

Recurrent adenoid cystic carcinoma of paranasal sinuses: A rare case report

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Abstract

Adenoid cystic carcinoma (ACC) is a distinct salivary gland neoplasm that predominantly occurs in the minor salivary gland. ACC is an uncommon tumor that progress slowly affecting 5% of paranasal sinuses with high distant metastasis rate in advance stages. This salivary gland tumor is difficult to diagnose both clinically and histopathologically due to its indolent presentation and diverse histopathological patterns. Hence, immunohistochemistry plays a decisive role in diagnosing this tumor. A recurrent case of ACC of paranasal sinuses in the maxillary sinus of a 41-year-old female patient without any extraoral abnormality is reported.

Keywords: Adenoid cystic carcinoma, immunohistochemical analysis, paranasal sinuses, recurrence

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INTRODUCTION

Adenoid cystic carcinomas (ACCs) are rare malignant tumors that usually affect salivary glands.^[1] ACC was first described by Robin, Lorian and Laboulbene in two of their articles published in 1853 and 1854, in which the cylindrical appearance of the tumor was discussed. Billroth, in 1859, has described the tumor as cylindroma and has also explained its great recurrent tendency. Spies in 1930 gave the name “ACC” in his discussion of cutaneous and noncutaneous tumors of the basal cell type. They comprise about 1% malignancies of all head and neck tumors and 6%–10% of salivary gland tumors.^[2] It affects both major and minor salivary glands accounting for about 15%–30% of submandibular gland, 50% of minor salivary glands and 2%–15% of parotid gland.^[3] Although there is no gender predilection, slight female predominance is reported.^[4] Usually, ACC occurs in 5th to

7th decade of life.^[5] The palate is the most common site of intraoral lesions.^[6] The most prognostic predictive factors are tumor size, grade, stage, margin status, lymph node involvement and perineural invasion. ACC rarely affects the paranasal sinuses (5%), among which maxillary sinus is most frequently affected for about (46%–63%), followed by the nasal cavity (20%), ethmoidal complex (10%), sphenoid sinus (5%), nasopharynx (20%) and frontal sinus (1%).^[7] The mean survival rate for 5, 10 and 15 years is 84.7%, 70.8% and 34%, respectively.^[8] Herein, we report a case of recurrent ACC in paranasal sinuses with an uncommon clinical presentation.

CASE REPORT

A 41-year-old female patient came to the outpatient department of our institution with a chief complaint of pain over the upper right side of the face and with

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Figure 1: Orthopantomogram-hyperdense areas seen in the entire right maxillary sinus

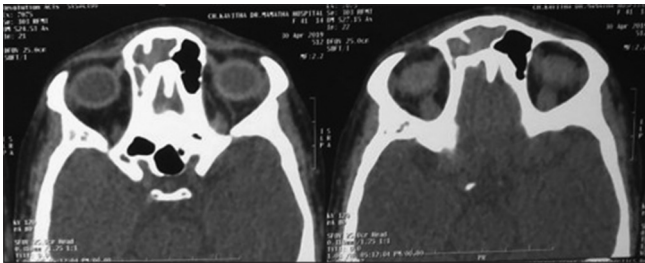


Figure 3: Computed tomography scan- irregular soft-tissue density lesion in the right entire maxillary sinus and nasal cavity extending to ethmoid and frontal sinus

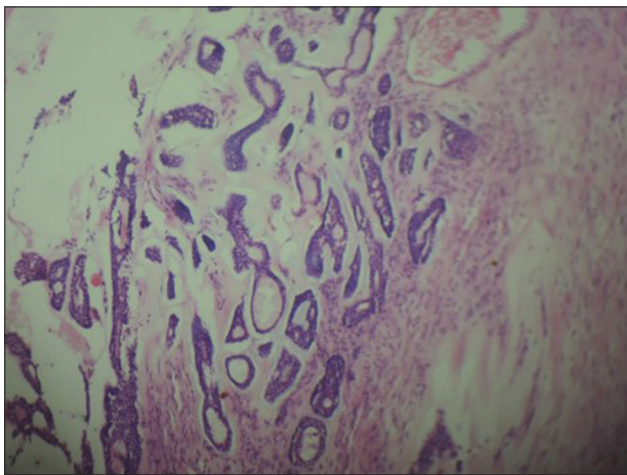


Figure 5: Cords and tubular areas. H&E, $\times 10$

nasal obstruction since 20 days. The patient medical history revealed a previous surgery for ACC in the right maxillary sinus 10 years back. The extraoral examination was within normal limits. Intraoral examination revealed mild tenderness over the upper right buccal vestibule. Orthopantomogram revealed hyperdense areas filling the entire right maxillary sinus [Figure 1] and contrast-enhanced computed tomography revealed irregular soft-tissue density lesion in the right maxillary sinus extending to ethmoid and frontal sinus, right nasal cavity showing heterogeneous enhancement in postcontrast series.[Figures 2 and 3] Based

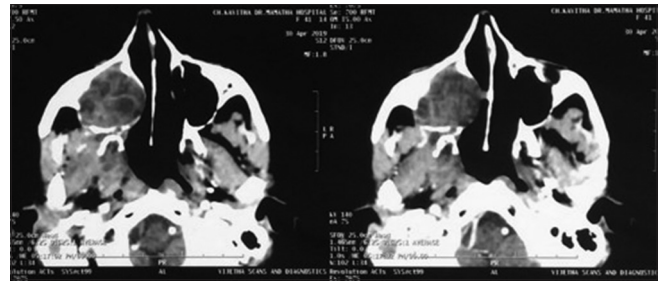


Figure 2: Computed tomography scan- irregular soft-tissue density lesion in the right entire maxillary sinus and nasal cavity



Figure 4: Gross picture- incisional biopsy- whitish brown in color, measuring about 1 cm \times 1.5 cm in size, soft in consistency, roughly trapezoidal in shape with smooth surface and ill-defined borders

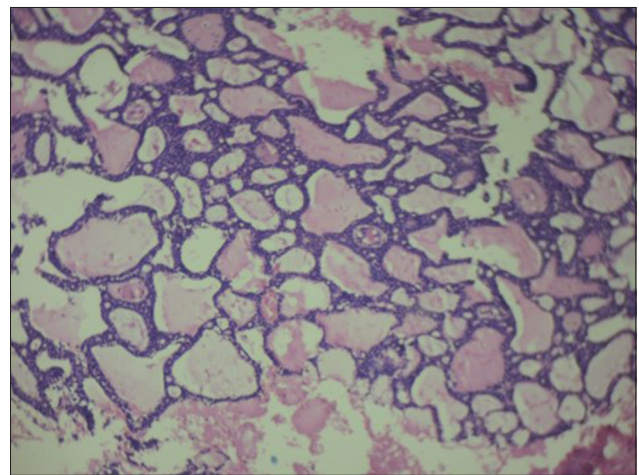


Figure 6: Swiss cheese pattern. H&E, $\times 40$

on clinical and radiographical features, the provisional diagnosis is suspected as malignancy of paranasal sinuses.

Incisional biopsy done in relation to the right maxillary sinus through Caldwell-Luc approach and specimen was sent for histopathological examination. Gross examination revealed three soft-tissue specimens, which are whitish brown in color, measuring about 1 cm \times 1.5 cm in size, soft

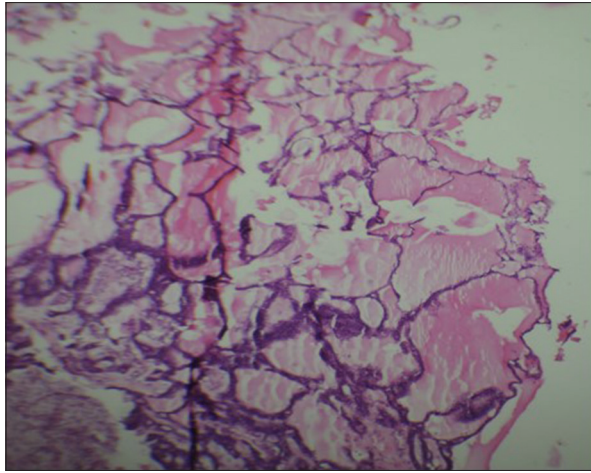


Figure 7: Cystic spaces with hyalinization. H&E, ×10

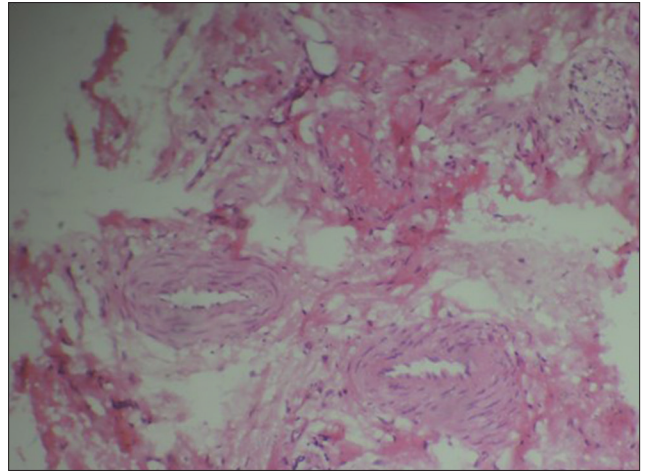


Figure 8: Perineural invasion. H&E, ×10

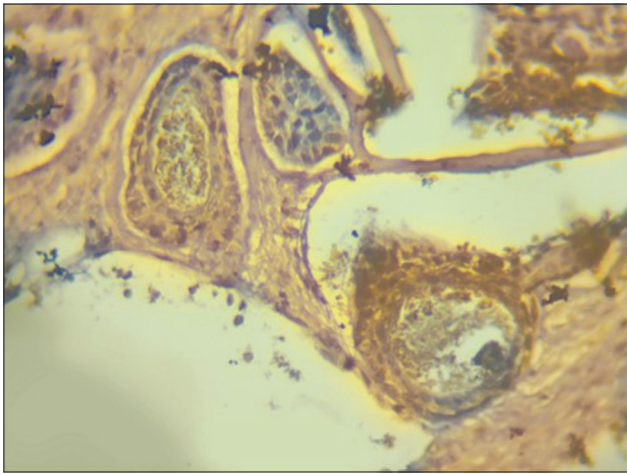


Figure 9: CD117. IHC, ×40 positive

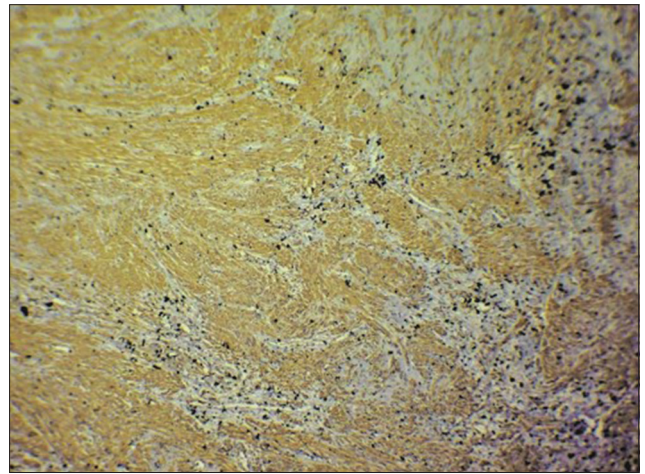


Figure 10: SMA. IHC, ×40 positive

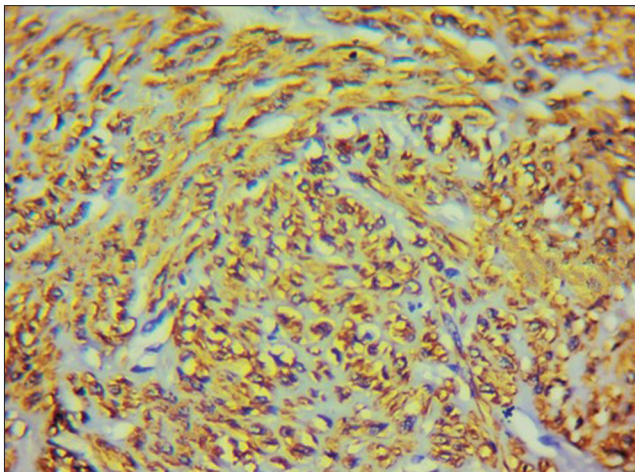


Figure 11: S100. IHC, X40 positive

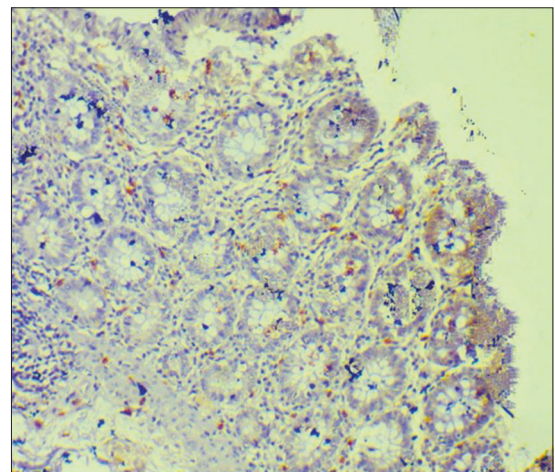


Figure 12: Vimentin. IHC, X40 negative

in consistency, roughly trapezoidal in shape with smooth surface and ill-defined borders [Figure 4]. Microscopically, the lesion showed cystic spaces surrounded by basophilic uniformly sized cuboidal cells showing hyperchromatism. The cystic spaces contain eosinophilic with hyalinization.

Focal areas also show cords and tubular areas of tumor cells with perineural invasion. Loose connective tissue stroma is also noted [Figures 5-8]. Based on histopathological features, the diagnosis is made as ACC of paranasal sinuses.

For confirmatory diagnosis, immunohistochemistry (IHC) staining done by markers such as CD117, alpha-smooth muscle actin, S100, Vimentin. Immunohistochemistry being the benchmark. The tumor cells showed strong positivity for CD117, alpha-smooth muscle actin, S100 and negative for Vimentin [Figures 9-12]. Based on clinical, radiographical, histopathological and immunohistochemical evaluation, a confirmatory diagnosis of recurrent ACC of paranasal sinuses was made and the patient was referred to higher centers for further treatment and management.

DISCUSSION

ACC is a rare epithelial tumor originating from mucus-secreting glands of the upper aerodigestive tract, which accounts for about 1% of all malignant neoplasms of the oral cavity and maxillofacial area.^[9] ACCs are the second-most common type of carcinoma arising in the salivary glands, following mucoepidermoid carcinoma.^[10] It mostly affects the minor salivary glands.

Clinically, ACC frequently appears as a small slow-growing lesion with indolence behavior but it is often discovered at an advanced stage.^[5] Based on tumor location, patients have variable clinical symptoms and signs, but the pain is a common clinical finding due to an early perineural invasion of neoplastic cells.^[10] It is a well-known fact that the pain arising from nondental causes might be confused as pain arising from the tooth, which leads to misdiagnosis. Park *et al.* reported an ACC of maxillary sinus misdiagnosed as chronic apical periodontitis.^[11] Therefore, pain associated to salivary gland malignant tumor may be misdiagnosed as pain from odontogenic origin. In a recent study of 4004 periapical lesions which were diagnosed as endodontically associated pathoses, nine were found to be malignant processes, seven of which were squamous cell carcinoma, one Langerhans cell histiocytosis and one ACC located in the mandible.^[12]

Histologically, ACC is composed by ductal and modified myoepithelial cells that typically have hyperchromatic, angular nuclei and frequently clear cytoplasm, organized in classic cribriform, tubular and solid patterns. The tumor is graded by the pattern of neoplastic cells arrangements in tubular, cribriform, solid nests and cords and solid type of ACC shows aggressive behavior with high recurrence rate and lymph nodes metastasis and most patients present at an advanced stage. The relationship between the histological pattern and the prognosis was suggested by Soares *et al.*^[5] According to Batsakis *et al.*, the survival rate was less in the solid pattern when compared with the cribriform and tubular patterns.^[13]

ACC can be misdiagnosed with other malignant neoplasms such as polymorphous adenocarcinoma (PAC), basaloid squamous cell carcinoma (BSCC), mucoepidermoid carcinoma (MEC) and benign tumors such as pleomorphic adenoma (PA) and basal cell adenoma (BCA). Immunohistochemical staining is a reliable approach to differentiate ACC from other tumors. IHC markers such as CD117, alpha smooth muscle actin, and S100 show a positive expression for ACC whereas vimentin gives a negative expression.

ACC and BSCC show cribriform and solid areas. The basement membrane material secreted by BSCC tends to dissect between tumor cells rather than to form cribriform spaces, as seen in ACC. Necrosis and basaloid cells with prominent nucleoli and coarse chromatin can be seen in both. However, BSCC shows few contrasting features such as single-cell necrosis, a brisk mitotic rate and greater degree of nuclear atypia. Unlike ACC, BSCC also shows foci of keratinization, attachment of rete pegs and presence of a surface squamous dysplasia. In addition, true lumina and mucosal carcinoma *in situ* changes are only seen in ACC. Few IHC markers help to differentiate ACC from BSCC. CD117 and alpha-smooth muscle actin were negative for BSCC, whereas strongly positive for ACC.

PAC occurs almost exclusively in the minor salivary glands and may contain overlapping histopathologic features with ACC. Both have common cell types and epithelial-myoepithelial relationship arranged in similar patterns (tubular, cribriform and solid), tendency for local infiltration and perineural spread. However, polymorphous architectural characteristics and foci of papillary growth and areas of single file single-cell infiltration are seen in PAC but not in ACC and typically has negative or low (<50% of cells) expression of CD 117 and alpha-smooth muscle actin compared with ACC and positive expression for vimentin, whereas ACC shows a negative expression. Other differential diagnoses can be PA, as sharply punched out; cribriform type glands are seen in both tumors. PA shows double-layered duct-like structure with an inner layer of dark, cuboidal cells and outer layer of clear cells which are seen in tubular variant of ACC. However, few contrasting features such as the presence of infiltration and perineural invasion are not seen in PA. Moreover, squamous metaplasia or chondroid metaplasia is seen only in PA and positive expression for vimentin and weak expression for CD117 helps to rule out ACC from PAC.

ACC can also be misdiagnosed as basal cell adenoma (BCA) because BCA may have areas mimicking ACC. However, it is encapsulated, does not show any invasive growth pattern

and lacks perineural invasion. In addition, the distinctive basosquamous whorls of BCA are helpful as squamous differentiation is not actually encountered in ACC. In addition, BCA shows a negative expression for CD117.

MECs and ACCs are two tumors which may create diagnostic confusion due to overlapping morphologic features, including cystic changes and mucinous secretions. Few IHC markers such as CD117 expressed weakly positive and negative for alpha-smooth muscle actin and S100 by mucoepidermoid carcinoma, which are contrasting expressed in ACC.

The current standard of care for head and neck ACC is complete surgical resection, and due to its high recurrence rates adjuvant radiotherapy is recommended. Usually, ACC shows distant metastasis to lung and bone.^[14] However, metastasis rates of ACC are lower and associated with poor prognosis in paranasal sinuses when compared to other head and neck sites.

CONCLUSION

ACC is a malignant tumor with indolent growth pattern and characterized by late high recurrence rate. Regardless of the benign histopathological appearance of ACC, it also shows the potential ability of local invasion with distant metastasis; however, the regional lymphatic spread is uncommon. Hence, IHC plays a major role in differentiating ACC from other tumors. Once ACC of the primary site presents recurrence, the prognosis is generally poor. The main treatment protocol for aggressive ACC is surgery with free surgical margins followed by adjuvant radiotherapy to reduce recurrence rates. This case report highlights the fact that although the oral cavity finding might be without any visible lesions, sharp unilateral pain might suggest underlying malignancy in the affected area as it was seen in this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bruzgielewicz A, Osuch-Wójcikiewicz E, Majszyk D, Chęciński P, Nyckowska J, Zawadzka B, *et al.* Adenoid cystic carcinoma of the head and neck – A 10 years experience. *Otolaryngol Pol* 2011;65:6-11.
2. Michel G, Joubert M, Delemazure AS, Espitalier F, Durand N, Malard O. Adenoid cystic carcinoma of the paranasal sinuses: Retrospective series and review of the literature. *Eur Ann Otorhinolaryngol Head Neck Dis* 2013;130:257-62.
3. Ali S, Palmer FL, Katabi N, Lee N, Shah JP, Patel SG, *et al.* Long-term local control rates of patients with adenoid cystic carcinoma of the head and neck managed by surgery and postoperative radiation. *Laryngoscope* 2017;127:2265-9.
4. Martínez-Rodríguez N, Leco-Berrocal I, Rubio-Alonso L, Arias-Irímia O, Martínez-González JM. Epidemiology and treatment of adenoid cystic carcinoma of the minor salivary glands: A meta-analytic study. *Med Oral Patol Oral Cir Bucal* 2011;16:e884-9.
5. Soares EC, Carreiro Filho FP, Costa FW, Vieira AC, Alves AP. Adenoid cystic carcinoma of the tongue: Case report and literature review. *Med Oral Patol Oral Cir Bucal* 2008;13:E475-8.
6. DeAngelis AF, Tsui A, Wiesenfeld D, Chandu A. Outcomes of patients with adenoid cystic carcinoma of the minor salivary glands. *Int J Oral Maxillofac Surg* 2011;40:710-4.
7. Lupinetti AD, Roberts DB, Williams MD, Kupferman ME, Rosenthal DI, Demonte F, *et al.* Sinonasal adenoid cystic carcinoma: The M. D. Anderson Cancer Center experience. *Cancer* 2007;110:2726-31.
8. Ouyang DQ, Liang LZ, Zheng GS, Ke ZF, Weng DS, Yang WF, *et al.* Risk factors and prognosis for salivary gland adenoid cystic carcinoma in southern china: A 25-year retrospective study. *Medicine (Baltimore)* 2017;96:e5964.
9. He S, Li P, Zhong Q, Hou L, Yu Z, Huang Z, *et al.* Clinicopathologic and prognostic factors in adenoid cystic carcinoma of head and neck minor salivary glands: A clinical analysis of 130 cases. *Am J Otolaryngol* 2017;38:157-62.
10. Ellington CL, Goodman M, Kono SA, Grist W, Wadsworth T, Chen AY, *et al.* Adenoid cystic carcinoma of the head and neck: Incidence and survival trends based on 1973-2007 Surveillance, Epidemiology, and End Results data. *Cancer* 2012;118:4444-51.
11. Park SY, Pi CY, Kim E, Lee Y. Adenoid Cystic Carcinoma of Maxillary Sinus Misdiagnosed as Chronic Apical Periodontitis. *J Oral Maxillofac Surg* 2017;75:1303.e1-000000.
12. Huang HY, Chen YK, Ko EC, Chuang FH, Chen PH, Chen CY, *et al.* Retrospective analysis of nonendodontic periapical lesions misdiagnosed as endodontic apical periodontitis lesions in a population of Taiwanese patients. *Clin Oral Investig* 2017;21:2077-82.
13. Batsakis JG, Luna MA, el-Naggar A. Histopathologic grading of salivary gland neoplasms: III. Adenoid cystic carcinomas. *Ann Otol Rhinol Laryngol* 1990;99:1007-9.
14. Bhayani MK, Yener M, El-Naggar A, Garden A, Hanna EY, Weber RS, *et al.* Prognosis and risk factors for early-stage adenoid cystic carcinoma of the major salivary glands. *Cancer* 2012;118:2872-8.