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Advanced adenoid cystic carcinoma of maxillary sinus: Rare case report and review of literature

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ABSTRACT

INTRODUCTION: Maxillary sinus adenoid cystic carcinoma (MSACC) is a rare malignancy with a propensity for distant metastasis**CASE PRESENTATION:** We report a case of a 55 years-old male who was admitted to our department with a complaint of right nasal obstruction and anosmia. Clinical examination, radiological investigations and histopathological examination found a mass compatible with advanced adenoid cystic carcinoma of the right maxillary sinus. Treatment consisted of radio chemotherapy.**CONCLUSION:** Adenoid cystic carcinoma of the maxillary sinus may represent a serious diagnostic challenge. It should be considered in the differential diagnosis of Sino nasal tumors even if it's rare.© 2021 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Adenoid cystic carcinoma (ACC) is a rare malignant epithelial tumor, described at first in 1859 as “cylindroma” by Bill Roth [1]. It's common in minor salivary glands around the mouth, uncommon in parotids and rare in paranasal Sinuses and nose [2].

This salivary gland tumor is hard to diagnose both clinically and histopathologically due to its indolent presentation and various histopathological patterns [3].

Characteristically it shows a very slow growth pattern, a high local recurrence rate, and distant metastases mainly to the lungs and bones [4].

The following case describes adenoid cystic carcinoma of the right maxillary sinus in a 55-year-old man whose diagnosis was delayed due to the indolence of his symptoms. This work is reported by following the surgical case report (SCARE) guidelines [5].

2. Case presentation

A 55-year-old male presented to the ENT department with the main complaints of right nasal obstruction and anosmia for 8 months who was treated at first as allergic rhinitis.

The medical history found no pharmacological allergies, no psychosocial problems including drug, smoking, no family genetic disease, and specially no history or family history of carcinoma.

Clinical examination revealed a subcutaneous swelling located in the anterolateral aspect of the right maxillary sinus, this tumefaction is fixed and of firm consistency. On the anterior rhinoscopy, we found a budding mass filling the right nasal cavity. Cervical lymph nodes were not enlarged or palpable on both sides.

A facial computed tomography (CT) scan showed the presence of a tumor process at the expense of the upper right maxillary bone invading the sphenoidal bone with maxillary and sphenoidal bone lysis as well as the right maxillary sinus, the right retro-ocular orbit, the cavum posteriorly and the right nasal fossa and the right ethmoidal cells. this process is in favor of a malignant process of maxillary localization (Fig.1)

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

A biopsy was taken from the mass under local anesthesia performed by a resident with 4 years of specialized training and on Histopathological examination showed undifferentiated tumor proliferation infiltrating the deep chorion of the right nasal mucosa and evoking either a cylindroma or a neuroendocrine-type carcinoma.

For confirmatory diagnosis, an immunohistochemical (IHC) (Fig.2) study was performed showing cytokeratin 7 positivity and PS 100 negativity. Based on clinical, radiographic and histopathological evaluation, a diagnosis of adenoid cystic carcinoma of the right maxillary sinus was made.

The patient was deemed inoperable due to the invasion of the infratemporal fossa

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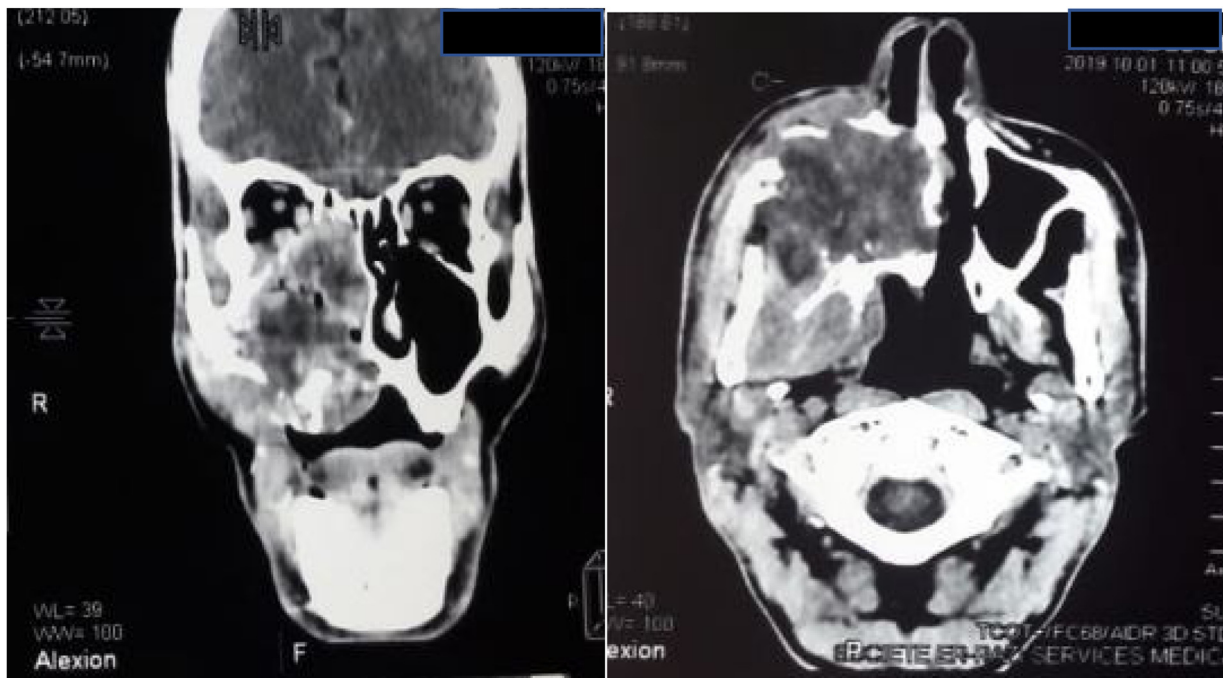


Fig. 1. axial and coronal view of facial CT Scan showing the extension of tumor.

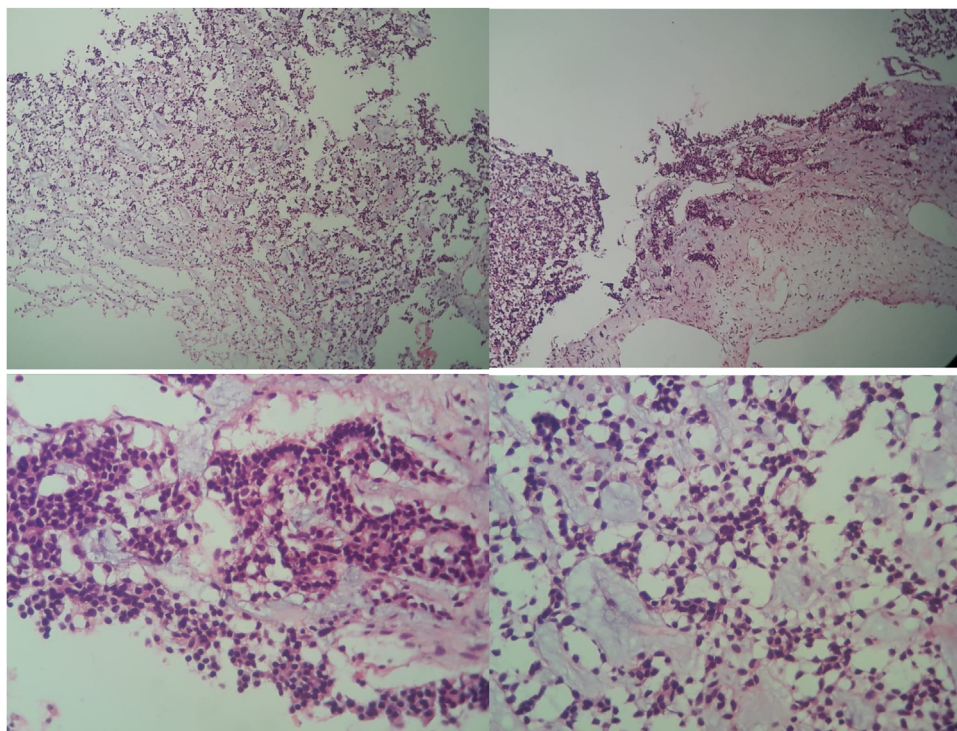


Fig. 2. Representative area of tumor showing infiltrating adenoid cystic carcinoma (Hematoxylin-eosin stain, x 10 and 40).

Because of these results, the patient was referred then to the department of Oncology for evaluation whose decision was to start radio chemotherapy.

The patient received several sessions of radiotherapy and chemotherapy based on Adriamycin, 5 FU and cisplatin.

The patient was followed in ENT and Ophthalmology consultation every 15 days for the first two months then once a month.

The patient died 13 months later with generalized metastases mainly in the lung.

3. Discussion

ACC is a neoplasm of secretory glands, such as the minor salivary, major salivary, ceruminous, and lacrimal glands. It is unusual in the maxillary sinus, where it constitutes only 0.3%–1.0% of all sinonasal tumors [6]. Maxillary sinus ACC is thought to originate from the minor seromucous glands that are found in the mucosa, underlying the respiratory epithelium in the nasal cavity and paranasal sinuses [1].

ACCs affect both men and women without any gender predilection, particularly in their fifth to seventh decades of life [7]. Although, the study of Gill and Frattali reported slight female predominance [8].

In the literature, MSACC is described as exhibiting slow growth insidiously into the sinus cavity, locally aggressive and relentless progression. However, it behaves much more aggressively than its counterparts in the major and minor salivary glands [7,9]. This growth pattern has been attributed to the fact that these tumors can remain in the submucosa of the maxillary sinus or spread in sheets below the periosteum without any symptoms or gross physical signs [9]. It also resulted from the structure of the nasal cavity and the para nasal sinuses. As the maxillary sinus and nasal cavity are air-filled spaces, the lesion first extends into the air space [1].

Additionally, this apparently indolent growth can lead to highly nonspecific signs and symptoms such as nasal obstruction, local pain, and epistaxis, and be misdiagnosed as sinusitis or allergic rhinitis, delaying correct diagnosis and appropriate treatment like our patient [7]. As the tumor progresses, it invades the surrounding bone and adjacent structures (orbit, pterygomaxillary fossa, meninges, and base of the cranium), causing more serious signs and symptoms like ophthalmoplegia, ophthalmalgia, ocular proptosis, headache, otalgia, and seizures [7].

CT scan or MRI are the mainstay of the diagnosis of ACC. The first one creates a 3D image helping the surgeon to project the surgical procedure and the second one is useful for detecting perineural invasion [8]. However, ACCs can show up as a benign or malignant process on imaging. Since the MR results of ACCs are not specific, histopathological examination should ensure a correct diagnosis. On T2-weighted images, lesions with hypointensity corresponded to highly cellular tumors (solid subtype) while lesions with hyperintensity were less cellular tumors (cribriform or tubular subtype) [10].

The diagnostic value of PET / CT 18F-fluorodeoxyglucose (FDG) for ACCs is limited due to metabolic activity, and therefore FDG uptake by these tumors is low [11].

Histologically, ACC is divided into a more differentiated cribriform type (oval islands of small, darkly staining cells with minimal cytoplasm) and a less differentiated solid form (hyperchromatic cells with indistinct cell borders and high mitotic activity). The cribriform type is further subdivided into the tubular type (cords of small dark epithelial cells). These histologic patterns are classified as follows: grade I, tubular; grade II, cribriform; and grade III, solid [12]. The cribriform type was the most common type found in the maxillary sinus, as it is also found in the sino-nasal tract [6] while the solid subtype is the least common. The prognosis for the solid subtype is the worst while that for the tubular subtype is the best. All 3 types have a tendency for perineural invasion [13].

ACC can be misdiagnosed with other malignant neoplasms such as basaloid squamous cell carcinoma, polymorphous adenocarcinoma, mucoepidermoid carcinoma and benign tumors such as basal cell adenoma and pleomorphic adenoma. To differentiate ACC from other tumors, the use of immunohistochemical staining is a reliable approach. IHC markers such as CD117, alpha smooth muscle actin, and S100 show a positive expression for ACC while vimentin gives a negative expression [12]. In our study, we found cytokeratin 7 positivity and PS 100 negativity

The choice of treatment depends on the stage, site, grade and behavior of the tumor. Most authors consider curative surgery and adjuvant radiotherapy the mainstay of ACC treatment [14].

Surgery is considered the most successful if the free tumor margin is at least of 2 mm [8]. However, complete resection is particularly difficult due to the complex local anatomy which limits aggressive surgery, and SNACC usually presents after having already largely involved adjacent vital structures, such as the carotid arteries, orbit, dura, brain, and cranial nerves.

Regardless our case we found an invasion of the infratemporal fossa.

Considering the limitations of the surgery and the advanced stage at which most SNACC tumors present, the majority of institutions have reviewed and implemented adjuvant radiotherapy [15]. As radiotherapy for ACC progressed, it was noted that 96% of tumors responded to radiation; however, the recurrence rate after radiotherapy was 94% [16,17]. This shows that ACC is radiosensitive but not radiocurable, which makes radiotherapy a singular ineffective treatment modality. In light of these results, pure radiotherapy has been indicated to treat unresectable T4 tumors, reduce tumor burden before surgical resection, improve the likelihood of achieving local control, and provide palliative therapy [12].

Chemotherapy has little role in this disease because ACC cells have low mitotic indices [8]. Sometimes it is used preoperatively to reduce tumor burden, postoperatively for residual tumor and for palliative care [12].

When comparing all treatment modalities in isolation, those who undergone surgery appear to have the highest survival rate [12].

Despite the ability of postoperative radiotherapy to increase survival in SNACC, no statistical difference was reported between the surgery cohort and the surgery with postoperative radiotherapy cohort [18,19].

In patients with head and neck ACC, those whose disease originates from the sinonasal tract have been reported to have the poorest prognosis. Local recurrence and distant metastasis were often observed in sinonasal ACC regardless of the treatment modality [12,20]. Several studies have reported that the rate of distant metastasis was approximately 40% in sinonasal ACC patients [20].

This poor prognosis of MSACC may be due in part to the late detection of extensive tumors and their surgical inaccessibility, preventing complete excision, along with the fact that its behavior is characterized by a slow-growing, locally aggressive relentless progression of disease [7]. Similar prognostic factors have been considered by others to explain the low survival rates found with MSACC [6,9,21,22].

4. Conclusion

Adenoid cystic carcinoma of the maxillary sinus is a rare tumor with a poor overall long-term prognosis, compared to other ACCs of the head and neck. It is characterized by slow growth, tendency to recurrence, local regional spread and distant metastasis which makes it particularly difficult to diagnose and treat.

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

Conflicts of interest

The authors declare that they have no competing interests.

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Ethical approval

I certify that this kind of manuscript does not require ethical approval by the Ethical Committee of our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

M. Lahjaouj: conception and design of the study
 O. Berrada: conception and design of the study
 A. Rayhane: acquisition of data
 Y. Oukessou: drafting the article
 S. Rouadi: drafting the article
 R. Abada: revising the article.
 M. Roubal: revising the article
 M. Mahtar: final approval of the version to be submitted

Registration of research studies

This is a case report that does not require a research registry.

Guarantor

Meryem Lahjaouj

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References

- [1] Y. Li, D. Hao, X. Song, C. Zhang, 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.* (2020).
- [2] V.P. Kumar, P.N. Rao, G.A. Kumar, Adenoid cystic carcinoma of nasal cavity—a case report, *Indian J. Otolaryngol. Head Neck Surg.* 55 (1) (2003) 43–46.
- [3] S.A. Wahed, K. Pradeepthi, M.J. Kiran, G.V. Rao, Recurrent adenoid cystic carcinoma of paranasal sinuses: a rare case report, *J. Oral Maxillofac. Pathol.* 24 (Suppl 1) (2020) S28.
- [4] S.A.R. Graça, H. Sequeira, G. Coelho, H. Costa, Adenoid cystic carcinoma of head and neck: apropos of a case, *Case Rep.* 2012 (2012), bcr0220125914.
- [5] R. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 9 (2020).
- [6] C.-S. Rhee, T.-B. Won, C.H. Lee, Y.-G. Min, M.-W. Sung, K.-H. Kim, et al., Adenoid cystic carcinoma of the sinonasal tract: treatment results, *Laryngoscope* 116 (6) (2006) 982–986.
- [7] M.F. Andrade, P.R. De Faria, S.V. Cardoso, M.R.M. Santos, F.L. Dias, A.L.A. Eisenberg, et al., 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.* 43 (11) (2014) 1313–1318.
- [8] K.S. Gill, M.A. Frattali, An unusual presentation of adenoid cystic carcinoma, *Case Rep. Otolaryngol.* 2015 (2015).
- [9] G.E. Kim, H.C. Park, K.C. Keum, C.G. Lee, C.O. Suh, W.J. Hur, et al., Adenoid cystic carcinoma of the maxillary antrum, *Am. J. Otolaryngol.* 20 (2) (1999) 77–84.
- [10] R. Sigal, O. Monnet, T. De Baere, C. Micheau, L.G. Shapeero, M. Julieron, et al., Adenoid cystic carcinoma of the head and neck: evaluation with MR imaging and clinical–pathologic correlation in 27 patients, *Radiology* 184 (1) (1992) 95–101.
- [11] H.-S. Jeong, M.K. Chung, Y.-I. Son, J.Y. Choi, H.J. Kim, Y.H. Ko, et al., Role of 18F-FDG PET/CT in management of high-grade salivary gland malignancies, *J. Nucl. Med.* 48 (8) (2007) 1237–1244.
- [12] Q. Husain, V.V. Kanumuri, P.F. Svider, B.M. Radvansky, Z. Boghani, J.K. Liu, et al., Sinonasal adenoid cystic carcinoma: systematic review of survival and treatment strategies, *Otolaryngol. Head. Neck Surg.* 148 (1) (2013) 29–39.
- [13] H. Kato, M. Kanematsu, K. Sakurai, K. Mizuta, M. Aoki, Y. Hirose, et al., Adenoid cystic carcinoma of the maxillary sinus: CT and MR imaging findings, *J. Radiol.* 31 (11) (2013) 744–749.
- [14] Y. Choi, S. Kim, S.Y. Kim, S.Y. Nam, S. Choi, J. Roh, et al., Prognostic factors of adenoid cystic carcinoma of head and neck, *J. Clin. Oncol.* 29 (15 suppl) (2011), 5584–5584.
- [15] A.A. Unsal, S.Y. Chung, A.H. Zhou, S. Baredes, J.A. Eloy, Sinonasal adenoid cystic carcinoma: a population-based analysis of 694 cases, in: *International Forum of Allergy & Rhinology*, Wiley Online Library, 2017, pp. 312–320.
- [16] B. Vikram, E.W. Strong, J.P. Shah, R.H. Spiro, Radiation therapy in adenoid–cystic carcinoma, *Int. J. Radiat. Oncol. Biol. Phys.* 10 (2) (1984) 221–223.
- [17] A.S. Spiers, D.L.W. Esseltine, J.C. Ruckdeschel, J.N. Davies, J. Horton, Metastatic adenoid cystic carcinoma of salivary glands: case reports and review of the literature, *Cancer Control* 3 (4) (1996) 336–342.
- [18] J. Conley, D.L. Dingman, Adenoid cystic carcinoma in the head and neck (cylindroma), *Arch. Otolaryngol.* 100 (2) (1974) 81–90.
- [19] S. Naficy, M.J. Disher, R.M. Esclamado, Adenoid cystic carcinoma of the paranasal sinuses, *Am. J. Rhinol.* 13 (4) (1999) 311–314.
- [20] S.Y. Seong, D.W. Hyun, Y.S. Kim, H.-J. Cho, J.-G. Lee, J.-H. Yoon, et al., Treatment outcomes of sinonasal adenoid cystic carcinoma: 30 cases from a single institution, *J. Cranio-maxillofacial Surg.* 42 (5) (2014) e171–e175.
- [21] D.E. da Cruz Perez, F.R. Pires, M.A. Lopes, O.P. de Almeida, L.P. Kowalski, Adenoid cystic carcinoma and mucoepidermoid carcinoma of the maxillary sinus: report of a 44-year experience of 25 cases from a single institution, *J. Oral Maxillofac. Surg.* 64 (11) (2006) 1592–1597.
- [22] A.D. Lupinetti, D.B. Roberts, M.D. Williams, M.E. Kupferman, D.I. Rosenthal, F. Demonte, et al., Sinonasal adenoid cystic carcinoma: the MD anderson Cancer center experience, *Cancer* 110 (12) (2007) 2726–2731.

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