Adenoid cystic carcinoma of the trachea mimicking asthma

Tracheal malignant neoplasms are rare. Their incidence is 140 times less than that of lung cancer.1 Tracheal tumors can be silent or deceptive in their clinical manifestations which can simulate many common ailments of the respiratory tract. The most common presenting symptoms of tracheal carcinomas include cough, hemoptysis, hoarseness and dyspnea.2 Dyspnea can be associated with wheezing, and several patients were treated for bronchial asthma for long periods of time before the discovery and diagnosis of a primary tracheal tumor.3,4 We report a patient with adenoid cystic carcinoma of the trachea who had recurrent attacks of wheezing and was misdiagnosed and treated as a case of bronchial asthma for over 10 years.

A 50 year old female was brought to the Emergency Room with history of dyspnea and cough of 2 days duration. On physical examination, the patient was anxious, dyspnic, mildly cyanotic and sweating. Pulse was 90 per minute, blood pressure 190/90, and respiratory rate 28 per minute. The trachea was central. Chest auscultation revealed decreased breath sounds, diffuse inspiratory and expiratory wheezes with prolonged expiratory phase. Heart sounds were normal. Chest x-ray showed slight cardiac enlargement. Electrocardiogram revealed low voltage and P pulmonale with Q waves in leads V1, V2 and V3.

The patient stated that she had repeated attacks of dyspnea, wheezing and dry cough for the last 11 years without any prolonged remissions or significant improvement in her condition. The onset of her illness started with a chest infection accompanied by severe wheezing, following delivery of one of her children. Since then she consulted many physicians who prescribed her numerous anti-asthmatic medications, including aerosols, and hydrocortisone, without any remarkable alleviation of her symptoms. Her attacks were triggered or aggravated by fumes, heat or physical exertion. There was no history of similar attacks during her childhood. Her father was asthmatic and died 15 years earlier and her mother was hypertensive and died of cerebrovascular accident.

The patient was admitted to the hospital for evaluation. Her work-up included blood gases analysis which revealed PH: 7.36, PO2: 77, CO2: 56, O2 Saturation: 95%, Base excess: +4. Spirometry showed the following: Forced vital capacity (FVC): 1.85 liters (predicted 3.55), forced expiratory volume in one second (FEV1): 0.85 liters (predicted 2.80), FEV1%: 47.5% (predicted 79%). These findings were consistent with severe obstructive ventilatory defect.

Otolaryngological examination revealed no lesions involving the vocal cords or the supraglottic region. A soft tissue tomogram of the neck showed 2 nodules below the glottis within the upper part of the trachea. On bronchoscopy, a smooth shiny nodule, 5 mm in diameter was seen arising from the left side of the trachea, 5 cm distal to the vocal cord. The nodule was partially obstructing the lumen of the trachea, and was followed by a similar nodule on the right side. Between the 2 nodules, the lumen of the trachea was almost totally obliterated. A biopsy was taken, and the tumor proved to be adenoid cystic carcinoma.

The segment of the trachea harboring the tumor was resected. Pathological examination showed a segment of trachea 3.8 cm in length by 2 cm in diameter. Two cartilagenous rings were identified. The outer surface exhibited a firm area measuring 1 x 0.4 cm laterally and posteriorly. On opening the trachea, 5 fleshy tan light red nodules ranging from 0.4 - 1.0 cm in diameter were seen (Figure 1). Two large nodules were present proximally, and it seems that these nodules were seen on bronchoscopy. Three smaller nodules next to each other were present in the

Figure 1 - Resected segment of trachea. The lumen is opened revealing several fleshy tumor nodules

Figure 2 - Photomicrograph of the tumor. Classical cylindromatous microcystic pattern of adenoid cystic carcinoma
distal part of the resected segment, and it appears that these were not visualized on bronchoscopy because they were masked by the proximal nodules. Histologically, all the nodules were identical. The tumor is composed of small basal type cells with scant cytoplasm and dark hyperchromatic nuclei and are arranged in the typical cribriform or cylindromatous pattern (Figure 2), which displayed numerous microcystic spaces which contained connective tissue mucin. The tumor cells were also arranged in cords. Tumor nests were seen surrounding tracheal cartilage, and infiltrating the entire thickness of the tracheal wall. The margins of resection were free of tumor.

Soon after the operation, the patient developed an Addisonian crisis due to cessation of her antiasthmatic medications before the operation which included hydrocortisone. She was given 600 mg hydrocortisone intravenously followed by 200 mg every 4 hours until she improved and recovered. On the 6th post operative day, she developed edema of both lower limbs, abdominal wall and sacral area. It was suspected that she had developed thrombosis of the inferior vena cava, and was placed on heparin. The edema and swelling subsided gradually.

Six weeks following the operation, bronchoscopy was carried out. It revealed narrowing of the lumen due to the presence of granulation tissue and silk sutures. Gradual dilation was performed and granulation tissue with debris and suture material were removed. An adult bronchoscope could be passed easily. During the following 6 months, the patient stopped wheezing and her breathing improved markedly.

Primary tracheal tumors are rare, with a reported frequency of 0.19% in patients with respiratory tract malignancy. One third to over half of these tumors are adenoid cystic carcinoma. In addition to its slow progression, this tumor has a tendency for local spread, perineural invasion, and post-operative recurrences.

Before 1960, most patients with tracheal neoplasms were treated by endoscopic removal of the tumor or had a biopsy followed by external orthovoltage irradiation or radon seed implantation. Advances in surgical techniques in the last 3 decades in the field of tracheal surgery, including resection and reconstruction without graft or prosthesis, made it possible to resect tracheal tumors that might previously have been considered unresectable.

Most of the symptoms of tracheal tumors are related to obstruction of the air passage leading to inspiratory and expiratory stridor which can simulate bronchial asthma as in this case. In spite of the well known long held dictum that all that wheezes is not asthma, this clinical fact is often forgotten. Many patients with upper airway obstruction are often treated for years for asthma, chronic bronchitis, emphysema or recurrent pneumonia. The causes of upper airway obstruction are numerous. They include vocal cord lesions, laryngeal edema, tumors both extrinsic and intrinsic, goiters, foreign bodies, mediastinal masses and several other conditions.

In patients with upper airway obstruction due to fixed obstructing lesion such as tracheal stenosis or tracheal tumor, as in the present case, pulmonary function testing with flow-volume loops is the mainstay of early diagnosis. In such cases, flow-volume loops reveal a limitation of flow with a plateau effect on both inspiration and expiration.

Due to the rarity of the tracheal malignant neoplasms, they are seldom considered in the differential diagnosis at onset of wheezing in an adult. The present case illustrates that an adult patient who had progressive stridor and dyspnea, without any significant response to conventional and acceptable modalities of treatment should have a radiological and endoscopic evaluation of the entire respiratory passages.

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