

A CHALLENGING CASE OF GIANT MAXILLARY AMELOBLASTOMA: NAVIGATING RARITY AND COMPLEXITY IN DIAGNOSIS AND TREATMENT

ANOUAR TITOU (DR) ¹, HAJAR EL YUBI (DR) ², ANOUAR BEN AMEUR EL YUBI (DR) ²,
ABDELLATIF OUDIDI (PR) ², NAWAL HAMMAS (PR) ³, DOUNIA KAMAL (PR) ¹, MOHAMED
NOUREDDINE EL ALAMI EL AMINE (PR) ²

¹*Department of Reconstructive and Maxillofacial Surgery, Hassan II University Hospital of Fez, Sidi Mohamed Ben Abdellah University, Faculty of Medicine, Pharmacy, and Dentistry, Fez, Morocco.*

²*ENT and Cervico Facial Surgery Department, Hassan II University Hospital of Fez, Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdellah University, Fez, Morocco.*

³*Anatomopathology Laboratory Department, Hassan II University Hospital of Fez, Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdellah University, Fez, Morocco.*

ABSTRACT

Background: Ameloblastoma is a rare, aggressive, benign odontogenic tumor that most commonly occurs in the mandible. Maxillary occurrences are rarely reported. **Case Presentation:** We report the case of a 61-year-old male presenting with right-sided unilateral nasal obstruction, characterized by a mass occupying the entire right nasal fossa upon anterior rhinoscopy. A computed tomography (CT) scan, combined with magnetic resonance imaging (MRI), revealed a locally advanced mass centered on the right maxillary sinus. The patient underwent wide excision of the mass, and histopathological analysis confirmed the diagnosis of ameloblastoma. : Naso-sinusal ameloblastomas are rare, benign, and slow-growing tumors, accounting for approximately 0.11% of nasal and sinus tumors. More common in men, they typically present between the ages of 20 and 30. Symptoms include nasal obstruction and epistaxis, with occasional facial swelling or headaches. These tumors often originate in the posterior maxillary region and extend into the maxillary sinuses and nasal cavity. Diagnosis is facilitated by imaging, where the tumors appear as solid, radio-opaque masses. Radical surgical resection is the preferred treatment, as it is essential for preventing recurrence. The prognosis is generally favorable, with no documented cases of malignant transformation or metastasis, and recurrences are rare when appropriate surgical management is performed. **Conclusion:** Medical imaging plays a crucial role in diagnosis and allows for the evaluation of tumor dissemination and anatomical involvement. The treatment of maxillary ameloblastoma typically involves surgical excision, with the complexity of management often linked to the extent of tumor spread.

KEYWORD: ameloblastoma, naso-sinusal tumor's, maxillary sinus.

INTRODUCTION

Ameloblastoma, initially referred to as adamantinoma, belongs to the group of odontogenic tumors [1]. It is a benign but aggressive tumor that most commonly affects the posterior mandible, with approximately 15% of cases occurring in the maxilla [2]. Histologically, naso-sinusal ameloblastomas are identical to those found in the oral cavity [2].

Medical imaging plays a key role in diagnosing ameloblastoma and assessing its spread and anatomical relationships. The management of maxillary ameloblastoma typically requires surgical intervention, though this procedure is often complicated by tumor recurrences during the disease course [3]. We report a unique case of a patient with a locally advanced giant ameloblastoma of the maxillary sinus, who underwent wide excision of the tumor and showed good clinical improvement at a one-year follow-up.

CASE PRESENTATION

A 61-year-old patient with an unremarkable medical history reported a 1-year history of right-sided unilateral nasal obstruction, accompanied by rhinorrhea. Clinical evaluation revealed a mass occupying the entire right nasal cavity, which was non-hemorrhagic on contact and painless (Figure 1).



Figure 1 (A budding process filling the entire right nasal fossa.)

No cervical lymphadenopathy was observed. A craniofacial computed tomography (CT) scan revealed a heterogeneous tissue lesion filling the right maxillary sinus, nasal cavity, ethmoid cells, and frontal and sphenoidal sinuses on the same side. This lesion caused bone lysis of various walls of the right

maxillary sinus and the temporal process of the zygomatic arch, with invasion of the retro-maxillary and zygomatico-masticatory spaces laterally. Inferiorly, it also caused lysis of the right alveolar and palatine processes of the maxilla. Medially, the mass invaded the nasal cavity, with complete lysis of the right turbinates and partial lysis of the nasal septum and ethmoidal septa. Superiorly, it caused partial lysis of the cribriform plate of the ethmoid on the right side, with intracranial extension affecting the anterior floor of the cranial base in that area (Figure 2).

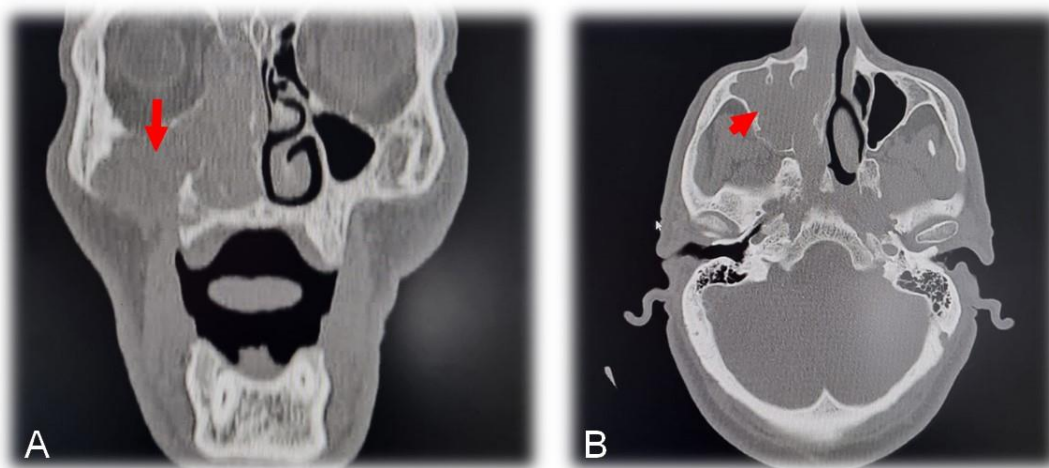


Figure 2 (Coronal (A) and axial (B) CT scans showing a heterogeneous tissue lesion process filling the right maxillary sinus, nasal cavity, ethmoid cells, and frontal and sphenoidal sinuses on the same side.)

Due to the intracranial extension, magnetic resonance imaging (MRI) was performed, which showed a poorly defined lesion in the right maxillary sinus, with restricted diffusion signal and enhancement after contrast injection. The lesion crossed the anterior floor of the cranial base, coming into contact with the right superior frontal gyrus after invading the dura mater, but without parenchymal involvement. The patient underwent surgery via a paralateral nasal approach, involving complete removal of the tumor through a right hemimaxillectomy (Figure 3).



Figure 3 (Right hemimaxillectomy via a paralateral nasal approach with complete tumor resection.)

This procedure also included opening the floor of the frontal sinus, marsupialization of the sphenoidal sinus, and anterior ethmoidectomy. Histologically, the tumor consisted of trabeculae and nodules with cystic areas. The histopathological study confirmed the diagnosis of ameloblastoma (Figure 4).

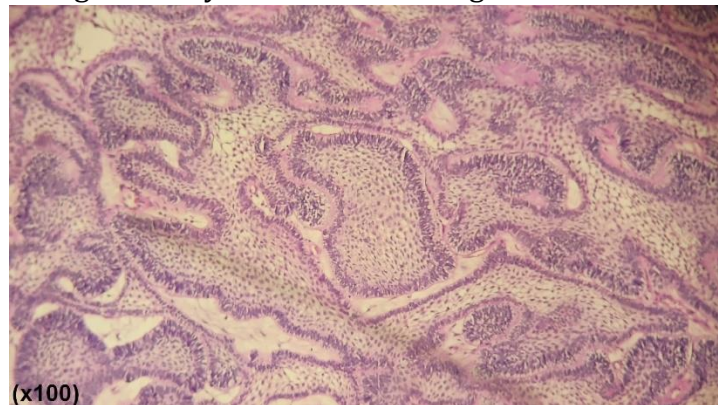


Figure 4 (Histological image showing a tumor proliferation consisting of nodules exhibiting a peripheral palisade of cells with a looser appearance in the center (x100).)

Postoperative follow-up was complicated by suture dehiscence and the development of a tissue defect. The patient subsequently underwent debridement of the edges of the right jugal tissue defect, followed by coverage with a frontal flap. Three months later, the flap was removed, and the patient showed good clinical improvement at the one-year follow-up.

DISCUSSION

Naso-sinusal ameloblastomas are tumors that develop from the epithelium of the nose and sinuses. They are benign, rare, and slow-growing [4]. These tumors account for about 0.11% of all nasal and sinus tumors and 11% of odontogenic tumors [4, 5]. Unlike their gnathic counterparts, naso-sinusal ameloblastomas are more common in men, with a male-to-female ratio of 3.8 to 1 [4]. Studies from Africa and Asia indicate that the highest frequency of these tumors occurs between the ages of 20 and 30 [6, 7]. Common symptoms include nasal obstruction and/or epistaxis. Less frequent manifestations may include facial swelling, headaches, and lacrimation [4, 8]. Most studies on maxillary ameloblastoma report a predilection for the posterior maxillary region, with early extension into the maxillary sinus, infratemporal fossa, and nasal cavity [9, 10].

Imaging plays a significant role in the diagnosis of maxillary ameloblastomas, where lesions in the nasal and sinus areas are often described as solid radio-opaque masses filling these cavities [4]. In contrast, ameloblastomas in the gnathic regions typically present as radiolucent lesions with a "honeycomb" or "soap bubble" appearance [11, 12].

According to the World Health Organization's (WHO) 2005 histological classification of head and neck tumors, ameloblastoma can be classified into four subtypes: multicystic or solid (follicular or plexiform), peripheral, desmoplastic, and unicystic [13].

The treatment of choice for naso-sinusal ameloblastomas is surgical resection [4, 14]. Radical surgery involves extensive excision with clear margins of 1.5 to 2 cm [5]. Successful treatment outcomes are strongly linked to the complete surgical removal of the lesion [8, 11]. In some clinical cases, such as late-stage diagnoses, conservative surgery may be considered, but complete tumor removal remains essential in determining the appropriate procedure [4, 14]. Radiotherapy has also been used, particularly for locally advanced, inoperable tumors or when surgery is refused [15].

The overall prognosis of naso-sinusal ameloblastomas is generally favorable [11]. Recurrence rates are higher after conservative surgery than after radical surgery [5]. Recurrences typically occur within 1 to 2 years following surgery, although cases of recurrence many years later have been reported [11]. Notably, there are no documented cases of malignant transformation, metastasis, or deaths attributed to naso-sinusal ameloblastomas [4, 8].

CONCLUSION

Naso-sinusal ameloblastomas are benign yet aggressive tumors. Medical imaging aids in diagnosis, allowing for the assessment of tumor spread and anatomical involvement. Surgical intervention is the standard management approach, with the complexity often related to the extent of tumor spread.

REFERENCES

1. Broca P: Recherches sur un nouveau groupe de tumeurs designees sous le nom d'odontomes (ed.1867). HACHETTE BNF, 2017.
2. Barrena, B.G., Phillips, B.J., Moran, K.J. et al. Sinonasal Ameloblastoma. *Head and Neck Pathol* 13, 247–250 (2019). <https://doi.org/10.1007/s12105-018-0933-3>
3. Ba B, Keita K, Coulibaly A, Ba M, Touré A, Koita H, Kassambara A, Théra TD, Guindo M, Diallo M, Coulibaly DT, Diombana ML. Les ameloblastomes des maxillaires au centre hospitalier universitaire d'odontosomatologie de Bamako [Maxillary ameloblastoma at the odonto-stomatology University Hospital in Bamako]. *Mali Med.* 2017; 32(2):1-8. French. PMID: 30079662.
4. Schafer DR, Thompson LD, Smith BC, Wenig BM. Primary ameloblastoma of the sinonasal tract: a clinicopathologic study of 24 cases. *Cancer.* 1998 Feb 15;82(4):667-74. doi: 10.1002/(sici)1097-0142(19980215)82:4<667::aid-cncr8>3.0.co;2-i. PMID: 9477098.
5. Vallicioni J, Loum B, Dassonville O, Poissonnet G, Ettore F, Demard F. Les améloblastomes [Ameloblastomas]. *Ann Otolaryngol Chir Cervicofac.* 2007 Sep;124(4):166-71. French. doi: 10.1016/j.aorl.2006.08.006. PMID: 17673157.
6. Adebayo ET, Ajike SO, Adekeye EO. A review of 318 odontogenic tumors in Kaduna, Nigeria. *J Oral Maxillofac Surg.* 2005 Jun;63(6):811-9. doi: 10.1016/j.joms.2004.03.022. Erratum in: *J Oral Maxillofac Surg.* 2005 Dec;63(12):1786. PMID: 15944979.
7. Darshani Gunawardhana KS, Jayasooriya PR, Rambukewela IK, Tilakaratne WM. A clinico-pathological comparison between mandibular and maxillary ameloblastomas in Sri Lanka. *J Oral Pathol Med.* 2010 Mar;39(3):236-41. doi: 10.1111/j.1600-0714.2009.00850.x. Epub 2010 Jan 11. PMID: 20070485.
8. Ereño C, Etxegarai L, Corral M, Basurko JM, Bilbao FJ, López JL. Primary sinonasal ameloblastoma. *APMIS.* 2005 Feb;113(2):148-50. doi: 10.1111/j.1600-0463.2005.apm1130210.x. PMID: 15723691.
9. Ogunsalu C, Scipio E, Williams N. Review of six cases of maxillary ameloblastoma from the West Indies: re-entry cryosurgery as prophylactic surgical intervention. *West Indian Med J.* 2009 Sep;58(4):398-403. PMID: 20099786.

10. Nastri AL, Wiesenfeld D, Radden BG, Eveson J, Scully C. Maxillary ameloblastoma: a retrospective study of 13 cases. *Br J Oral Maxillofac Surg.* 1995 Feb;33(1):28-32. doi: 10.1016/0266-4356(95)90082-9. PMID: 7718524.
11. Speight PM, Takata T. New tumour entities in the 4th edition of the World Health Organization Classification of Head and Neck tumours: odontogenic and maxillofacial bone tumours. *Virchows Arch.* 2018 Mar;472(3):331-339. doi: 10.1007/s00428-017-2182-3. Epub 2017 Jul 3. PMID: 28674741; PMCID: PMC5886999.
12. Weissman JL, Snyderman CH, Yousem SA, Curtin HD. Ameloblastoma of the maxilla: CT and MR appearance. *AJNR Am J Neuroradiol.* 1993 Jan-Feb;14(1):223-6. PMID: 8427095; PMCID: PMC8334476.
13. Barnes, Leon & Eveson, John & PA, Reichart & Sidransky, D.. *World Health Organization Classification of Tumours. Pathology & Genetics. Head and Neck Tumours. Tumours of the oral cavity and oropharynx.* 168-175 (2005).
14. Kumar, A., Sarswat, S., & Sharma, S. (2014: Ameloblastoma presenting as a sinonasal tumor: rare entity . *Int J Adv Health Sci.* 1:28-31.
15. Reynolds WR, Pacyniak J. Ameloblastoma: radioresistant or radiosensitive. *J Mo Dent Assoc* (1980). 1982 Nov-Dec;62(6):36-8. PMID: 6961229.