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Case Report

Adenoid Cystic Carcinoma of the Tongue: A Rare Case Report

Nupur Bansal, Deepak Kumar, Paramjeet Kaur, Ashok K Chauhan, Anil Khurana
Department of Radiotherapy, Pt BDS PGIMS, Rohtak-124001, Haryana

***Corresponding author**
Nupur Bansal
Email: drnupurbansal@gmail.com

Abstract: Adenoid cystic carcinoma is a malignant neoplasm arising from the salivary glands. The most striking feature of this tumor is that it is locally aggressive, with perineural invasion, tendency to metastasize, high recurrence rates. The parotid gland is the most commonly affected site in the head and neck region, tongue being relatively rare. The current study presents a rare case of 6 years old female child presenting as a recurrent case of adenoid cystic carcinoma of tongue.
Keywords: Adenoid cystic, carcinoma, tongue, radiotherapy

INTRODUCTION

Adenoid cystic carcinoma is an epithelial malignant neoplasm originating from the salivary glands [1]. Billroth termed it as cylindroma in 1859. Then, the lesion was further coined as adenoid cystic carcinoma by Foote and Frazell in 1953 [1]. It represents about 2-4% of malignant tumors of head and neck region and about 5-10% of all malignant salivary gland neoplasms [2]. The peak incidence of disease occurs mainly among women between 5^{th} and 6^{th} decades of life [2]. The most frequent site is parotid gland (30%) followed by disease of submandibular region. Approximately 40% of adenoid cystic carcinomas are found in minor salivary glands [3]. Clinically, painless slow growing mass is the usual complaint. It has propensity to invade peripheral nerves and associated with high recurrence rates [3]. Histopathologically, defines patterns as cribriform, tubular and solid type. Cribriform type is most common. The solid variant is associated with worse prognosis. Surgery remains the mainstay of treatment. Radiotherapy is recommended mainly as adjuvant treatment after surgery and in selected cases as primary treatment [3].

CASE REPORT

A 6-year-old girl presented at our institute with a complaint of swelling on left side of tongue for past 1 month. The swelling increased gradually in size. It was not associated with pain or difficulty in swallowing. The clinical examination revealed a nodule of ~ 2x2cm on the tip of tongue and adjacent right lateral border of anterior tongue, which was non tender, hard in consistency. Figure 1 is showing the lesion on anterior part of tongue as already described. She had a history of being operated for a similar swelling at tongue about one year back. Partial glossectomy has been done at that time. Histopathological report confirmed the findings of adenoid cystic carcinoma, solid variant, with growing edge of tumor going around nerve bundle. Immunohistochemistry analysis demonstrated CD99 positive, CD 117 positive, HMB-45 was expressed in cells around minor salivary glands. Patient was diagnosed as a recurrent case of adenoid cystic carcinoma of tongue. Baseline investigations and metastatic work up have shown normal findings. She was treated with external beam radiotherapy; 60Gy/30Fr/6weeks was planned and given. Computed Tomography scan of the head and neck region described a defined area on tip of the tongue, in midline and bilateral paramedian location. Figure 2 is representing the same. No abnormal contrast enhancement was seen in the scan and the lesion might be the result of post radiotherapy fibrosis. She was asymptomatic after one year of follow up.

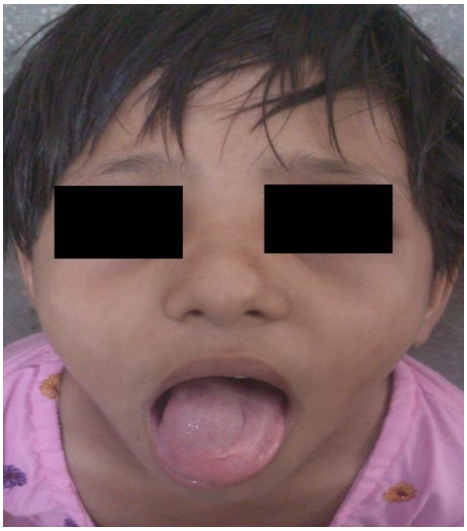

Fig 1: Patient presenting as lesion at tongue

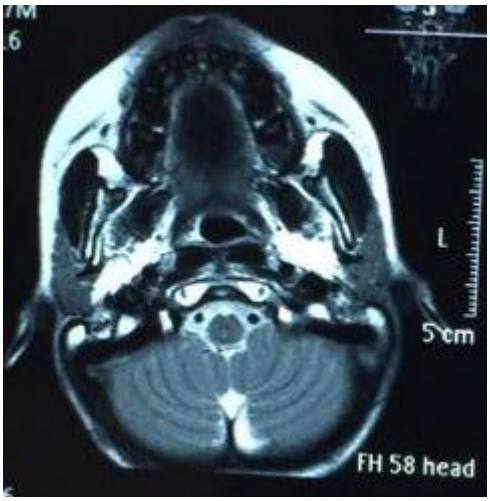

Fig 2: CT image of the patient: showing post radiotherapy changes at tongue

DISCUSSION

The WHO definition of Adenoid cystic carcinoma (ACC) is, "A basaloid tumor consisting of epithelial and myoepithelial cells in various morphological configurations including solid, tubular and cribriform patterns." [4]. ACC predominantly occurs in women and has a peak incidence in the fifth and sixth decades of life [4]. ACC is locally aggressive tumor with infiltrating tendency and is characterized by perineural invasion, leading to high recurrence. It is one of the most biologically harmful tumors of the head and neck. Lung is the most common site of distant metastases. Other sites include bone, liver, kidney and prostate [1].

Histologically, ACC has three different variables - cribriform, tubular and solid [4]. A good prognosis is influenced by the histopathology type, this relationship has been studied by Batsakis, who proposed that the histological subtypes of lower-grade malignancy (tubular and cribriform) have a better prognosis than those of higher grade malignancy (solid) as the latter are associated with early distant metastases, recurrences and a higher mortality rate [5]. ACCs have been histologically graded on the basis of their solid component: Grade 1, no solid component; grade 2, <30% solid component; and grade 3, ≥30% solid component [14-16]. The solid subtype presents the worst prognosis, with a survival of 34% at 10 years, in contrast to 76% of the cribriform and 100% of the tubular subtypes [1]. Polymorphous low grade adenocarcinoma and pleomorphic adenoma with cribriform pattern, should be considered in the differential diagnosis due to similarities in the histological pattern, however, final diagnosis can be made with the help of immunohistochemistry markers [3]. Vimentin expression is well known in some epithelial cells; thus ACC appears to frequently contain vimentin. Markers of neuroendocrine differentiation (S-100, MMB-45), c-KIT (CD117), a receptor tyrosine kinase, is well known to be expressed in ACC [40, 41] [6].

The primary objective in treating the patients of ACC of tongue is local control, preservation of function and prevention of distant metastasis, so early detection of the disease is beneficial. Surgery is the mainstay of treatment and the extent of surgery depends on the size of the tumor. Partial glossectomy, hemiglossectomy, total glossectomy with or without preservation of the larynx, or total glossectomy with or without reconstruction can be done [5].

Compared with other malignancies, it is difficult to excise ACC completely leading to positive surgical margins in most of the cases. The role of adjuvant radiotherapy is controversial. Though, it is advised to give post-operative radiotherapy in cases of positive surgical margins. Some studies have demonstrated the role of adjuvant radiotherapy following surgery in all patients regardless of the margin status, with an increase in the local control rate at 5 or 10 years. However, in some other studies radiotherapy is reserved for advanced stages and as adjuvant in positive microscopic margins [1].

Chen *et al.;* found that the independent predictors of local recurrence were T4 disease, perineural invasion, omission of postoperative radiotherapy and major nerve involvement. Surgery and adjuvant radiotherapy, to doses in excess of 60 Gy offer the best chance of curing patients in resectable ACC of the head and neck [7]. Patients with advanced, unresectable disease can be treated with definitive radiotherapy alone. Miglianico et al stated that the improvement in survival rates doesn't depend on the loco regional control of the disease in patients treated with surgery alone versus with combined modality therapy [8]. There is no uniform consensus on the role of adjuvant chemotherapy. Some studies have put forward the importance of adjuvant chemotherapy due

to the high risk of hematogenous dissemination. Chemotherapy can be given either after surgery or radiotherapy. Concomitant chemo-radiotherapy is an alternative for patients with advanced disease to preserve the function of the involved organ, but it is relatively inefficacious in treating ACC [1].

ACC presents a poor prognosis and it is therefore necessary to carry out long-term follow-up (Tables II and III). It has been reported to be a recurrent disease following treatment, ranging from 19 to 51% with a median time to recurrence of 2.7 years. The reported overall survival in ACC of the head and neck ranges from 51-56.5% and 32.5-34% at 5 and 10 years, respectively. In conclusion, the current study presents a noteworthy case of c-kit positive posterior tongue ACC treated with surgery and adjuvant radiotherapy [1].

Targeted therapy also seem to influence the treatment of ACC. High proportion (85%) of ACCs stain positively for epidermal growth factor receptors (EGFR) and may respond to agents that act as EGFR inhibitors. Also, c-kit (CD-117), a tyrosine kinase receptor which is involved in the growth and development of tissues, is expressed in some cases of ACC [9]. Alcedo *et al.;* identified favourable results in two cases of unresectable ACC who were treated with imatinib mesylate. However, disease progression is seen during the treatment with imatinib mesylate [10]. Though, the use of imatinib mesylate in direct treatment of non-resectable or metastatic disease remains controversial, which is also due to its high cost [1].

CONCLUSION

The primary treatment objective in treatment of adenoid cystic carcinomas is local control, function preservation, and prevention of distant metastasis. Tongue lesions remain challenging for diagnosis and treatment due to their anatomical location and rarity of disease presentation at these sites. Early diagnosis, the histopathological pattern and the presence or absence of distant metastases helps in timely intervention and improved survival of the patient.

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