

# Adenoid Cystic Carcinoma Of The Trachea Treated As COPD

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## Abstract

Primary adenoid cystic carcinoma of the trachea is a rare malignant neoplasm. We present the case of a 64 year old man, ex-smoker, who presented at the University Hospital of Larissa complaining of dyspnea on exertion, chest tightness and wheezing for more than one year. By that time the patient had been misdiagnosed and treated as chronic obstructive respiratory disease (COPD) without any improvement of his symptoms. Computed tomography of the chest revealed a tumor which caused a 90% obstruction of the trachea lumen. Bronchoscopy was performed and the pathological diagnosis was adenoid cystic carcinoma of the trachea. The literature is reviewed.

## CASE HISTORY

A 64 year-old male, farmer, ex-smoker for 4 years with a smoking history of 40 pack-years, presented at the University Hospital of Larissa complaining of dyspnea on exertion, chest tightness and wheezing. The patient had a long-standing history of his symptoms, lasting for over one year. He had been previously diagnosed with chronic obstructive pulmonary disease (COPD) and received treatment with inhaled bronchodilators with no significant improvement of his symptoms. He presented a weight loss of about 8 kg in the past few months but he denied any other symptoms, including fever, cough, chest pain, increased sputum production or hemoptysis.

On physical examination he was a pleasant apparently healthy man with body temperature 36,8°C, pulse rate 85 beats/min, blood pressure 130/80 mmHg, respiratory rate 21 breaths/min and oxygen saturation 97% on room air. Auscultation disclosed stridor and expiratory wheezing over both hemithoraces. No other abnormal findings were found in the rest of the physical examination. In the laboratory tests, the patient's hemoglobin was 11.6 g/dL %, white blood cell count was 6,800/ $\mu$ L (with 60% neutrophils and 36% lymphocytes), and platelet count was 282,000/ $\mu$ L, while the values for urea nitrogen, creatinine and electrolytes were within the normal range.

The patient's chest x-ray on admission (Figure 1), shows a

well-defined homogenous opacity, located in the distal trachea, causing a significant stenosis of the tracheal lumen, while both lung fields are clear.

## Figure 1

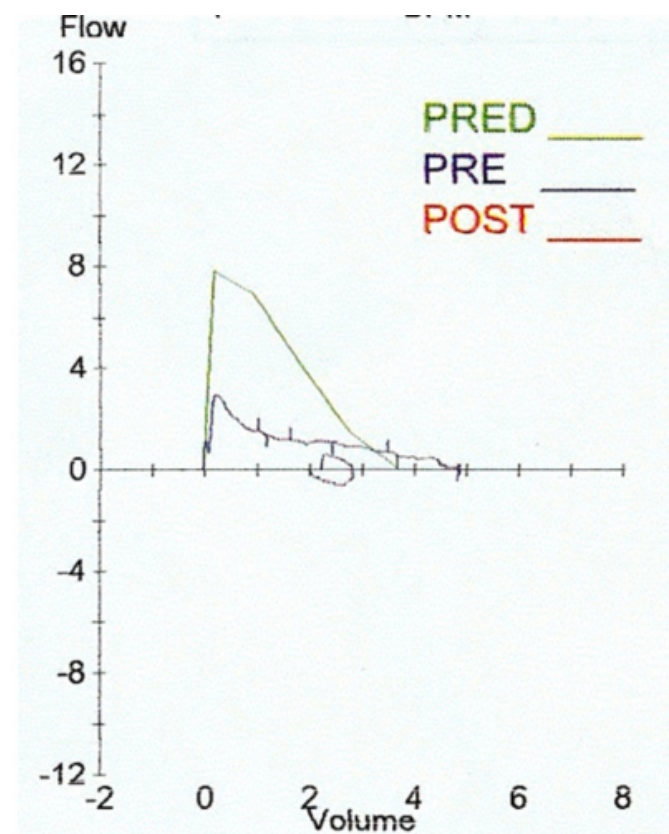
Figure 1: Chest x- ray on admission.



Simple spirometry (Figure 2), demonstrated an FEV<sub>1</sub> of 1,69L (58% predicted), an FVC of 4,90L (132% predicted), and FEV<sub>1</sub>/FVC 35%. An expiratory flow-volume loop was performed at the same time and it is also shown.

**Figure 2**

Figure 2: The patient's expiratory flow volume loop.

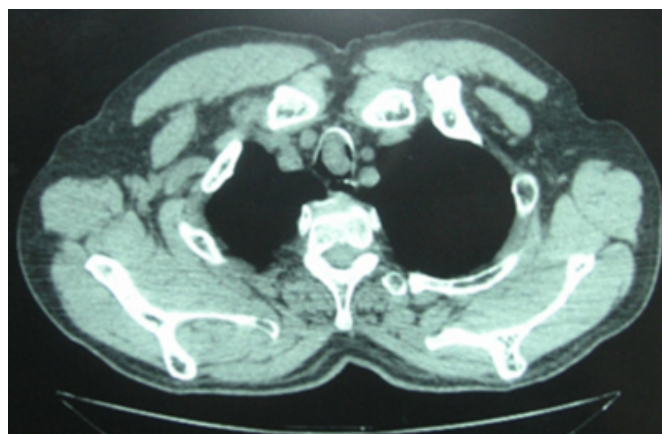


The spirometric values were suggestive of a severe obstructive pattern; however, the expiratory limb of the flow-volume curve was suggestive of upper airway obstruction. At the time of the initial interpretation of the patient no inspiratory limb of the flow-volume curve had been performed to the patient. A subsequent flow-volume curve was typical of fixed upper airway obstruction (not presented).

Taking into account the finding in the chest x-ray, the patient underwent a computed tomography of the chest, performed after the intravenous injection of contrast material, which revealed a soft tissue mass occluding 90% of the trachea (Figure 3).

**Figure 3**

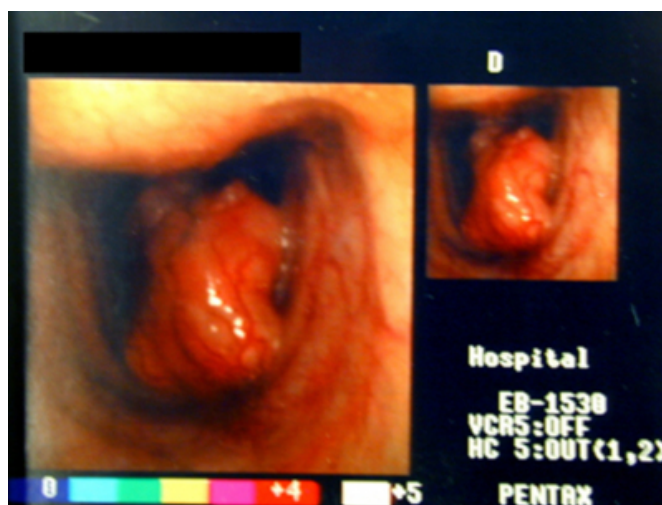
Figure 3: CT scan of the chest showing a soft tissue mass occluding 90% of the trachea.



Bronchoscopy demonstrated an exophytic mass just under the vocal chords which occluded approximately 70% of the tracheal lumen (Figure 4). The histopathologic examination of an endobronchial biopsy revealed an adenoid cystic carcinoma.

**Figure 4**

Figure 4: Endoscopic view of the smooth, shiny, exophytic mass in the tracheal lumen



The patient was advised to undergo surgical resection of the mass with adjuvant radiation therapy. As far as we know he has not been operated yet.

## DISCUSSION

Primary adenoid cystic carcinoma (ACC) of the lung is a very rare malignant neoplasm and accounts for one per thousand of all respiratory tract cancers. (1) Usually it arises in the lower trachea, main stem or lobar bronchi. (2) Peripheral ACC of the lung is uncommon and represents

approximately 10% of the pulmonary ACC. (3) Most of the times, ACC appears as an endobronchial exophytic lesion that is located submucosally and usually is covered by intact mucosa. Macroscopically ACC is a shiny, smooth, sessile polypoid mass, as was the case of our patient.

Presenting symptoms are usually cough, dyspnea, hemoptysis, hoarseness, evidence of airway obstruction or post-obstructive pneumonia. The symptoms depend on the tumor's size and location. Tumors with important occlusion of the trachea usually cause stridor and wheezing. Such findings in a current or ex-smoker along with an obstructive pattern in spirometry are the reason why these patients are often misdiagnosed and treated as COPD, and this was the case of our patient. However, the tumor can occlude 75% of the trachea before causing symptoms.

Chest X-ray is not sensitive enough for the diagnosis of endobronchial ACC, however it can show the ACC associated atelectasis and post obstructive pneumonia. (3) Most commonly the endobronchial ACC is identified by Computed Tomography (CT) of the chest and, since most endobronchial ACC are endoscopically visible, the tumor can be detected by fiberoptic bronchoscopy. (3) Computed tomography shows the tumor which is a homogenous mass encircling the posterolateral trachea with thickening of the tracheal wall and usually has a broad base. (4)

The pathologic diagnosis can be established by tissue biopsies or fine needle aspiration (FNA) under bronchoscopy. Because of their submucosal location and the fact that they usually are covered by intact mucosa, the diagnosis cannot be established by techniques such as bronchial brushing or washing cytology. Usually endobronchial biopsy or fine needle aspiration cytology is required.

Primary ACC of the respiratory tract usually grows slowly and is thought to arise from ductal/myoepithelial cells of the submucosal glands. Its usual cytologic appearance consists of small neoplastic basaloid cells with high nuclear/cytoplasmic ratios which arrange in clusters and often are associated with presence of cyanophilic hyaline basement membrane material. (3) Pathologic examination usually reveals local invasion beyond the wall of the trachea in all patients. Microscopic examination sometimes shows extension in the submucosal and perineural lymphatics well beyond the grossly visible or palpable limits of the tumor. (5)

The primary ACC of the lung is a low grade malignancy and

has very good prognosis. Although it is important to pay attention to the local extensiveness of neoplasm, (6) ACC grows slowly and is metastasizing late. Metastases are usually hematogenous. Local recurrence after resection can also occur. (7) Pulmonary metastases have also been reported. Lymphatic metastases are uncommon. (8) The therapeutic procedure comprises surgical treatment if possible. If the ACC arises in the trachea, surgical resection and anastomosis has been reported. For unresectable tumors, radiation therapy and chemotherapy could be effective. Laser resections have also been reported. (9) After complete resection, the five year survival rate is approximately 80 percent. Aggressive surgical resection is important for the improvement of survival in this condition, although this tumor shows low grade malignancy and grows slowly. (11)

In conclusion, primary tumors of the major airways may present with symptoms suggesting obstructive disorders and may cause an obstructive spirometric pattern. Simple diagnostic procedures, such as the chest x-ray and the flow-volume loop in simple spirometry may be suggestive of the diagnosis as in the case of our patient.

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