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A surgical conundrum: Managing primary tracheal adenoid cystic carcinoma with preservation of larynx

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Abstract

Title: A surgical conundrum: managing primary tracheal adenoid cystic carcinoma with preservation of larynx.

Introduction: Tracheal tumors are extremely uncommon. The incidence of tracheal tumors is 0.09–0.2% out of which 10% are Adenoid cystic carcinoma. The origin of adenoid cystic carcinoma is from the submucosal glands of the tracheal tree. It is typically regarded as a low-grade malignancy with a favorable prognosis.

Case Presentation: A 35 years old male patient came to Out Patient Department with chief complaints of change in voice since 6 months and stridor since 1 week. On Indirect laryngoscopy there was left sided vocal cord palsy with submucosal swelling in subglottic area compromising the airway. Contrast Enhanced Computed Tomography revealed a hyperintense lesion of about 5.5 cm craniocaudally in the posterolateral aspect of the subglottic trachea and inferior border of the cricoid pushing left thyroid lobe anterolaterally along with significant obstruction of the airway at the level of cricoid. An emergency tracheostomy was done at lower level to secure the airway. Flexible bronchoscopy showed a 1.5-2cm stenotic segment obstructing 50 to 75% of the airway in a subglottic region with left vocal cord palsy. A biopsy of the lesion took externally which came as an Adenoid Cystic Carcinoma. On Magnetic Resonance Imaging there was no involvement of the nerve or metastasis. Wide local excision of the lesion involving tracheal resection with cricotracheal anastomosis and Montgomery Tube insertion was done followed by chemoradiation. Montgomery tube was successfully removed and the patient satisfactorily decannulated.

Conclusion: Adenoid cystic carcinoma should be considered as a differential diagnosis in patients with changes in voice and stridor. As adenoid cystic carcinoma is very slow-growing tumor so instead of being so radical a combined approach that gives a functional larynx and better quality of life should opt.

Keywords: Trachea; Adenoid Cystic Carcinoma; Larynx; Cricoid; Subglottis

1. Introduction

Tracheal tumors are extremely uncommon. The incidence of tracheal tumors is 0.09–0.2% out of which 10% are Adenoid cystic carcinoma [1]. Adenoid cystic carcinoma (ACC) is the second most common primary malignant tumor of the trachea, after squamous cell carcinoma [2]. The origin of adenoid cystic carcinoma is from the submucosal glands of the tracheal tree. It is typically regarded as a low-grade malignancy with favorable prognosis. The clinical and pathologic features of adenoid cystic carcinoma of the trachea were first reported in 1859 by Billroth. Dyspnea and wheezing, which are brought on by central airway blockage, are the most common symptoms of patients with ACC of the airway [3,4]. Both complete and incomplete resections procedures with acceptably less mortality and long survival expectation can be done successfully in most of the patients [5].

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2. Case Presentation

A 28 year old male patient presented to ENT Out Patient Department with chief complaints of change in voice since 6 months and breathing difficulty since 1 week. There was no history of swelling in the neck, dysphagia, or hematemesis. On Indirect laryngoscopy and subsequently 70 degree scopy there was left-sided vocal cord palsy with appreciable submucosal swelling down in the subglottic area compromising the airway. We have done Contrast Enhanced Computed Tomography (CECT) which revealed a hyperintense lesion of about 5.5 cm craniocaudally in posterolateral aspect of subglottic trachea and inferior border of cricoid pushing left thyroid lobe anterolaterally along with significant obstruction of airway at the level of cricoid (Fig. 1.).

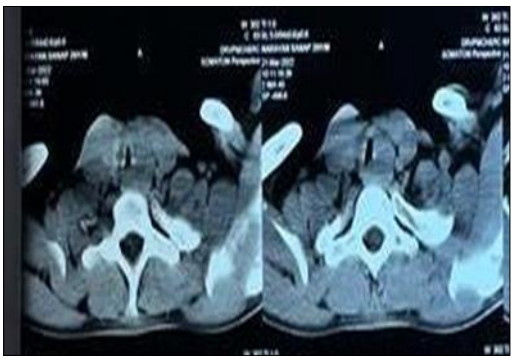

Figure 1 CECT showing narrowing of airway at the level of cricoid

On Magnetic Resonance Imaging (MRI) there was no involvement of the nerve or metastasis. Low Emergency Tracheostomy was done to relieve the worsening stridor in ward itself. After stabilising the patient we have done Fiberoptic Brochoscopy which showed a 1.5-2cm stenotic segment obstructing 50 to 75% of the airway in the subglottic region. The stenotic segment was lying about 1 to1.5 cm below the level of true vocal cords. As the growth was submucosal so we preffered to do biopsy externally to get the representative sample. The biopsy came out as adenoid cystic carcinoma. After all evaluation and detail discussion with patient and his attendant we have decided to proceed with the surgery but with the goal of preservating the larynx as patient was very young and adenoid cystic carcinoma is a very slow growing tumor. Wide local excision involving resection of two tracheal rings and inferior margin of cricoid keeping microscopic margin positive was done along with cricotracheal anastomosis after supralaryngeal drop with Montgomery Tube insertion [Fig.2, 3, 4, 5].

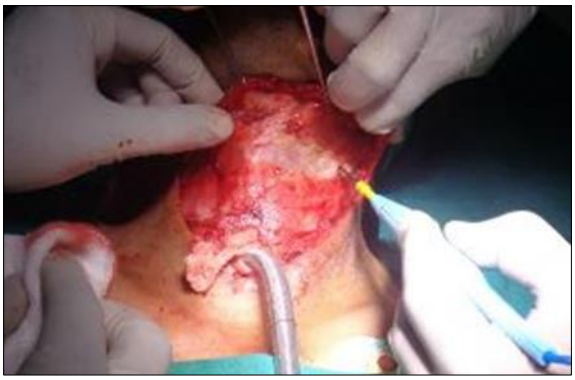

Figure 2 Horizontal neck skin crease incision with subplatysmal flap elevation

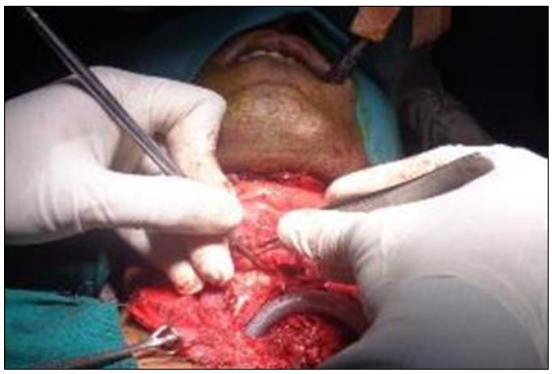

Figure 3 Preparation for tracheal resection

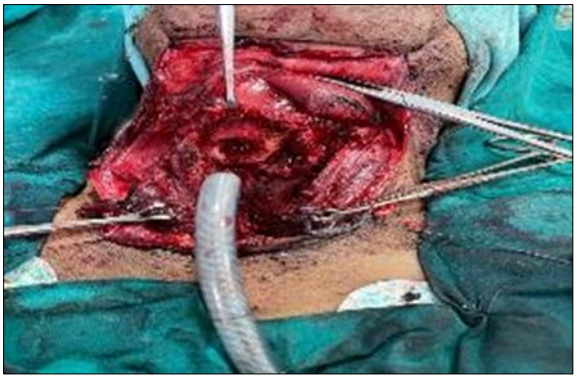

Figure 4 Tracheal resection displaying the preserved right thyroid lobe and lower margin of cricoid

Figure 5 Supralaryngeal drop with cricotracheal anastomosis

We have preserved the right lobe of thyroid also. Histopathology report came as adenoid cystic carcinoma with superior margin positive of cricoid cartilage (Fig. 6).

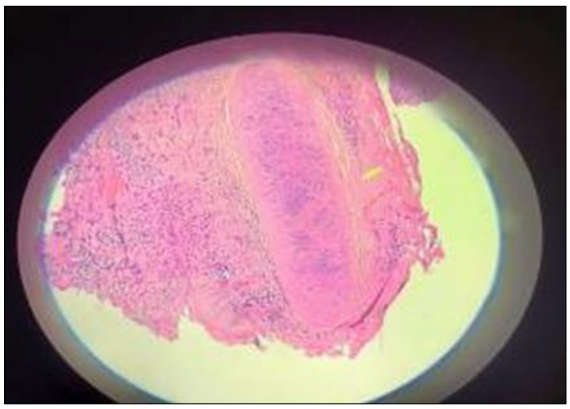

Figure 6 Histopathology showing tracheal cartilage with tumor cells

After suture removal we have sent patient for oncology opinion for ChemoRadiotherapy (CTRT). The patient received six cycles of chemotherapy with cisplatin and 33 cycles of radiotherapy for positive margins in the cricoid area. After completion of radiation therapy the Montgomery tube was successfully removed and the patient was satisfactorily decannulated. On last follow up of 6 months the patient was doing fine and there was no any respiratory difficulty. We were able to successfully treat adenoid cystic carcinoma of trachea by tracheal resection and anastomosis while preserving right hemi thyroid and larynx. Patient is disease free and nicely communicating at the moment without tracheostomy. The patient was followed up after completion of radiation post 6 weeks with X-ray Soft Tissue Neck showing patent airway.

3. Discussion

Primary tracheal malignancy is a rare condition with an annual incidence of about 1 per 1 million. Squamous cell carcinoma and adenoid cystic carcinoma are the two histological categories that make up the majority of tracheal malignancies, each accounting for 49% and 23% of cases respectively [7]. Since tumors may not produce symptoms until they obstruct 75% of the tracheal luminal diameter, diagnosis is frequently postponed [8]. In our case also 50 to 75% obstruction was present. The preferred course of treatment for ACC patients is complete surgical resection [6]. A laryngectomy is not recommended for treating local disease because up to 59% of patients with positive airway margins following tracheal resection, adjuvant radiotherapy decreases local recurrence instead. The patient would be left aphonic with a mediastinal tracheostomy, poor respiratory function, and a dismal quality of life after a total laryngectomy with tracheal resection [7]. In our case, we preserved right sided thyroid lobe along with the complete larynx after tracheal resection and anastomosis thus minimizing the need for lifelong thyroid supplements and giving the patient a functional larynx, greater self-esteem, and a higher quality of life.

4. Conclusion

Adenoid cystic carcinoma should be considered as a differential diagnosis in young patients with a change in voice and stridor. Adenoid cystic carcinoma is a slow-growing tumor, therefore rather than being so radical a combined approach that results in a functional larynx and improved quality of life should be chosen, especially in young individuals as this will boost their self-confidence.

Compliance with ethical standards

Disclosure of conflict of interest

All Authors stated that there is no conflict of interest regarding this publication.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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