Original Research Paper

Volume - 14 | Issue - 12 | December - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Histopathology

ADENOID CYSTIC CARCINOMA OF MAXILLARY SINUS:A RARE CASE REPORT

Dr.Gaurangi G. Velip	Senior Resident in Department of Pathology Goa Medical College
Dr. Premila De Souza Rocha	Associate Professor in Department of Pathology Goa Medical College
Dr. Samiksha Naik Talaulikar*	Senior Resident Department of Surgery Goa Medical College *Corresponding Author
Dr.Sunidhi Vernekar	Senior Resident in Department of ENT Goa Medical College
Dr. Shubhra S. Ghadi Amonkar	Junior Resident in Department of Pathology Goa Medical College

ABSTRACT Adenoid Cystic Carcinoma of the maxillary sinus is a rare tumour. It is a slow growing,aggressive,with high rates of local and distal recurrence with a overall poor prognosis[1].Thus causing a considerable diagnostic challenge since the symptoms mimic more common ailments such as sinusitis or allergic rhinitis,particularly in the early stage.We report a case of a 35 year old female who presented to opd with proptosis and facial swelling.The clinical,radiological and histopathological examination found a mass compatible with adenoid cystic carcinoma of maxillary sinus.

KEYWORDS : Adenoid cystic carcinoma,Maxillary sinus

INTRODUCTION

Adenoid Cystic Carcinoma is a carcinoma of primary salivary gland or minor salivary gland of upper aerodigestive tract. It is characterized by the biphasic ductal and myoepithelial differentiation.It is an aggressive tumour with a high propensity for local and distal recurrence. Adenoid cystic carcinoma of the maxillary sinus is a rare tumour with a poor overall long term prognosis[1][2]

CASE PRESENTATION

A 35 year old female presented to opd with complaints of watering and proptosis of left eye and left side facial swelling since 4 months. A facial CT and MRI scan revealed a soft tissue mass epicentered in the left maxillary sinus causing destruction of the floor of the left orbit ,anterior and medial wall of the maxillary sinus with extension into the soft tissues of the left cheek.Biopsy of the mass was done which revealed adenoid cystic carcinoma. Later, the patient underwent a total maxillectomy surgery.

Grossly, the total maxillectomy specimen showed the erosion of the anterior,medial and lateral walls of the maxillary sinus.The brownish white tumoral mass filling the maxillary cavity measured 6.5x4.5x4.5 cm in size. Cut section revealed whitish homogenous appearance.No areas of necrosis or hemorrhage were seen.

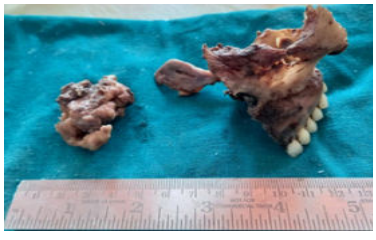


Figure 1: Total maxillectomy specimen along with the irregular brownish white tumoral mass

Figure 2: Cut section of the tumoral mass revealing whitish homogenous appearance

Microscopically,the tumoral mass revealed a cellular neoplasm with a mixed tubular and cribriform architecture, composed of small hyperchromatic cells with mild pleomorphism and high nuclear to cytoplasmic ratio .Perineural invasion was seen.No solid areas were noted.Mitosis was 5/10hpf with no nuclear atypia.The tumour was seen infiltrating the bony trabeculae and the nasal mucosa. All the features were compatible with diagnosis of adenoid cystic carcinoma of maxillary sinus.

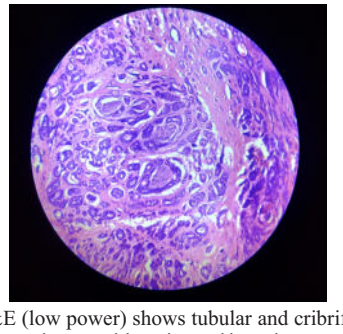


Figure 3:H &E (low power) shows tubular and cribriform pattern of adenoid cystic carcinoma with perineural invasion

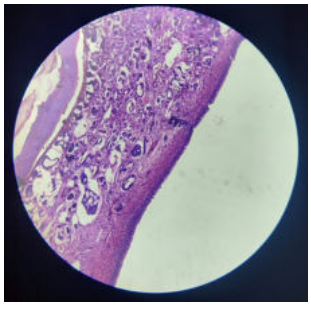
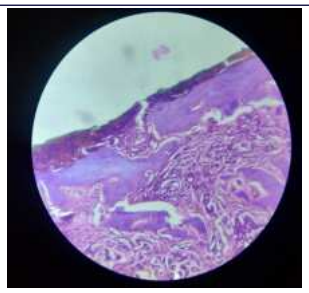


Figure 4:H&E (low power) showing adenoid cystic carcinoma infiltrating the nasal mucosa

Figure5 :H&E(low power) showing adenoid cystic carcinoma infiltrating the bony trabeculae of maxillary sinus



DISCUSSION

Adenoid Cystic Carcinoma is a Carcinoma of the secretory glands such as the major salivary glands,minor salivary glands,ceruminous and lacrimal glands[2].The most commonly affected is the parotid gland,among the major salivary glands.Whereas,the minor salivary glands,which are affected include those of oral cavity,sinonasal tract,nasopharynx,oropharynx and trachea.There is no gender predilection and the mean age of occurrence is 57 years[1].The most common site for the distant metastasis is lung.[1]

Maxillary sinus is an unusual site of occurrence for the adenoid cystic carcinoma,where it constitutes 0.3-1% of all sinonasal tumours[2]. Maxillary sinus adenoid cystic carcinoma(MSACC) is thought to originate from the seromucinous glands which are found in the respiratory epithelial mucosa lining the nasal cavity and the paranasal sinuses[3].MSACC is slowly growing tumour,locally aggressive with high rates of local and distant recurrence.MSACC behaves more aggressively as compared to its counterparts in the major and minor salivary glands.This is due to the fact that these tumours can remain in the submucosa of the maxillary sinus or spread in sheets below the periosteum without any signs or symptoms.Since the nasal cavity and the maxillary sinus are air filled spaces,the tumour first extends into the air space.[4][5]

Clinical features of MSACC are non specific such as nasal obstruction,local pain and epistaxis.Thus often misdiagnosed as sinusitis or allergic rhinitis which leads to the delay in the appropriate treatment.As the tumour progresses ,it invades the bone and the adjacent structures such as orbit,meninges,base of cranium etc leading to serious consequences such as opthalmoplegia,proptosis and seizures[4]

CT scan and MRI are the mainstay for the diagnosis of MSACC[6]. PET CT scan is of limited value,since the FDG uptake by these tumours is low[7].

Grossly,ACC appears as a infitrative and ill defined ,firm mass.

Histologically,Adenoid cystic carcinoma is a biphasic tumour ,composed of ductal and myoepithelial cells.The ductal cells are cuboidal with eosinophilic cytoplasm.Whereas,the myoepithelial cells have dark angulated nuclei and scanty cytoplasm giving a basaloid appearance[1].

ACC has three patterns:Tubular,cribriform and solid pattern.
The tubular pattern contains simple tubules lined by inner ductal and outer myoepithelial cells.Cribriform pattern is made up of predominantly myoepithelial cells with myxoid or hyalinized globules.Solid pattern are solid nests made up of basaloid cells.Some studies have shown that the presence of the solid growth is a predictor of the adverse outcome.

Adenoid cystic carcinoma has a high propensity for perineural invasion[1].

The features of the high grade transformation of ACC are:marked nuclear atypia,increased mitosis(>10/10hpf) and comedo necrosis[8].

ACC can be misdiagnosed with other malignant tumours such as epithelial myoepithelial carcinoma,basal cell adenocarcinoma, polymorphous adenocarcinoma, basaloid squamous cell carcinoma and HPV related sinonasal carcinoma.

To differentiate ACC from other tumours,the use of IHC is a reliable approach.CK7,CAM5.2 (ductal component),CD117,alpha SMA and S100 show a positive expression for ACC.While,Vimentin gives a negative expression[9]

60-90% of ACC have characteristic and diagnostic fusions involving MYB,MYBL1 or NFIB gene[10].

The choice of the treatment depends upon the grade ,stage ,site and behaviour of the tumour.Mostly Curative surgery and adjuvant radiotherapy is considered as the mainstay of treatment by most authors.

Due the limitations of surgery ,majority of institutions have implemented the use of radiotherapy[11].However, it was noted that 96% of the tumours responded to radiation but the recurrence rate after radiotherapy was 94%[12][13].This shows that adenoid cystic carcinoma is radiosensitive but not radiocurable.Chemotherapy has a little role in ACC since the tumour cells have low mitotic index[6].Many studies have reported that the rate of distant metastasis was approximately 40% in sinonasal ACC patients[14].

CONCLUSION

Maxillary sinus adenoid cystic carcinoma is a rare tumour with an overall poor long term prognosis,compared to other Adenoid cystic carcinoma of head and neck.It is characterised by slow growth and tendency for recurrence which makes it difficult for diagnosis and treatment.

Thus,MSACC should be considered in the differential diagnosis in patients with slowly growing swelling or fleshy mass in and around the nose, even if it is rare.

REFERENCES

1) van Weert S, Reinhard R, Bloemena E, Buter J, Witte BI, Vergeer MR, Leemans CR. Differences in patterns of survival in metastatic adenoid cystic carcinoma of the head and neck. Head Neck. 2017 Mar;39(3):456-463. doi: 10.1002/hed.24613. Epub 2016 Oct 24. PMID: 27775851.
2) Rhee CS, Won TB, Lee CH, Min YG, Sung MW, Kim KH, Shim WS, Kim YM, Kim JW. Adenoid cystic carcinoma of the sinonasal tract: treatment results. Laryngoscope. 2006 Jun;116(6):982-6. doi: 10.1097/01.mlg.0000216900.03188.48. PMID: 16735899.
3) Li Y, Hao D, Song X, Zhang C. Computed tomography and magnetic resonance imaging of adenoid cystic carcinoma in the maxillary sinus: a retrospective study with radiologic-histopathologic correlations. Oral Surg Oral Med Oral Pathol Oral Radiol. 2021 Jan;131(1):111-121. doi: 10.1016/j.oooo.2020.06.019. Epub 2020 Jul 4. PMID: 32712057.
4) Andrade MF, de Faria PR, Cardoso SV, Santos MR, Dias FL, Eisenberg AL, Santos IC, Loyola AM. Adenoid cystic carcinoma of the maxillary sinus: a clinical-pathological report of 10 years of experience from a single institution. Int J Oral Maxillofac Surg. 2014 Nov;43(11):1313-8. doi: 10.1016/j.ijom.2014.06.016. Epub 2014 Jul 31. PMID: 25088528.
5) Kim GE, Park HC, Keum KC, Lee CG, Suh CO, Hur WJ, Kim KM, Hong WP. Adenoid cystic carcinoma of the maxillary antrum. Am J Otolaryngol. 1999 Mar-Apr;20(2):77-84. doi: 10.1016/s0196-0709(99)90015-7. PMID: 10203156.
6) Gill KS, Frattali MA. An Unusual Presentation of Adenoid Cystic Carcinoma. Case Rep Otolaryngol. 2015;2015:826436. doi: 10.1155/2015/826436. Epub 2015 Dec 24. PMID: 26819792; PMCID: PMC4706871.
7) Jeong HS, Chung MK, Son YI, Choi JY, Kim HJ, Ko YH, Baek CH. Role of 18F-FDG PET/CT in management of high-grade salivary gland malignancies. J Nucl Med. 2007 Aug;48(8):1237-44. doi: 10.2967/jnumed.107.041350. Epub 2007 Jul 13. PMID: 17631549.
8) Seethala RR, Hunt JL, Baloch ZW, Livolsi VA, Leon Barnes E. Adenoid cystic carcinoma with high-grade transformation: a report of 11 cases and a review of the literature. Am J Surg Pathol. 2007 Nov;31(11):1683-94. doi: 10.1097/PAS.0b013e3180dc928c. PMID: 18059225.
9) Husain Q, Kanumuri VV, Svider PF, Radvansky BM, Boghani Z, Liu JK, Eloy JA. Sinonasal adenoid cystic carcinoma: systematic review of survival and treatment strategies. Otolaryngol Head Neck Surg. 2013 Jan;148(1):29-39. doi: 10.1177/0194599812464020. Epub 2012 Oct 12. PMID: 23064210.
10) Brayer KJ, Frerich CA, Kang H, Ness SA. Recurrent Fusions in MYB and MYBL1 Define a Common, Transcription Factor-Driven Oncogenic Pathway in Salivary Gland Adenoid Cystic Carcinoma. Cancer Discov. 2016 Feb;6(2):176-87. doi: 10.1158/2159-8290.CD-15-0859. Epub 2015 Dec 2. PMID: 26631070; PMCID: PMC4744535.
11) Unsal AA, Chung SY, Zhou AH, Baredes S, Eloy JA. Sinonasal adenoid cystic carcinoma: a population-based analysis of 694 cases. Int Forum Allergy Rhinol. 2017 Mar;7(3):312-320. doi: 10.1002/alr.21875. Epub 2016 Nov 11. PMID: 27863150.
12) Vikram B, Strong EW, Shah JP, Spiro RH. Radiation therapy in adenoid-cystic carcinoma. Int J Radiat Oncol Biol Phys. 1984 Feb;10(2):221-3. doi: 10.1016/0360-3016(84)90007-5. PMID: 6323361.
13) Spiers AS, Esseltine DL, Ruckdeschel JC, Davies JN, Horton J. Metastatic Adenoid Cystic Carcinoma of Salivary Glands: Case Reports and Review of the Literature. Cancer Control. 1996 Jul;3(4):336-342. doi: 10.1177/107327489600300405. PMID: 10765225.
14) Seong SY, Hyun DW, Kim YS, Cho HJ, Lee JG, Yoon JH, Kim CH. Treatment outcomes of sinonasal adenoid cystic carcinoma: 30 cases from a single institution. J Craniomaxillofac Surg. 2014 Jul;42(5):e171-5. doi: 10.1016/j.jcms.2013.08.002. Epub 2013 Sep 9. PMID: 24080140.