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Case Report

Case Report on Adenoid Cystic Carcinoma of Larynx: A Rare Tumour

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Abstract

Adenoid cystic carcinoma (ACC) of larynx is a rare malignant tumour with unknown aetiology. It accounts for less than 1% of all laryngeal malignancies worldwide. ACC of larynx is slowly progressive and arises from accessory salivary glands. It is known for multiple recurrences and distant metastasis, which may occur even 5 years after treatment. In this article we report a case of 57 year old women with laryngeal adenoid cystic carcinoma who presented with a 5-month history of hoarseness of voice and noisy breathing. The patient underwent elective tracheostomy and biopsy followed by total laryngectomy with bilateral neck dissection. As laryngeal adenoid cystic carcinomas are well known for relapses and metastasis, regular follow up is necessary for early detection.

Keywords: Adenoid Cystic Carcinoma; Laryngeal Malignancy; Laryngectomy; Minor Salivary Glands

Introduction

Adenoid cystic carcinomas account for 1% to 5% of all head and neck malignancies and mostly occur in minor salivary glands, which are found extending from paranasal sinuses to the larynx [1,2].

Adenoid cystic carcinomas are rare in larynx due to the sparseness of accessory salivary glands and contributes to less than 1% of laryngeal malignancies. ACC which were previously known as cylindromas [3] arise from sub mucosal minor salivary glands and the aetiology remains unknown. Laryngeal adenoid cystic carcinomas are found mainly in the age group between 50 - 60 years with no significant difference among gender [4].

Laryngeal adenoid cystic carcinomas arise from sub epithelial glands and 2/3 of these tumors occur in the subglottis [5]. Laryngeal adenoid cystic carcinomas most commonly present as sub mucosal masses and are difficult to detect early as they spread in a sub mucosal fashion and therefore, most of the patients are diagnosed late i.e. in advanced stage. ACC commonly spreads by perineural extension and frequently recur following initial treatment. Distant metastasis may occur with lung being the most frequent site.

Case Report

A 57-year-old female, presented to our opd with a history of hoarseness of voice and noisy breathing since 5 months. The patient had no history of dysphagia, cough, laryngeal or prelaryngeal pain. The patient was a known case of diabetes mellitus. Indirect laryngoscopy detected growth over the right vocal cord with fixation of right vocal cord and growth extending into sub glottis. The laryngeal mucosa appeared healthy with no areas of necrosis. The supra-glottic area was normal. On neck examination, the patient had no palpable mass or lymph nodes and there was no impairment of laryngeal motion.

Figure 1

Computerized tomography (CT) of the neck with contrast medium showed the presence of mass in right true vocal cord extending caudally to involve right half of sub-glottic area with nodular intra luminal projection. Superiorly it is extending to involve right false vocal cord. Right half of cricoid cartilage is involved along its posterior aspect.

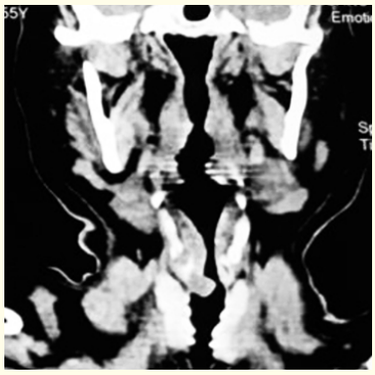

Figure 2

Elective Tracheostomy followed by direct laryngoscopy and biopsy of the growth was planned considering the extension of mass to prevent any post-operative stridor and it also helped for better visualization. A biopsy was taken from the tumour and sent for histopathology. HPE report was s/o adenocarcinoma and pathologist advised IHC markers for confirmation and exact tumour typing. After IHC tumour panel the diagnosis was confirmed as adenoid cystic carcinoma. The patient was treated surgically and total laryngectomy with bilateral neck dissection was done. The surgical margins were free of tumour and the final pathology report showed negative margins. Seven days post-operatively, the patient was discharged. Since this patient had no lymphatic spread and no perineural invasion, she was not scheduled for adjuvant radiotherapy. The patient was explained regarding mandatory regular and long term follow up to look for recurrence or metastasis.

Discussion

Adenoid cystic carcinoma is extremely rare in larynx due to the sparseness of accessory salivary glands. Adenoid cystic carcinomas are slowly progressive tumours with high recurrence rate and are known for late distant metastasis.

According to the pattern of disease distribution, the supraglottic area is the most commonly involved area followed by the glottis and the subglottic area [6]. Ellington., *et al*. [7] in their study found that the subglottic region is the most common primary site for laryngeal ACC with 58.2%. There are no distinct predisposing factors for laryngeal adenoid cystic carcinoma [8].

The clinical signs and symptoms of laryngeal ACC may vary according to the tumour location, size and extent. Supraglottic tumours present with dysphagia, glottic tumors present with hoarseness or dyspnea, where as stridor and airway obstruction are symptoms of subglottic tumours.

Early diagnosis is difficult and often missed as the disease spreads in submucosal fashion and therefore, most of the patients with laryngeal ACC are diagnosed late i.e. in the advanced stage. Subglottic tumours invade much deeper by the time they are diagnosed.

ACC of larynx has a high rate of neurological involvement and characterized by local invasion, multiple recurrences and rarely metastasis to cervical lymph nodes. The most common site for distant metastasis is the lung followed by bone and liver. Even early lesions have the potential for distant metastasis. Distant metastasis (especially in the lungs) can occur even after a long period of treatment [9] and therefore, imaging is necessary for evaluation at the time of follow-up.

Histopathological analysis is essential pre-operatively as it is difficult to differentiate from squamous cell carcinoma based on clinical history. Adenoid cystic carcinomas are composed of epithelial and myoepithelial cells and are basal cell like malignant tumours. Although neck metastasis can occur, it is found only in 10 to 15% of the cases [10].

CT is recommended in the pre-operative evaluation to assess the location of tumour, its extension and also to look for regional and distant metastasis. In our case, CT with contrast medium was done and it showed the presence of mass in right vocal cord extending caudally to involve right half of sub-glottic area with nodular intra luminal projection. Superiorly it is extending in to right false vocal cord. Right half of cricoid cartilage is involved along its posterior aspect.

The treatment options for laryngeal ACC remain controversial but according to most of the authors, treatment of choice is wide-margin local excision (partial or total laryngectomy depending on the location and size of the tumor) with radical neck dissection for patients with confirmed nodal metastasis [11,12].

Postoperative radiation is advocated by Coca-Pelaz., *et al.* [13] as radiotherapy not usually radiocurable postoperative radiotherapy can be used as an adjuvant modality in cases with positive surgical margins or cases with perineural spread. The dose is a prerequisite for the efficacy, the recommended dose is 60 Gy or greater and of-

ten it is observed that if the dose is less than 60 Gy the chances of relapse is more [14-16].

The use of chemotherapy in ACC remains controversial and it is recommended as palliative therapy in patients with advanced disease. The role of chemotherapy in ACC of the head and neck is usually confined to advanced (nonresectable), recurrent, or metastatic disease.

Five-year survival rates are low for laryngeal ACC i.e. 12 - 17% [17] therefore, more effective treatment options are required. As laryngeal adenoid cystic carcinomas are known for multiple relapses and distant metastasis the patient should be explained about regular and long-term follow-up in order to detect relapse and metastasis.

Conclusion

Adenoid cystic carcinoma of larynx is rare and it is a slowly progressing cancer. As early diagnosis is often difficult, a high degree of suspicion is needed and late diagnosis at times can be fatal. The treatment of choice is wide-margin surgery with or with out postoperative radiotherapy. Post-operative radiotherapy is advocated for cases with perineural spread or positive margins. All the patients require long-term surveillance in order to detect relapses and distant metastasis.

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