 3

The patient was a 49 year old housewife who presented with complaints of coughing, chest pains on the right, weight loss and dyspnea which had started and progressed in the past two months. No noteworthy findings were present at the physical examination, and routine blood laboratory investigations. A rigid bronchoscopy was performed under general anesthesia. The intermediate bronchus lumen was observed to be almost completely obstructed. Histological examination results of the biopsy specimens were reported as adenoid cystic carcinoma. The lesion was considered operable. A right pneumonectomy was performed followed by radiotherapy.

Case 4

The patient was a 22 year old young woman whose complaints of dyspnea had started a week ago. Her blood pressure and heart rates were normal. Pulmonary sounds were decreased in the left hemithorax and dullness was found at percussion. A rigid bronchoscopy was performed under general anesthesia and a tumor completely obstructing the left main bronchus at a site 3 cm from the main carina was detected. Pathology of the biopsy specimens reported adenoid cystic carcinoma. On the CT scan, a consolidation causing a volume loss in the left lung, and a compensatory hypertrophy in the right lung was seen. She was admitted to our thoracic surgery department and pneumonectomy was performed.

Discussion

Adenoid cystic carcinoma of the lungs are rare and locally infiltrative tumours. They usually present a slow clinical progress. After squamous cell carcinomas they are the second most common neoplasms of the trachea. The majority (80%) of these tumours arise either from the trachea or the main bronchi and 10-15% from the periphery of the lung (4). The tumors originating from the trachea arise from the lower or upper third section of the trachea, from the lateral or posterolateral walls. A recent review revealed 174 patients with tracheal adenoid cystic carcinoma reported since 1998 (3). The age range was 15 to 80 years and there was no sex predominance. Accordingly in our cases, three of the 4 patients were also in this reported age range and no sex predominance was present.

The etiology of these tumours is unknown and there is no evidence based data regarding the role of cigarette smoking.

The generally recognized characteristics of adenoid cystic carcinoma consist of their locally infiltrative nature, a tendency toward local recurrence and usually a prolonged natural clinical course. Histologically they can be cribriform (cylindromatous), tubular or solid tumours, the most frequent and predominant being in cribriform pattern. This type is characterized by nests of tumor cells containing numerous sharply outlined luminal spaces and sometimes containing mucinous secretions within their lumens (5). The solid pattern has a tendency to grow predominantly extraluminally and to be more likely to metastasize.

The chest roentgenogram is often normal or shows only minimal abnormalities. In our cases, only one had an abnormal chest roentgenogram (homogeneous opacity in the left upper hemithorax).

The most frequent complaints are wheezing or stidor, dyspnea, hemoptysis and coughing. Most of our patients had been on bronchodilator therapy prior to the diagnosis.

The clinical course is often prolonged. Because local recurrence and pulmonary metastases are frequent even after curative resection, surgery alone may not be adequate and adjuvant radiotherapy is recommended (1,6). Since this report is based on retrospective data covering 5 years, the four year clinical course of only one patient is known to date.

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