Case Report

Journal of Dental and Maxillofacial Surgery

Adenoid Cystic Carcinoma of Buccal Mucosa: A Rare Presentation

Kumar A¹, Rawat G^{2*}, Gupta S² and Deb S³

¹Department of Surgical Oncology, Dharamshila Narayana SS Hospital, Delhi, India

²Department of Head & Neck Oncology, Dharamshila Narayana SS Hospital, Delhi, India

³Department of Pathology, Dharamshila Narayana SS Hospital, Delhi, India

***Correspondence:** Garima Rawat, Department of Head & Neck Oncology, Dharamshila Narayana SS Hospital, Delhi, India, Tel: +91 9873776634; E-mail: garima3103@gmail.com

Received: May 11, 2018; Accepted: June 22, 2018; Published: June 27, 2018

Abstract

Adenoid Cystic Carcinoma (ACC) is an unusual slow growing, aggressive neoplasm of salivary gland, constituting less than 1% of all head and neck malignancies. It commonly affects adults in the fourth to sixth decades and typically involves minor salivary glands of palate followed by parotid, and submandibular glands. We present here a case of a 16-year-old female diagnosed with ACC involving the buccal mucosa and abutting the distal end of stenson's duct along with the surgical management and follow up.

Keywords: Adenoid cystic carcinoma; Minor salivary gland; Buccal mucosa; Perineural invasion

Introduction

Salivary gland neoplasms comprise a diverse group of tumors with varied histological characteristics and clinical behavior patterns. Salivary gland neoplasms show infiltrative behavior and late loco-regional recurrence and distant metastasis, these mandate a proper diagnosis and treatment [1].

Adenoid cystic carcinoma (ACC) is a rare tumor accounting for less than 1% of all head and neck malignancies and 10% of all salivary gland neoplasms. It is the fourth most common malignant epithelial salivary gland neoplasm with 50% occurring intraorally [2]. Extraorally, parotid gland (25%) is the single most common site of origin. It presents a widespread age distribution, peak incidence between the fifth and sixth decades of life, with preponderance for females. Most of the cases of ACC are submucosal and they appear as smooth, domed shaped swellings without overlying ulcerations [3]. Pain is an important symptom of this tumour due to its tendency

for perineural spread [2]. Lymph node metastases are rare although hematogenous spread, often to the lungs is characteristic.

Histologically, ACC exhibits three growth patterns: cribriform, tubular, and solid. The solid growth pattern is associated with a worse prognosis, caused by advanced stage and development of distant metastases. The choice of therapy is influenced by site, stage, histologic grade, and biologic behavior of the ACC [4]. Surgical excision with wide margins is the treatment of choice [5]. The minor salivary gland ACCs have a worse prognosis than those of the major salivary glands.

Here we present a case of ACC involving the buccal mucosa in a 16-year-old female with emphasis on the surgical management and follow up. Till date only six case reports of ACC involving the buccal mucosa have been published in the English literature, with none occurring before third decade.

Case Report

A 16 year old female patient reported to the outpatient department of a tertiary care hospital with the chief complaint of pain and swelling on right side of buccal mucosa since 4 months. The pain was mild, intermittent in nature and relieved on taking medication. The swelling was initially small in size, barely perceptible to the patient and it slowly increased in size over a period of 4 months to its present size. Patient reported when she observed visible facial asymmetry of face. Patient went to GTB hospital, where sialoendoscopy and biopsy was done from the lesion and histopathology findings were suggestive of Adenoid Cystic Carcinoma. From there, patient was referred to our hospital for management of the same. Medical history & personal history was non contributory. On inspection, there was facial asymmetry with slight swelling evident over right side of cheek without any signs of inflammation (Figure 1A). Intraorally, on inspection, there was mild inflammation over region of stenson's duct. On palpation, intraorally, firm, nontender, nodular swelling was evident measuring approximately 1.5×1 cm in size (Figure 1B). The overlying mucosa was normal in colour, texture and was afebrile to touch. On stimulation of salivation, the salivary flow from the stenson's duct was normal. On lymph node examination, no lymph nodes were palpable. Patient was advised further investigations including sialography of parotid gland, and Magnetic Resonance Imaging (MRI) of face and neck. MRI findings were suggestive of a circumscribed mass in right buccal space at the distal end of stenson's duct, overlying the buccinators muscle (Figure 2). The mass was approximately 1.08×0.7×1.13 cm without any significant lymphadenopathy.

Based on clinical, histopathology and imaging findings, a final diagnosis of Adenoid Cystic Carcinoma of buccal mucosa and distal end of stenson's duct was made. In order to rule out metastasis, MRI Brain and Chest X-ray were also done. There was no evidence of any distant metastasis. It was planned to carry out wide local excision via intraoral approach followed by reconstruction of new opening of stenson's duct. The patient was operated under general anaesthesia after giving circumferential incision around the lesion. Before giving incision, the opening of stenson's duct was identified and duct was cannulated (Figure 3A and 3B). After the fibres of buccinators were dissected, demarcation of the lesion was noticed and it was excised in toto (Figure 3C). The excised specimen (Figure 4) was sent for frozen section to evaluate the margin status. The reports of frozen section stated that the margins were free of tumour. Thereafter,

the mucosal defect was reconstructed with buccal pad of fat. Hemostasis was achieved and wound was packed with gauze. The cannula was left in place. The histopathological examination of the excised specimen revealed the tumour composed of large islands of basaloid tumor cells partially separated by fibrous connective tissue septae. The tumour cells were surrounding variably sized cyst like spaces containing pale basophilic material resembling a swiss cheese pattern (Figure 5A). The tumour cells were seen in and around the neural bundles also (Figure 5B). Thus, a final diagnosis of Adenoid Cystic Carcinoma with perineural invasion was given. After one week post surgery, the patient was examined and satisfactory healing of surgical site was evident. The cannula was removed and patient was advised oral food intake (Figure 6). Patient was referred to Department of Radiotherapy where it was planned to administer 30 cycles of Radiotherapy. After completion of radiotherapy, the surgical site had healed uneventfully. Patient is kept under regular follow up initially at an interval of 2 months up to 1 year.

Figure 1: A) Extraoral photograph showing slight facial swelling on right cheek region. B) Intraoral photograph showing palpable swelling near opening of Stenson's duct of right side.

Figure 2: MRI section depicting 1.08 x 0.7cm circumscribed mass in right buccal space at the distal end of stenson's duct, overlying the buccinators muscle (arrow).

Figure 3: Intra operative photographs showing.
Insertion of cannula into stenson's duct.
Curvilinear incision around the tumour.
Reconstruction of surgical defect using buccal fat pad.

Figure 6: Post-operative photograph (One month after surgery).

Discussion

Malignant neoplasms of the salivary glands in the head and neck are relatively rare, accounting for only less than 7% of all neoplasms, of which about 10% are Adenoid Cystic Carcinomas [1,2]. ACC was first described by Theodor Bilroth in 1856, as cylindromas, in his histological studies where he described it as long amorphous compartments called "cylinders". It is only recently that the tumor has been renamed as adenoid cystic carcinoma (ACC) (Kaiser) [1]. ACCs are mostly reported in the minor salivary glands, and less frequently in the major salivary glands. ACC of minor salivary gland origin occurs most frequently in the hard palate [2,3]. In the present case, patient presented with a lesion in buccal mucosa; a rare site of presentation. In English literature, only 6 cases of ACC involving buccal mucosa have been reported. The features of these cases have been tabulated in table 1.

ACC occurs predominantly in fourth to sixth decade of life with a female predilection of 3:2 [3]. However, in our case, patient is a young female in 2nd decade. It usually presents as a slow growing, firm, and unilobular mass. Pain is usually a common and important associated symptom, occasionally occurring before clinical evidence of the disease [3,4]. Similar symptoms were presented in this patient as well. The most characteristic clinical feature of this malignancy includes its slow growth rate, delayed onset of distant metastasis, increased local recurrences, and peri-neural invasion (14-22%) [5]. In the present case, there was no evidence of distant metastasis. The incidence of distant metastasis in ACC, most often the lung, is difficult to estimate, but is certainly dependent on the length of time that patients are followed, usually more than 15 to 20 years, but ranges from 35 to 50% [6].

Microscopically, the ACC is composed of a mixture

Figure 4: Excised specimen of tumour mass with arrow depicting the distal end of Stenson's duct.

Figure 5: Photomicrograph showing.

Table 1: Reported cases of ACC of buccal mucosa.

Author	Age/ sex	Location	Pain	Metastasis	Treatment modality	Recurrence
Singh S [11]	50/M	Right buccal mucosa	Yes (Mild)	-	Surgical excision	-
Ajila V [5]	48/F	Left buccal mucosa	Yes	-		-
Nailk LRK [12]	48/M	Right buccal mucosa	Yes (Mild)	-		
Kumar AN [9]	26/F	Right buccal mucosa	Yes (palpation)	-	Surgical excision	-
Vidyalakshmi S [3]	34/F	Left buccal mucosa	Yes	-		-
Dalirsani Z [8]	47/M	Right buccal mucosa	Yes (palpation)	-	Surgical excision followed by Radiotherapy and chemotherapy	Yes (To Frontal bone)

of myoepithelial cells and ductal cells that can have a varied arrangement. Histopathologically, ACC presents three patterns, cribriform, tubular and solid. The cribriform (glandular) pattern is the most classic and best recognized appearance, characterized by islands of basaloid epithelial cells that contain multiple cylindric, cyst-like spaces resembling Swiss cheese. These spaces often contain a mildly basophilic mucoid material, a hyalinized eosinophilic product, or a combined mucoid hyalinized appearance [7,8]. The microscopic evaluation of the present case showed features of cribriform variant.

The accepted treatment modality for localized ACC is radical surgery with complete resection and negative surgical margins. Cases with clinically positive cervical lymph nodes warrant modified radical neck dissection [6,9,10]. Radiation is optional for patients with small tumors (T1N0), but should be considered for those who have low grade tumors with perineural invasion or evidence of tumor spillage during surgery [11]. Elective radiation of clinically negative regional nodes to a lesser dose has also been proposed for patients whose tumors are located in lymphatic rich areas. Radiotherapy has proved to provide a better locoregional control when it was given post-operatively [10,12]. However, radiotherapy alone is not sufficient for treating this tumour per se. accepted protocol is to administer 60gy of radiation in a dose of 5 cycles per week for 6 weeks. We followed the similar protocol in our case. In our patient, we preferred surgical excision as the treatment because histopathologically the lesion was cribriform type which has better prognosis than the other variants but post operative radiotherapy was started on account of perineural invasion evident on histopathological examination of excised tumor mass. The nature of the

primary tumour, location, histomorphology, and its tendency to metastasize to distant sites, positive margins at the surgical site, determine the survival rate and prognosis. Tumors in major salivary gland with cribriform pattern, and no distant metastasis tend to have better prognosis [12,13].

Conclusion

ACC of minor salivary gland should be treated aggressively with aim of achieving negative surgical margins in order to prevent recurrence. Also, patient should be adequately followed up to monitor distant metastasis and recurrence.

References

1. Garg V, Roy S, Khanna KS, Bakshi PS, Chauhan I. Adenoid Cystic Carcinoma of Buccal Mucosa: A Rare Case Report. Indian J Otolaryngol Head Neck Surg. 2016 Sep;68(3):370–373.
2. Jaiswara C, Dhiman NK, Singh AK, Sharma NK, Verma V, Pandey A. Adenoid cystic carcinoma of the floor of the mouth – A rare presentation. J Oral Biol Craniofac Res. 2016 Nov;6(Suppl 1):S65–S69.
3. S V, R A. Adenoid Cystic Carcinoma of the Buccal Mucosa: A Case Report with Review of Literature. J Clin Diagn Res. 2014 Mar;8(3):266–268.
4. Bradley PJ. Adenoid cystic carcinoma of the head and neck: a review. Curr Opin Otolaryngol Head Neck Surg. 2004 Apr;12(2):127–132.
5. Ajila V, Hegde S, Nair GR, Babu SG. Adenoid cystic carcinoma of the buccal mucosa: A case report and review of the literature. Dent Res J (Isfahan). 2012;9(5):642–646.

6. Dillon PM, Chakraborty S, Moskaluk CA, Joshi PJ, Thomas CY. Adenoid cystic carcinoma: A review of recent advances, molecular targets, and clinical trials. *Head Neck*. 2016 Apr;38(4):620–627.
7. Neville BW, Damm DD, Allen CM, Bouquot JF (2002) Salivary gland pathology. In: Neville BW, Damm DD, Allen CM, Bouquot JF (eds) *Oral and maxillofacial pathology*. W.B. Saunders, Philadelphia, pp 389–496.
8. Dalirsani Z, Mohtasham N, Pakfetrat A, Delavarian Z, Ghazi A, Rahimi SA, et al. Adenoid Cystic Carcinoma of the Buccal Mucosa with Rare Delayed Frontal Bone Metastasis: A Case Report. *Journal of Dental Materials and Techniques*. 2016 Dec 1;5(4):208–212.
9. Kumar AN, Harish M, Alavi YA, Mallikarjuna R. Adenoid cystic carcinoma of buccal mucosa. *BMJ Case Reports*. 2013 Jun 11; 2013:bcr2013009770.
10. Triantafyllidou K, Dimitrakopoulos J, Iordanidis F, Koufogiannis D. Management of adenoid cystic carcinoma of minor salivary glands. *J Oral Maxillofac Surg*. 2016; 64:1114–1120.
11. Singh S, Gokkulakrishnan, Jain J, Pathak S, Singh KT. Adenoid Cystic Carcinoma of Buccal Mucosa. *J Maxillofac Oral Surg*. 2010 Sep;9(3):273–276.
12. Naik KL, Shetty P, Hegde P. Adenoid cystic carcinoma of buccal mucosa with extensive hyalinization: A unique case report. *Annals of Tropical Medicine and Public Health*. 2013 Sep 1;6(5):571.
13. Gondivkar SM, Gadgil AR, Chole R, Parikh RV. Adenoid cystic carcinoma: a rare clinical entity and literature review. *Oral Oncol*. 2011 Apr;47(4):231–236.

Copyright: © **Kumar et al.** This is an Open Access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.