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CLINICAL REPORT

Adenoid Cystic Carcinoma of Trachea: A Diagnostic and Therapeutic Challenge

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Abstract Primary tumors of the trachea, which may be benign or malignant, account for fewer than 0.1 % of tumors. Adenoid cystic carcinoma is the second most common tracheal tumor. It poses a diagnostic and therapeutic challenge because of its slow growth and tendency for local recurrence and late metastasis. We present a case of adenoid cystic carcinoma of trachea which was misdiagnosed and mistreated as asthma initially because of its inherent indolent progression. It is important to be acquainted with this condition to prevent delay in diagnosis and provide timely treatment.

Keywords Adenoid cystic carcinoma · Tracheal malignancy · Airway · Stridor · Asthma

Introduction

Adenoid cystic carcinoma (ACC) is a malignant tumor of salivary glands with tendency of neural invasion. It forms 10 % of the overall head and neck tumors and if present in the airway, it arises from the submucosal glands of the tracheobronchial tree [1]. Tracheal tumors are infrequent cause of dyspnoea because of their relatively rare occurrence [2]. We present a case where ACC of trachea was missed on the pretext of symptoms that impersonated a commoner ailment, asthma and was treated with partial tracheal resection and stenting followed by adjuvant postoperative radiotherapy.

Case Report

A 45 years old male presented to ENT emergency with noisy breathing for 2 days. History revealed difficulty in breathing for past 1 year. The physicians had diagnosed this to be a case of asthma for which he had been put on bronchodilators, but with little relief. There was no history of any change in character of voice, difficulty in swallowing, blood in sputum or swelling in neck. Indirect laryngoscopic examination revealed a hazy mass in subglottic region with mobile vocal cords. There was no significant lymphadenopathy. An urgent soft tissue X-ray of the neck showed a radio-opaque shadow in upper tracheal lumen (Fig. 1a). An emergency low tracheostomy was done bypassing the growth which immediately relieved stridor. A biopsy was taken retrogradely through the tracheostome which disclosed ACC. Computed tomographic scan revealed a soft tissue growth in tracheal lumen causing airway attenuation at the level of D1 (Fig. 1b).

Peroperatively, a 3 × 2 cm tumor was seen attached to anterior wall of trachea at the level of 3rd and 4th tracheal rings. Posterior and lateral tracheal walls were free of tumor. The tumor and previous tracheostome was excised taking 1.5 cm visible margins. Thyroidectomy was done along with preservation of bilateral superior and recurrent laryngeal nerves. A neotracheostome was created and the tracheal defect was reconstructed using strap muscles after stenting. Stent was removed after 3 weeks.

Histopathologic specimen showed all the margins to be free from tumor. Patient received 36 cycles of radiotherapy and is doing fine after 1 year of surgery.

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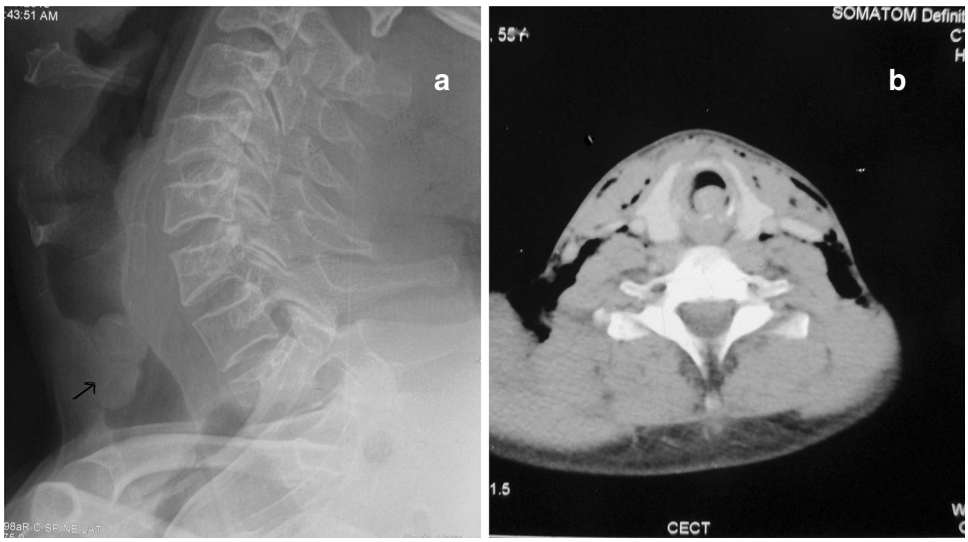

Fig. 1 **a** X-ray soft tissue neck showing mass attached to anterior wall (marked with *black arrow*) in the cervical trachea just below the cricoid cartilage. **b** CT scan of neck revealed a mass attached to the anterior tracheal wall just below the cricoids cartilage, involving almost the entire tracheal lumen

Discussion

ACC was earlier named as 'cylindroma' by Billroth in 1856. It occurs most commonly in salivary glands and with less frequency in other places like uterine cervix, breast, skin, aerodigestive tract and lungs [3]. It is a slow-growing tumor with tendency of local recurrence and late metastasis typically over a long period, thereby offering a poor prognosis. Almost the entire clinico-pathological course of ACC has been deduced by tracing salivary glands because of rarity of this type of tumor elsewhere. The incidence of primary tracheal tumors is <0.2 per 100,000 people per year with a prevalence of 1 per 15,000 autopsies. The most common tracheal tumor is squamous cell carcinoma followed by ACC, which together represent two-thirds of the tumors in airway [4]. Respiratory tumors arising from glandular tissue in trachea constitute 10 % of all the tracheobronchial tumors.

ACC may present with symptoms of upper airway obstruction (dyspnoea, stridor) and can be mistaken as asthma, bronchitis and chronic obstructive pulmonary disease. Diagnosis often gets delayed because of large functional reserve of tracheal lumen which does not cause any apparent symptom till occurrence of 50–75 % of luminal occlusion [5]. Other presentation can be cough or hemoptysis because of mucosal irritation and ulceration or voice change or dysphagia as a result of direct invasion of structures in the vicinity. Because of its indolent growth, patients with ACC of trachea can have symptoms of obstruction for years without compromising the airways absolutely, thereby getting diagnosed only in the late stage. Chest radiographs are rarely diagnostic but often done as the first investigation. Most of the times, tracheal tumors get easily overlooked on X-rays, hence CT is the standard technique to detect the tumor and its extent. MRI has special significance in ACC cases where it may help delineate neural and other soft tissue invasion.

Lawrence and Mazur examined eight cases of ACC from different anatomic sites and found all of them sharing similar ultrastructural and light microscopic pictures. Each of them showed a fine set of microstructural features like pseudocysts, intercellular spaces, basal lamina and true glandular lumens which are specific for ACC, when present together [6]. Wegner revealed structural differentials in the central and peripheral ACC tissue. The cells in the peripheral part showed lower degree of maturation than the central ones with fewer collagen fibres in peripheral stroma and proeoglycans and glycosaminoglycans dominating the periphery in the absence of histoformative features [7].

The treatment choice of ACC is complete surgical resection of the tumor. A high chance of tumor positivity of margins despite taking adequate margin of palpable or macroscopically involved tumor tissue is an intimidating characteristic of this tumor arising in trachea. In these cases, post-operative radiotherapy becomes indispensable [8]. Surgery is absolutely contra-indicated in the presence of multiple positive lymph nodes, involvement of more than 50 % of trachea, mediastinal invasion of unresectable organs, a mediastinum that has received a maximum radiation dose of more than 60 Gy and distant metastasis [5].

ACC of the airways is a difficult disease to study because of paucity of the condition, thereby excluding any potential prospective study of its natural history. However, studies so far suggest similar behaviour of ACC in trachea to other head and neck subsites [1, 3, 4].

Conclusion

ACC of trachea is an uncommon entity, though not rare. It can be easily confused with asthma because of similar clinical presentation. Therefore, it is important to be aware of this tumor to prevent any delay in its diagnosis and treat it completely.

Compliance with Ethical Standards

Conflict of Interest None.

Consent Written informed consent was obtained from the patient for publication of this Case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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