Original Research Paper Medicine

ADENOID CYSTIC CARCINOMA A RARE PRESENTATION IN THE FLOOR OF THE MOUTH – CASE REPORT

PRATHIMA SREENIVASAN	Professor and Head of the Department, Department Of Oral Medicine and Radiology, Kannur Dental College, Anjarakandy,Kannur, Kerala.
RAJEEV.A.K	Postgraduate Student, Department Of Oral Medicine and Radiology, Kannur Dental College, Anjarakandy,Kannur,Kerala.
CHETAN POOVAN VASANTHAN	Postgraduate Student, Department Of Oral Medicine and Radiology, Kannur Dental College, Anjarakandy,Kannur,Kerala.
PRABATH RAMAKARISHNAN	READER, Department Of Oral Medicine and Radiology, Kannur Dental College, Anjarakandy,Kannur,Kerala.

ABSTRACT Adenoid cystic carcinoma (ACC) is an unusual salivary gland malignancy that remains poorly understood.It is characterized by the proliferation of ductal (luminal) and myoepithelial cells in cribriform, tubular, solid, and cystic forms. Standard treatment, including surgery with postoperative radiation therapy, has attained reasonable local control rates, but distant metastases do not allow any improvement in the survival rate. In this paper we aim to present a case report of a 66 year old female patient with a rare reason of a linear solitary ulcer in the floor of the mouth, with a detailed discussion of the differential diagnosis and the investigative work up.It is a slow growing but aggressive neoplasm with a tendency for recurrence.

KEYWORDS : Adenoid cystic carcinoma; Floor of the mouth; Malignant; Salivary gland

1.INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare malignant tumour comprises less than 1% of all head and neck cancers and 20–25% of all salivary gland carcinomas.[1]The term 'adenoid cystic carcinoma' was coined in the year 1928 by Spies[2].About 30% of all ACC occurs in the minor salivary glands, and these have worse prognosis than those of the major salivary glands. The submandibular gland involvement is reported in 15%-30% of all cases, and in the parotid gland in 2%-15% of cases.[3]ACC are slowgrowing, locally invasive growth and is known for its perineuralspread, local recurrence, and distant metastasis to the lungand bone.[4]Commonest site of involvement is the palate.[5]Other less frequent intraoral sites are the lower lip, retromolar region and buccal mucosa.Cervical lymph node metastasis is found in 8–13% of cases. Distant metastasis is found in up to 50% cases in lungs and bones.[5]Clinically ACC lesions usually characterized by its small size and slow growth.[6]

2. CASE REPORT

A 66-year-old female patient reported to Department of Oral Medicine and Radiology with a chief complaint of burning sensation in the mouth since one month, which was sudden in onset, continuous mild burning sensation. And the severity increases while having spicy food. There was no history of mechanical or thermal or chemical trauma. Past medical and dental history was not significant. No abnormalities found on extra oral examination. On intra oral examination a solitary linear ulcer noted in the left lingual vestibule, extending anteroposteriorly from the lingual aspect of first premolar region and extends posteriorly. The exact posterior extension was difficult to define clinically. The ulcer has a rounded and hyperplastic margins, floor is deep and covered with slough. Surrounding areas appears normal. On palpation, the ulcer is tender, base of the ulcer is indurated and it bleeds on touch. It is fixed to the underlying structures.

An OPG was advised, there were no specific findings on the OPG. Since there was no signs of bone involvement on the OPG and in order to visualize the exact extension of the lesion an MRI was performed and the report suggested a lesion measures 3.4 X 1.2 X 1.6 cm in size and which was isointense on T1 and hyperintense on T2lies lateral to sublingual gland. There were no evidence for neurovascular or osseous involvement and there was no sign of lymphadenopathy or perineural extension. An incisional biopsy was performed and the histopathology report showed possibility of two conditions includes adenoid cystic carcinoma (ACC) or adenoid squamous cell carcinoma. An immunohistochemistry (IHC) was performed using the same sample, in order to confirm the diagnosis. IHC report showed an over expression of Ki-67, CD-117 and P-53, which was diagnostic of ACC. Patient, was then referred to oncology department and underwent surgical excision of the lesion with radical neck dissection followed by radiation therapy. Currently patient is under follow-up.

3.DISCUSSION

Adenoid cystic carcinoma (ACC) is a rare malignant tumor comprises less than 1% of all head and neck carcinomas and 20-25% of all salivary gland malignancies.[1] ACC arises from the epithelial cells of mucous secreting glands.[7] It is slow growing aggressive tumor and is known for its perineural invasion.[1] The peak incidence noted in sixth and seventh decades of life, with a slight female preponderance.[8] Most ACCs arise in the minor salivary glands (60%). ACC of minor salivary gland origin occurs most frequently in the hard palate.[6]Foote and Frazell[9] were the first to describe that ACC canoccur in the major and minor salivary glands. And they also mentioned that the tumors were usually small with an incomplete capsule and will show variations in histology and that had a propensity to perineural spread.Conley and Dingman[10]described ACC as one of the most biologically destructive and unpredictable tumors of the head and neck. It has got high recurrence rate even when radical excision has been performed.ACC usually will be a localized disease at the time of diagnosis,[11] and is characterized by a wide local infiltration and the associated pain is mainly due to perineural invasion.[12]loco-regional lymphatic disease is uncommon, usually associated with late distant metastasis and local recurrence.[12]ACC is known for its late distant metastasis.[18] Lung being the commonest site of involvement, and lung involvement is reported in more than 50% of the cases.[1]

There is a strong positive correlation between the site of origin and prognosis. The more favorable prognosis with major salivary gland ACC as compared to minor salivary gland[2] and is due to chance for an earlier diagnosis due to increased accessibility. ACC of the nasal cavity and paranasal sinuses has got worse prognosis than in any other area of the head and neck.[15] Tumor staging and histological subtyping is considered as one of the most reliable indicators of overall prognosis.[14] Szanto et al.[16] introduced histopathological grading as cribriform or tubular (grade I), less than 30% solid (grade

II), or greater than 30% solid (grade III). The cribriform variant demonstrates significantly poor prognosis in terms of local recurrence rate. 1972 Eby et al[13]described that solid subtype has a worse prognosis than the other two forms with a100% possibility for recurrence.Interms of distant metastases and long-term survival.The tubular pattern has got a better prognosis.[14]

ACCs arising from sites close to the cranial base (nasopharynx, nasal cavity, and maxilla) have a significantly increased risk of local recurrence.[3] This is related to the difficulty of obtaining a clear resection margin at the cranial base because of the difficulties associated with the surgery, intracranialextension of the tumor along nerves, and restrictions on the limits of resection due to the proximity of neural and vascular structures.

The use of immunehistocytochemistry(IHC), and the staining pattern of p53, bcl-2, P-glycoprotein are useful in diagnosis and differentiation of ACC from other malignancies.[1]The p53 alteration is an independent prognostic marker and the proteins known for their radio- and chemotherapy resistance can be overexpressed in some ACCs, suggesting that those molecules could influence the outcome of new therapeutic management.[17]

The minor salivary gland tumors should be treated by local radical excision and postoperative radiotherapy.[19] ACCs arising in major salivary glands are treated surgically, followed by possible addition of adjunctive radiotherapy.The average survival rates reported for patients with surgery followed with radiotherapy is3 years 80% and 5 years 78%.[1,18]Mendenhall WM, et al[20] reported a 60% 3 year survival rate with surgery alone and 56% with radiotherapy alone. 10- to 20-year survival rates are very low with any kind of therapy.[21]Survival times after appearance of distant metastases among patients with isolated lung metastases and those with bone metastases with or without lung involvement were 54 and 21 months, respectively.[1] The size of tumor at presentation and the development of local treatment failure are the two factors most predictive of distant metastases.

4. CONCLUSION

Solitary ulcers are a common clinical presentation for a wide range of clinical conditions. The causes range from simple trauma, resistant infection to malignancies. Solitary ulcers can also be the first and only presentation of underlying systemic diseases. Accurate diagnosis is important in identifying the underlying condition and thereby reduces the morbidity in disease management.Optimal treatment of ACC has not yet been fully established.Most authors advocate the use of surgical excision and postoperative radiotherapy.Careful tumor staging and grading with documentation of perineural invasion and margin status continue to be important prognostic tools.IHC is helpful for the diagnosis confirmation and a routine chest radiographic examination is a mandatory for all the patients who are under follow up as lung is being the common location for a metastasis.

FIGURES

Figure.1:A solitary linear ulcer in the left lingual vestibule

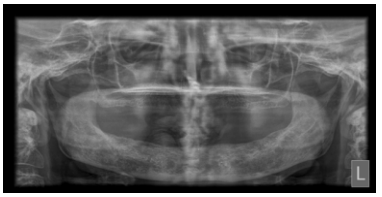

Figure.2: A Panoramic image shows no specific findings.

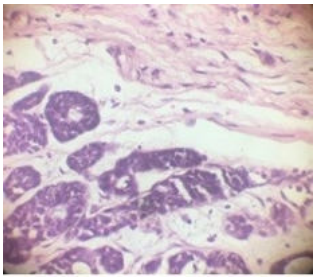

Figure.3: Photomicrograph (H & E stained) revealed numerous duct-like structures and tubules containing mucinous substance; ducts were lined by cuboidal epithelium. Various islands of hyperchromatic, basophilic, and isomorphic cells surrounded by hyalinizedstroma were seen.

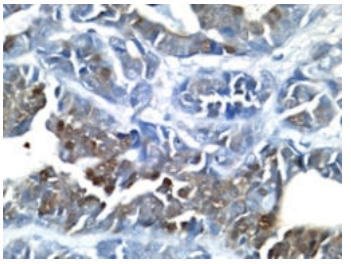

Figure.4: IHC staining pattern of p53

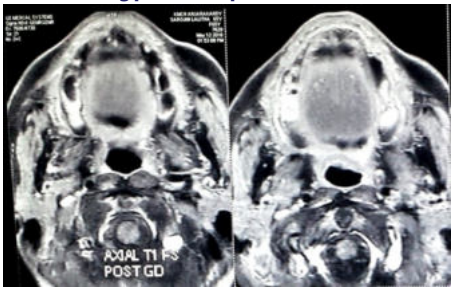

Figure.5: MRI showing Small altered signal intensity lesion which is isointense on T1 and Hyperintense on T2,Lies lateral to sublingual gland

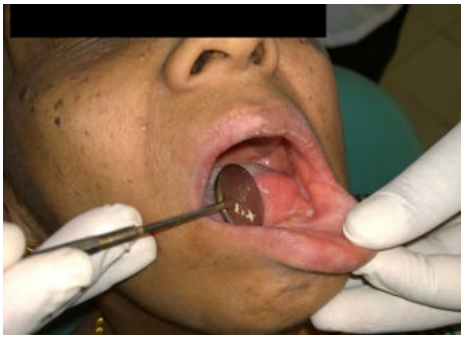

Figure.6: Postoperative follow-up after 3months

5. REFERENCES

1. S. Chummun, N. R. McLean , Adenoid cystic carcinoma of the head and neck. British Journal of Plastic Surgery (2001), 54, 476--480
2. Giannini PJ, Shetty KV, Horan SL, Reid WD, Litchmore LL. Adenoid cystic carcinoma of the buccal vestibule: a case report and review of the literature. Oral Oncol. 2006; 42:1029.
3. Khan S, Agwani K, Bhargava P, Kumar SP. Adenoid cystic carcinoma presenting as an ulcer on the floor of the mouth: a rare case report. J Korean Assoc Oral Maxillofac Surg 2014; 40: 253-7.
4. Ji-Young Song. Adenoid cystic carcinoma of the sublingual gland: A case report. Imaging Science in Dentistry 2016; 46: 291-6.
5. Chandresh Jaiswara, Neeraj Kumar Dhiman, Akhilesh Kumar Singh, Naresh Kumar Sharma, Vishal Verma, Arun Pandey. Adenoid cystic carcinoma of the floor of the mouth – A rare presentation. J Oral Biol Craniofac Res. (2015).
6. Mahajan A, Kulkarni M, Parekh M, Khan M, Shah A, Gabhane M. Adenoid cystic carcinoma of hard palate: a case report. Oral Maxillofac Pathol J. 2011;2:127.
7. J.P. Agarwal , Intraoral adenoid cystic carcinoma: Prognosticfactors and outcome.

Oral Oncology (2008) 44, 986– 993.
8. Fordice J, Kershaw C, E1-Naggar A, Goepfert H. Adenoid cystic carcinoma of the head and neck: predictors of morbidity and mortality. Arch Otolaryngol Head Neck Surg 1999; 125: 149-52.
9. Foote FW, Frazell EL. Tumours of the major salivary glands. Cancer. 1953;6:1065–1133.
10. Conley J, Dingman DL. Adenoid cystic carcinoma in the head and neck (cylindroma). Arch Otolaryngol. 1974;100:81–90.
11. Yu-Chin Lin et al , Clinicopathological features of salivary and non-salivary adenoid cystic carcinomas. Int. J. Oral Maxillofac. Surg. 2012; 41: 354–360.
12. B. Bianchi et al , Adenoid cystic carcinoma of intraoral minor salivary glands . Oral Oncology (2008) 44, 1026– 1031.
13. Eby LS, Johnson DS, Baker HW. Adenoid cystic carcinoma of the head and neck. Cancer 1972; 29: 1160-8.
14. Spiro RH, Huvos AG. Stage means more than grade in adenoid cystic carcinoma. Am J Surg 1992; 164: 623-8.
15. Howard DJ, Lund VJ. Reflections on the management of adenoid cystic carcinoma of the nasal cavity and paranasal sinuses. Otolaryngol Head Neck Surg. 1985;93: 338–341.
16. Szanto PA, Luna MA, Tortoledo ME, et al. Histologic grading of adenoid cystic carcinoma of the salivary glands. Cancer. 1984;54:1062–1069.
17. Kolude B, Adisa AO, Lawal AO, Adeyemi BF. Strong immunohistochemical expression of C-kit may characterize adenoid cystic carcinoma of the salivary gland. J Oral Maxillofac Surg Med Pathol. 2014;26(4):549–553.
18. Haddad A, Enepekides DJ, Manolidis S, Black M. Adenoid cystic carcinoma of the head and neck: a clinicopathologic study of 37 cases. J Otolaryngol 1995; 24: 201-5.
19. Bradley PJ. Submandibular gland and the minor salivary gland neoplasms. CurrOpinOtolaryngol Head Neck Surg. 1999;7:72–78.
20. Mendenhall WM, Morris CG, Amdur RJ, Werning JW, Hinerman DP, Villaret DP. Radiotherapy alone or combined with surgery for adenoid cystic carcinoma of the head and neck. Head Neck 2004;26(2):154–62.
21. Westra WH. The surgical pathology of salivary gland neoplasms. Otolaryngol Clin N Am. 1999;39:919–943.