

DENTAL ARTICLE

Adenoid Cystic Carcinoma of Sublingual Salivary Gland Obstructing the Submandibular Salivary Gland Duct

Venkata Suneel Kumar, Venkata Sarath Prathi, Rakesh Kumar Manne, Swapna Beeraka, Kannan Natarajan

Department of Oral Medicine and Radiology, Narayana Dental College and Hospital, Nellore, Andhra Pradesh, India

Address for correspondence:

Dr. Venkata Suneel Kumar,
Department of Oral Medicine and
Radiology, Narayana Dental College and
Hospital, Nellore, Andhra Pradesh, India.
E-mail: suneelkv55@gmail.com



Received : 24-07-2013

Accepted : 06-09-2013

Published : 29-10-2013

ABSTRACT

Sublingual salivary gland malignancies are extremely rare and account for only 0.3-1% of all epithelial salivary gland tumors. Here, we report a case of adenoid cystic carcinoma (ACC) of the sublingual salivary gland that presented as a swelling in the right anterior floor of the mouth obstructing the submandibular duct. Sublingual salivary gland ACC obstructing the submandibular duct is rare and only three cases have been reported in the literature until date. We discuss the different patterns of ACC seen during the pathologic investigations and its radiologic features.

Key words: Adenoid cystic carcinoma, sublingual salivary gland, submandibular salivary duct

INTRODUCTION

The term "adenoid cystic carcinoma" (ACC) was first used by Spies in 1930. This tumor was first described by Billroth as "Cylindroma" in 1859. It is a malignant tumor originating from the salivary glands. The sublingual gland is a rare site for this tumor. It is more common in the minor salivary glands (>25%) rather than in the major salivary glands (about 5%). The tumor is characterized by slow and relentless growth with a tendency for extensive local invasion.^[1] A particular feature

of ACC is the tumor's propensity for perineural invasion that may result in the spread along the involved nerves. ACC has a high recurrence rate and is prone to develop distant metastasis.^[2] A few point mutations in known cancer genes, including KRAS, BRAF and TP53 have been reported in ACCs. Recently, recurrent translocation t (6;9)(q22-23;p23-24) was identified resulting in the fusion of the v-myb myeloblastosis viral oncogene homolog (MYB) gene on chromosome 6 to the nuclear factor I/B gene on chromosome 9 in a major number of cases. However, the extent to which other genes contribute to the disease is not well understood.^[3] Most commonly followed the method of treatment for ACC seems to be surgical resection combined with radiotherapy.^[4]

A 64-year-old man presented to the Department of Oral medicine and Radiology with a 3-month history of swelling in the right anterior floor of the mouth. The swelling was initially small in size and gradually increased to the present

Access this article online

Quick Response Code:



Website:

www.clinicalimaging-science.org

DOI:

10.4103/2156-7514.120793

Copyright: © 2013 Kumar VS. 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 author and source are credited.

This article may be cited as:

Kumar VS, Prathi VS, Manne RK, Beeraka S, Natarajan K. Adenoid Cystic Carcinoma of Sublingual Salivary Gland Obstructing the Submandibular Salivary Gland Duct. J Clin Imaging Sci 2013;3:510. Available FREE in open access from: <http://www.clinicalimaging-science.org/text.asp?2013/3/2/10/120793>

size. The patient had noted reduced salivation for 2 months along with a little discomfort in speech and mastication due to the size of the tumor. Extra-oral examination revealed a palpable right submandibular lymph node measuring around 2 cm × 2 cm in size, tender and soft in consistency suggestive of an inflammatory nature of the lesion. Intra-oral examination revealed a tender, solitary, soft, and sessile swelling in the right anterior floor of the mouth measuring about 3 cm × 2 cm extending mesiodistally from central incisor to premolar region and superoinferiorly from the depth of the lingual vestibule to the level of occlusion. Examination also revealed an ulcer over the swelling [Figure 1]. Considering the history of reduced salivation and discomfort in mastication, provisional diagnosis of submandibular salivary sialolith and ranula was considered.

RADIOLOGIC FEATURES

ACC may require ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) for evaluating perineural, vascular, or skull base infiltration. ACC may demonstrate echo-free cystic areas on US.^[5] The limitation of US includes its inability to evaluate the deep lesions.^[6] Because CT is less accurate than MRI for detecting the extent of disease, MRI is preferred over CT.^[7] In case of bone pain, CT is the favored modality. ACC is best evaluated by using T1-weighted and fat-suppressed T2-weighted and gadolinium enhanced MRI.^[6] The lesion exhibits homogenous hypointense signals with ill-defined margins on T1-weighted MRI and after injecting gadolinium intravenously ACC shows continuous rapid or gradual enhancement pattern.^[7] On T2-weighted MRI, the signal intensity of ACC depends on the histological type of the lesion. Low signal intensity corresponds to the solid type



Figure 1: 64-year-old-male with swelling in the right anterior floor of the mouth diagnosed with adenoid cystic carcinoma. Intraoral photograph shows smooth surface with well-defined borders (solid arrow).

with poor prognosis and high signal intensity corresponds to either the cribriform or tubular subtypes with better prognoses.^[8]

Mandibular cross-sectional occlusal radiograph of the 64-year-old patient showed no identifiable sialoliths [Figure 2]. The patient's socioeconomic status did not allow use of the more expensive diagnostic imaging modalities. With a provisional diagnosis of ranula, surgical excision of the swelling was planned.

PATHOLOGIC FEATURES

Surgical excision of the swelling was done under local anesthesia. The swelling was found to begin from the sublingual duct and to obstruct the submandibular duct [Figures 3 and 4]. Histological investigation of the excised tissue stained with hematoxylin and eosin revealed basaloid epithelial cells arranged in cyst like spaces (duct like appearance), with many of the spaces presenting with hyalinized and mucoid appearance. The cells appeared small and uniform with deeply basophilic nuclei and a predominantly cribriform architecture suggestive of ACC arising from the right sublingual gland [Figure 5]. Considering the patient's age and socioeconomic status, no further investigations or radiotherapy was done. Patient was advised to return for a follow-up examination once in 3 months because of the relentless growth of ACC which has a tendency for extensive local invasion. Follow-up examination after 9 months showed no recurrence.

ACCs are characterized by histological and cellular heterogeneity. ACCs may show any one of the following patterns: Solid, trabecular, tubular, and cribriform.^[9] Among these cribriform type is the most common variant. Cribriform form shows the presence of extensive sheets, uniform bands and is composed of small, darkly stained,

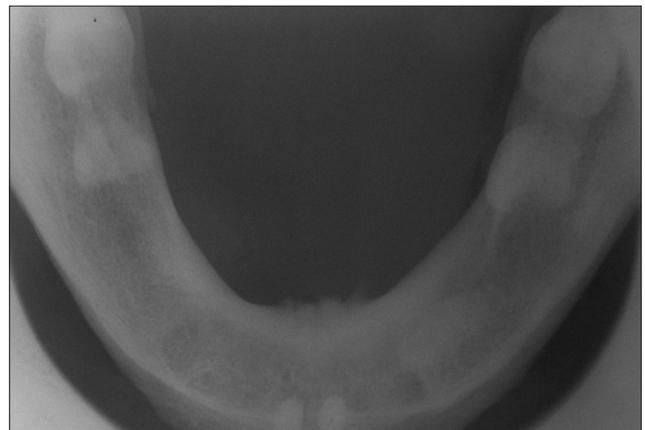


Figure 2: 64-year-old-male with swelling in the right anterior floor of the mouth diagnosed with adenoid cystic carcinoma. Mandibular cross-sectional occlusal radiograph shows no pathological changes.

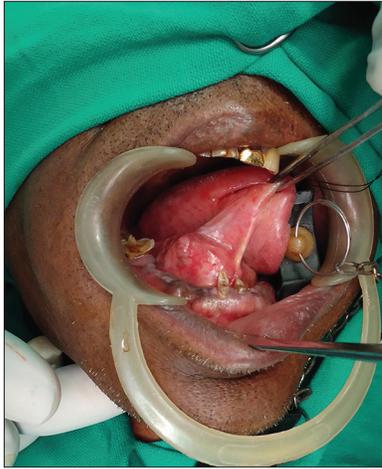


Figure 3: 64-year-old-male with swelling in the right anterior floor of the mouth diagnosed with adenoid cystic carcinoma. Intraoral clinical photograph shows surgical excision.

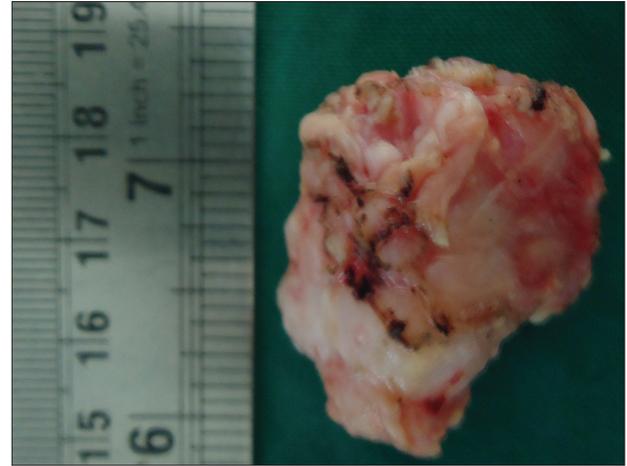


Figure 4: 64-year-old-male with swelling in the right anterior floor of the mouth diagnosed with adenoid cystic carcinoma. Photograph shows the surgically excised mass.

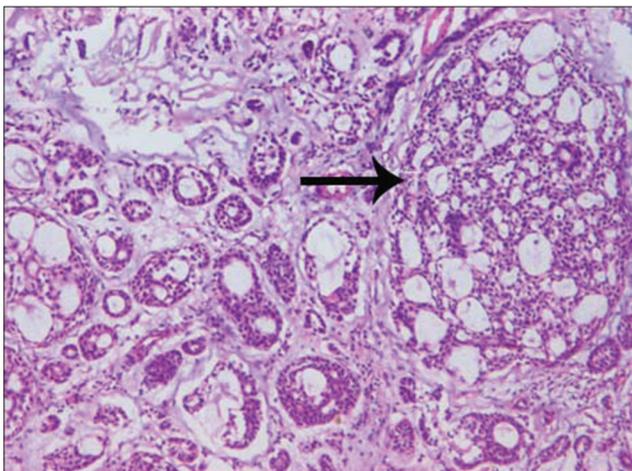


Figure 5: 64-year-old-male with swelling in the right anterior floor of the mouth diagnosed with adenoid cystic carcinoma. Photomicrograph of the Hematoxylin and Eosin stained tissue ($\times 10$) shows multiple basaloid epithelial cells with isomorphic and basophilic nuclei arranged in cribriform architecture (solid arrow).

slightly separated basal cells with duct like structures, which may contain secretory products. It may contain round to oval, intercellular spaces termed pseudocysts. The present case showed the most common cribriform pattern of ACC. According to many authors histological subtype influence the prognosis of the disease.^[9] Histological subtypes of tubular and cribriform ACC have a better prognosis than lesions with trabecular and solid patterns.

DISCUSSION

ACC of the sublingual salivary gland causing obstruction of the submandibular duct is exceedingly rare: only three cases have been reported in the literature.^[1,2,8] The peak incidence is seen during the fifth and sixth decades of life and shows a female predominance. Most of the patients with ACC present with a mass, which has been

there for months or years. Pain is felt by a few patients and is more often associated with advanced or recurrent tumors.^[10] In the present case, a 64-year-old-male patient presented with swelling of only 3-months duration and experienced mild pain, which may be associated with traumatic ulceration on the swelling rather than due to the tumor itself.

Possible treatment modalities of ACC include surgical therapy, radiotherapy, chemotherapy, and combined therapy.^[11] Radical surgery combined with radiotherapy is the commonly followed method of treatment.^[4] Post-operative radiotherapy is indicated for tumors associated with perineural spread, local recurrence, and distant metastases. According to Avery et al., radiation often produces tumor regression. ACC presents a limited response to chemotherapy.^[12] Alcedo et al., opened up a new possibility in the pharmacological treatment of this tumor by demonstrating its response to imatinib mesylate, a potent inhibitor of KIT tyrosine kinase, an enzyme involved in the pathogenesis of this tumor.^[13] Several genomic investigations have been carried out in ACCs to identify biological markers of therapeutic potential. These efforts, however, have been largely unrewarding and more investigations of new targets are needed.^[9] In the present case, only surgical therapy was preferred and further management was not done due to patient's socio-economic status. In spite of only surgical management, 9-month follow-up showed no recurrence. This may be due to better prognosis associated with the less aggressive cribriform type of the lesion. Further studies are required to validate treatment with only surgery in cases of tubular and cribriform forms of ACCs.

CONCLUSION

ACC of sublingual glands is rare and tumors causing obstruction to submandibular duct are extremely rare. Review of the literature shows that until date, only three cases have been reported. US, CT, and MRI are helpful tools in the diagnosis of ACCs. The diagnosis can be confirmed and the subtype identified by histological investigations. Most common method of treatment for ACC seems to be surgical resection combined with radiotherapy. The present case was managed only with surgical modality and 9-month follow-up showed no recurrence. Long-term follow-up and newer biological markers of therapeutic potential are necessary to validate use of only surgery to manage tubular and cribriform ACCs.

REFERENCES

1. Whear NM, Addy JM. Adenoid cystic carcinoma of the sublingual gland - An unusual presentation. *Br J Oral Maxillofac Surg* 1993;31:113-6.
2. McFall MR, Irvine GH, Eveson JW. Adenoid cystic carcinoma of the sublingual salivary gland in a 16-year-old female – Report of a case and review of the literature. *J Laryngol Otol* 1997;111:485-8.
3. Stephens PJ, Davies HR, Mitani Y, Van Loo P, Shlien A, Tarpey PS, et al. Whole exome sequencing of adenoid cystic carcinoma. *J Clin Invest* 2013;123:2965-8.
4. Triantafyllidou K, Dimitrakopoulos J, Iordanidis F, Koufogiannis D. Management of adenoid cystic carcinoma of minor salivary glands. *J Oral Maxillofac Surg* 2006;64:1114-20.
5. Benson BW. Salivary gland radiology. In: White SC, Pharoah MJ, editors. *Oral Radiology: Principles and Interpretation*. 5th ed. New Delhi, NA: Elsevier; 2004. p. 675.
6. Yousem DM, Kraut MA, Chalian AA. Major salivary gland imaging. *Radiology* 2000;216:19-29.
7. Rastogi R, Bhargava S, Mallarajapatna GJ, Singh SK. Pictorial essay: Salivary gland imaging. *Indian J Radiol Imaging* 2012;22:325-33.
8. Saito M, Nishiyama H, Maruyama S, Oda Y, Saku T, Hayashi T. Adenoid cystic carcinoma of sublingual gland involving the submandibular duct. *Dentomaxillofac Radiol* 2008;37:421-4.
9. Mitani Y, Roberts DB, Fatani H, Weber RS, Kies MS, Lippman SM, et al. MicroRNA profiling of salivary adenoid cystic carcinoma: Association of miR-17-92 upregulation with poor outcome. *PLoS One* 2013;8:e66778.
10. Andersen LJ, Therkildsen MH, Ockelmann HH, Bentzen JD, Schiødt T, Hansen HS. Malignant epithelial tumors in the minor salivary glands, the submandibular gland, and the sublingual gland. Prognostic factors and treatment results. *Cancer* 1991;68:2431-7.
11. Gondivkar SM, Gadail AR, Chole R, Parikh RV. Adenoid cystic carcinoma: A rare clinical entity and literature review. *Oral Oncol* 2011;47:231-6.
12. Avery CM, Moody AB, McKinna FE, Taylor J, Henk JM, Langdon JD. Combined treatment of adenoid cystic carcinoma of the salivary glands. *Int J Oral Maxillofac Surg* 2000;29:277-9.
13. Alcedo JC, Fábrega JM, Arosemena JR, Urrutia A. Imatinib mesylate as treatment for adenoid cystic carcinoma of the salivary glands: Report of two successfully treated cases. *Head Neck* 2004;26:829-31.

Source of Support: Nil, **Conflict of Interest:** None declared.