

Adenoid Cystic Carcinoma of Buccal Mucosa: A Rare Case Report

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Abstract: Adenoid cystic carcinoma is an uncommon, slow growing malignant salivary gland tumor that is characterized by wide local infiltration, perineural spread, propensity to local recurrence and distant metastasis. Among intra oral adenoid cystic carcinoma, buccal mucosa is among the rarest sites. In this paper, the authors present a case of adenoid cystic carcinoma affecting the buccal mucosa in a 36-year-old male patient. We have discussed the clinical features, histopathology, diagnosis and treatment along with a brief review of the relevant literature. The peculiarity of the lesion and the site we made is the key factor in the presentation

Keywords: Adenoid cystic carcinoma, malignant salivary gland tumor, buccal mucosa

1. Introduction

Salivary gland neoplasms constitutes a diverse group of tumors, exhibiting different histological characteristics and variable clinical behavior patterns^{1,2}. Adenoid cystic carcinoma (ACC) was first described by Theodor Bilroth in 1856, as cylindromas. The term 'ACC' was introduced by Ewing (Foote and Frazell) in 1954.^{3,4}

It presents a widespread age distribution, peak incidence occurs predominantly among women, between the fifth and sixth decades of life⁴. The majority of the tumours arise in the major salivary glands, minor salivary glands of the oral cavity and mucous glands of the upper respiratory tract.⁵

According to recent statistics 26.8% of cases occur in the parotid, 24% in the submandibular gland, 20.5% in the palate, 5% in the tongue, around 4% each in the lips and buccal mucosa and 1.2% in the sublingual gland⁶. In the present case, ACC was located on the rare region of the oral cavity (buccal mucosa) of a 36-year-old patient with the histopathological features showing cribriform pattern with no perineural invasion.

2. Case Presentation

A 36 year male patient reported to our Department with complaints of discomfort in the lower right inner cheek region since 20 days. Pt gave a history of notice of a fibrous band in the inner cheek one month back (Figure 1). No change in the size of the band, no change in the structure No history of pus discharge and no pain. On palpation a fibrous band palpated over the right buccal mucosa which was seen 1 cm below the occlusal plane in 16 17 region, firm in consistency, no tenderness was present on palpation. On head and neck examination, no palpable lymph nodes were present. On the basis of clinical details, Soft tissue

calcification on the right buccal mucosa and Calcification of the minor salivary gland were considered as provisional diagnosis.

3. Investigations

Incisional biopsy was taken from right buccal mucosa. Microscopic sections showed lesional cells arranged as lobules, nests and file like pattern in few areas. Focal duct like areas seen with single layer of cells lining these spaces with secretions present in few. The cells do not show polymorphism and have minimal eosinophilic cytoplasm. The nuclei show pale staining and dispersed nucleoli and appear granular. Cells infiltrating into adjacent connective tissue and muscle fibers. Focal areas of hyalinization are noted. The histopathology was suggestive of Adenoid cystic carcinoma. (Figure 2)

MRI of neck showed enhanced thickening of right posterior buccal mucosa 22x11x18 mm extending into the right retromolar trigone and right superior gingivobuccal with no obvious bone erosion and abutting the right lateral pterygoid and masseter muscle .No significant cervical lymph node enlargement.

4. Treatment

Regarding the surgical technique, in our patient, we have done wide local excision of the lesion with a free margin of 1.5 cm anteriorly, 1.5 cm posteriorly, 1.5 cm superiorly, and 1.5 cm inferiorly, with upper alveolectomy, measuring 6cm anteroposteriorly, 5 cm mediolaterally, 5 cm superoinferiorly bearing one tooth and marginal mandibulectomy also done , segment of mandible measuring 6cm in length bearing one tooth associated with selective neck dissection . Level 1 -4 lymph nodes has been dissected out. Left radial artery forearm free flap was taken out to

reconstruct the resected portion. Complete surgical removal of the lesion was performed (figure 3). Adjuvant Radiotherapy was performed 30 fractions, 200cGy with total dose of 6000cGy for 10 days.

Outcome and Follow-Up

After surgical excision of the tumour and radiotherapy, the margins of the tumour were clear microscopically and a regular follow-up of the patient with repeated scans and radiographs has not revealed any signs of recurrence or metastasis until today (Figure 4)

5. Discussion

Adenoid cystic carcinoma was first described by Theodor Bilroth in 1856, as cylindromas, in his histological studies where he described it as long amorphous compartments “cylinders”. It is only recently that the tumor has been renamed as adenoid cystic carcinoma (ACC) (Kaiser), called which is a rare tumor accounting for 1 % of all head and neck malignancies and 10 % of all salivary gland neoplasms.⁷

ACC is an uncommon form of malignant neoplasm that arises most commonly in the major and minor salivary glands of the head and neck. ACC make up about 6 % of all salivary gland tumors. They make up 15–30 % of submandibular gland tumors, 30 % of minor salivary gland tumors, and 2–15 % of parotid gland tumors. The long natural history of this tumor, its propensity for perineural invasion, and its tendency for local recurrence are well known. Although it presents a widespread age distribution, peak incidence occurs predominantly among women, between the 5th and 6th decades of life. It is a slowly growing but highly invasive cancer with high recurrence rate. Lymphatic spread to local lymph nodes is rare. Hematogenous spread, however, occurs often in the course of the disease. Perineural spread of ACC has long been recognized. The literature revealed that the region of Gasserian ganglion to be the most common site of involvement (35.8 %) ⁸. Microscopically, the ACC is composed of a mixture of myoepithelial cells and ductal cells that can have a varied arrangement. Histopathologically, ACC presents three patterns, cribriform, tubular and solid.⁸

The prognostic factors of ACC depend on tumour site, tumour stage, the presence of perineural invasion and tumour grade. Tubular and cribriform subtypes have better prognosis than solid subtypes. We performed a thorough microscopic evaluation of the specimen for marginal clearance ^{9,10,11} The treatment aspect of the tumor is chiefly surgical although in some cases surgery has been successfully coupled with radiotherapy. Radiation therapy as the sole treatment modality seems to be inadequate for ACC. Unlike most other type of salivary gland carcinoma, distant metastasis is far more frequent in ACC (as lungs, liver, kidneys, pelvic girdle). Lymphatic spread is far less common in this tumor.

6. Conclusion

Adenoid cystic carcinomas have a variable prognosis. The 5-year survival rate is 75 %, but 10-year survival rate is only 20 %, and survival at 15 years is about 10 %.⁸ Other factors which adversely affect the prognosis are perineural spread, distant metastasis and recurrent local lesions. The present case was seen as a fibrous band on the buccal mucosa giving a picture of calcification of the minor salivary gland or any calcification on the buccal mucosa, but the histopathological examination revealed a adenoid cystic carcinoma, so it is important to consider it as a differential diagnosis of any localized lesion involving buccal mucosa. The margins of the lesion were free from tumor cells and the patient has not reported recurrence till date.



Figure 1: Pre operative photograph

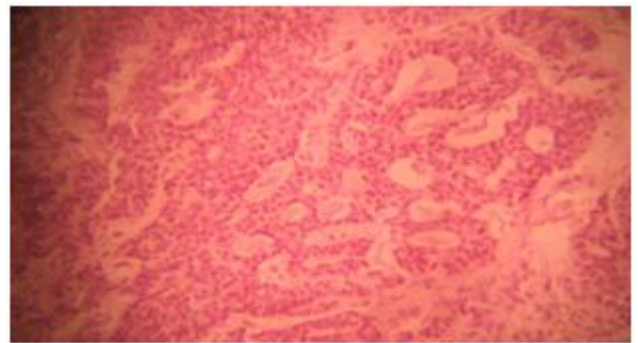


Figure 2

Microscopic sections showed lesional cells arranged as lobules, nests and file like pattern in few areas. Focal duct like areas seen with single layer of cells lining these spaces with secretions present in few areas



Figure 3: Intraoperative photograph

Post operative photograph



Figure 4

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