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Case Report

Pure endoscopic transsphenoidal treatment of skull base ameloblastoma with intracranial extension: Case report and literature review

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ABSTRACT

Background: Ameloblastoma is a benign locally invasive lesion that represents 1% of all oral tumors. Epidemiological characteristics are variable in the literature. The most common origin sites are mandible and maxilla. Rarely presents metastasis, but the skull base, lymph nodes, and the lung are described as metastatic sites. Low recurrence rates were reported by the authors when surgical treatment achieved complete resection.

Case Description: A female patient, 19 years old presenting moderate headache associated with nausea, vomiting, left facial hypoesthesia, and low visual acuity. Resonance image showed a heterogeneous expansive solid formation in sphenoid bone and clivus with neoplastic aspect. Signs of dissemination due to contiguity and invasion of skull base structures, especially cavernous sinus and internal carotid artery, determining also compression of the brainstem. First, an endoscopic biopsy was performed with otorhinolaryngology service. The pathological study showed histological characteristics of ameloblastoma. After, the patient was submitted to endoscopic surgery for resection of tumor.

Conclusion: Ameloblastoma is a rare tumor with benign behavior and slow growing. It arises from odontogenic epithelium and accounts 1% of all oral tumors. The mandible and maxilla are the most common sites of origin. Ameloblastoma with intracranial involvement is a rare presentation with few literature reviews. A long time illness course and multiple surgeries are characteristics present in the majority of cases described. Total resection surgery is the treatment of choice and endoscopic transnasal resection is a viable option.

Keywords: Ameloblastoma, Endoscopy, Skull base surgery, Transsphenoidal

INTRODUCTION

Ameloblastoma is a rare tumor with slow growing and benign behavior. It arises from the dental epithelium and accounts for 1% of all oral tumors.[20] Clinical studies do not show a predilection for sex, race, or age.[1]

Despite benign oncological behavior, ameloblastomas become locally aggressive when the tumor invades the skull base.[18] Ameloblastic carcinoma is an exceptionally rare and aggressive malignant tumor that can arise from a malignant transformation of ameloblastomas.[21]

The mandible is the most common site of ameloblastoma. Approximately 75-85% of the cases originate from this bone; the minority originates from maxilla (15-20%).[11] Sphenoid bone is not a typical site of origin.

The literature shows 14 cases of ameloblastoma with intracranial invasion.[20,21,18,7,10,14,1,4,19,16,8,23,6,22] We present a 15th case that corresponds to a giant ameloblastoma originating from the sphenoid bone with extension to the nasal cavity, paranasal sinuses, and to skull base, which was treated by a purely endoscopic endonasal transsphenoidal approach. The present case is the only one reported with sphenoid origin and with pure endoscopic treatment when compared to the previously published cases.

CASE REPORT

A 19-year-old female patient was referred to our hospital with moderate headache associated with nausea, vomiting, left facial hypoesthesia, and low visual acuity. These symptoms started 2 months before admission. On the neurological exam, Glasgow Coma Scale 15, bilateral papilledema, low visual acuity, left facial hypoesthesia, and absent vomiting reflex were present. Furthermore, after hospitalization, the patient evolved with dysphagia.

The magnetic resonance image (MRI) [Figure 1a-d] showed a large heterogeneous expansive formation in sphenoid bone and clivus with neoplastic aspect. Signs of dissemination due to contiguity and invasion of skull base structures, especially cavernous sinus and internal carotid artery, determining also compression of the brainstem and optic chiasm.

In view of the atypical radiological aspect, we initially opted for an endoscopic transnasal biopsy in August 2017. The pathological study showed odontogenic epithelial islands composed of peripheral palisade columnar cells at basal layer, hyperchromatic. The cells show reverse polarization away from basement membrane (Vickers-Gorlin change). The edematous center mimics the stellate reticulum of the enamel organ. No dentin or enamel formation was found. Other patterns are also seen featuring acanthomatous with squamous metaplasia and variable keratinization of stellate reticulum-like cells, and plexiform with cords and sheets of anastomosing odontogenic epithelial cells. These characteristics defined ameloblastoma as diagnosis [Figure 2a-e].

After the biopsy, we concluded that the maximal resection would be the best initial treatment. In September 2017, the patient underwent to a pure endoscopic transnasal transsphenoidal approach to the skull with a total resection of the lesion. There was mild bleeding and the lesion was very heterogeneous with some areas highly calcified [Figure 2f].

The initial endoscopic approach was chosen because it allows brainstem and optic nerves decompression, with less risk of damage to nervous and vascular structures, in addition to being a suitable surgical route for resection of the lesion in question when compared with other skull base approaches.

After surgery, the patient presented good evolution and the MRI control image demonstrated excellent local control of tumor [Figure 3a-d]. No adjuvants therapies were needed. The patient's follow-up continues after 2 years of surgery with improvement of symptoms and no evidence of lesion regrowth on the radiological exams.

DISCUSSION

Ameloblastoma is a benign locally invasive lesion responsible for 1% of all oral tumors.[15] The first case of ameloblastoma was report in 1879 by FALKSON[5] MALASSEZ[13] in 1885 used the term "adamantinoma-epithelioma." The current denomination was made by IVY and CHURCHILL.[3] Previously called adamantinoma, they are epithelial tumors of odontogenic origin, with slow growth and high incidence of recurrence after surgical excision.[11]

Epidemiological characteristics are variable in the literature. MAGLIOCA[12] refers an equal gender distribution with mean age presentation of 39 years old. However, OLAITAN[15] in a series of 315 Nigerian patients reported male dominance (61.9%) and common presentation between 30 and 40 years.[15,17]

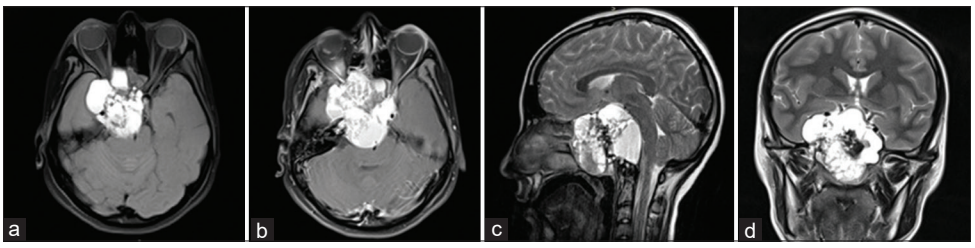

Figure 1: Preoperative MRI (a) T1 axial without contrast, (b) T1 axial with contrast, (c) T2 sagittal, (d) T2 coronal – heterogeneous expansive formation involving the skull base, mainly the sphenoid sinus and clivus with neoplastic aspect and dissemination to cavernous sinus, determining compression of the brainstem, surrounding vascular structures, right optical nerve, and optical chiasm.

The most common sites of origin are mandible and maxilla with 80% and 20%, respectively. The posterior mandible is the most common, responsible for 66% of all cases.[12,20,16] The sphenoid bone is a rare site of origin of this tumor with some reports in the literature.

Usually asymptomatic and with low growth rate, ameloblastomas can be found accidentally in routine dental exams. Rarely presents metastasis, however the skull base, lymph nodes, and the lung are possible metastatic sites. The treatment of these lesions is associated to multiple surgeries and radiation therapy witch is indicated when a subtotal resection of tumor occurs.[17,20] The duration of disease and increased number of recurrences appear to be risk factors for intracranial involvement.[20]

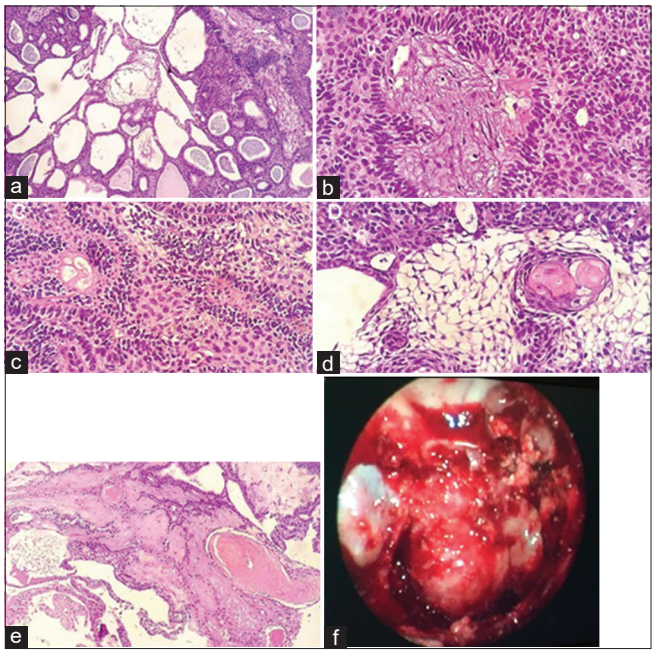

Figure 2: The classic histologic features characterized by islands of odontogenic epithelium in fibrous connective tissue; may be cystic (a). Odontogenic epithelial islands composed of peripheral palisading columnar cells at basal layer, hyperchromatic, cells show reverse polarization (b), palisading basal cells and stellate reticulum (c), the central edematous, and mimic the stellate reticulum of the enamel organ (d), featuring acanthomatous with squamous metaplasia (e), heterogeneous lesion with some areas highly calcified (f).

In treatment of ameloblastoma, surgery is the first choice. There is no doubt that the initial extent of ameloblastoma resection is an important factor that influences the rate of recurrence and the prognosis of disease.

In our case, the transnasal endoscopic transsphenoidal approach was successfully used with the inherent benefits to this minimal invasive approach, where no skin incision is required and with reduced manipulation of vascular and nervous tissues, as well satisfactory decompression of the optic nerves and of the brainstem.[20]

In other cases described in the literature of ameloblastomas with intracranial invasion, the authors used transcranial approaches in treatment, as is described at Table 1. Above, we have written the advantages of the endonasal endoscopic minimally invasive approach.

Transcranial approaches increase the manipulation of vascular and nervous tissues; however, they allow better control of other structures, in addition to offering a wider route of dissection of the lesion. In our case, the choice was for the endonasal endoscopic approach, because a large resection of tumor, in addition to satisfactory optic nerves and brainstem decompression were possible. Another reason was the bone origin of this tumor. This tumor was centered at sphenoid bone witch facilitated the endoscopic approach.

The radiotherapy is reported as adjuvant therapy in tumors with incomplete surgical resection or with recurrence.[9] Other adjuvant treatments are reported. The BRAF inhibitor was used in cases with lung metastasis based in molecular activity mutations.[2] The BRAF is a human gene that encodes a protein called B-Raf. This protein is involved in sending signals inside cells which are involved in directing cell growth.

Low recurrence rates were reported by the authors when a total resection surgery occurs (total tumor resection includes the dental and alveolar structures).[17] A long-time survival was reported in 81.8% of cases in a Nigerian study associated

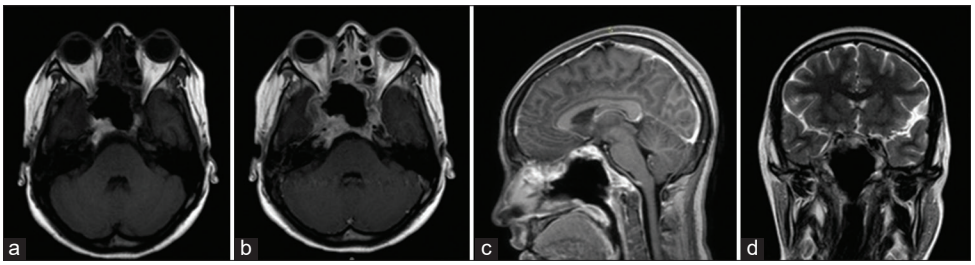

Figure 3: Postoperative MRI, (a) T1 axial without contrast, (b) T1 axial with contrast, (c) T1 sagittal with contrast, (d) T2 coronal – control image showing excellent local control of lesion, normalization of the brainstem anatomy, and absence of compression of the optic pathways.

Table 1: Ameloblastoma cases with skull base invasion.

No.	Authors/Year	Year	Sex	Age	Primary tumor location	Extracranial Involvement	Treatment	Follow-up	Outcome/Number of Surgeries
1	Harrer *et al*.[7]	1970	F	52	Mandible	Yes (Lung)	Open Surgery	15 years	Multiple Recurrence. 2 surgeries. Death
2	Kyriazis *et al*.[10]	1971	F	73	Maxilla	No	Open Surgery	7 years	Multiple Recurrence. 2 surgeries. Death
3	Oka *et al*.[14]	1986	M	27	Mandible	Yes (Femur)	Open Surgery	3 years	Multiple Recurrence. 2 surgeries. Death
4	Bredenkamp *et al*.[1]	1989	M	53	Maxilla	No	Radiotherapy alone	1 year	Primary. Good clinical condition.
5	Eliasson *et al*.[4]	1989	F	40	Maxilla	No	Open Surgery	4 years	Multiple Recurrence. 2 surgeries. Death
6	Scaccia *et al*.[19]	1991	M	53	Maxilla	No	Open Surgery	19 years	Multiple Recurrence/ 1 surgery. Death
7	Philips *et al*.[16]	1992	M	65	Mandible	No	Open Surgery and Radiotherapy	18 years	Multiple Recurrence. 3 surgeries. Good clinical condition
8	Sato *et al*.[18]	1994	M	79	Maxilla	No	Open Surgery	2 years	Multiple Recurrence. 1 surgery. Good clinical condition
9	Hayashi *et al*.[8]	1997	M	63	Mandible	No	Open Surgery	6 months	Primary. 1 surgery. Visual acuity has been limited.
10	Zarbo *et al*.[23]	2003	F	14	Maxilla	Yes (Pelvis, L2 Body and Femur)	Open Surgery and Radiotherapy	19 years	Multiple Recurrence/ 4 surgeries. Death
11	Goldenberg *et al*.[6]	2004	F	77	Mandible	No	Open Surgery	7 years	Multiple Recurrence/ Unknown. Death
12	Leibovitch *et al*.[11]	2006	M	73	Maxilla	No	Open Surgery	6 months	Primary. 1 surgery. Good clinical condition.
13	Yoshida *et al*.[22]	2009	F	70	Maxilla	No	Open Surgery	6 years	Primary/ 1 surgery. Good clinical condition
14	Woodroffe *et al*.[20]	2013	M	70	Maxilla	No	Open and Endoscopic Surgery	4 years	Multiple Recurrence/2 Surgeries. Good clinical condition
15	Author's case	2020	F	19	Sphenoid	No	Endoscopic Surgery	2 years	Primary/1 surgery. Good clinical condition

with radical surgery (follow-up ranged 6 months to 13 years). In this study, all the cases were originated in the mandible.[15]

In a literature review, we found 15 cases of ameloblastoma with intracranial invasion including the present case [Table 1]. The mean age was 55.2 years (range 14–79 years). No gender predominance was found. The majorly of the cases arose in the maxilla 9 (60%), followed by the mandible 5 (33.3%) and only one case (the present case) arose in the sphenoid bone. The extracranial involvement was observed in three cases (20%): Two cases in others distant bones and one case to the lung. Of the 15 cases, only two had treatment with an endoscopic approach (one is the current case and the other was operated with combined access), 12 were treated with open surgery and one case with radiotherapy alone.

CONCLUSION

Ameloblastoma with intracranial involvement is very rare, with only a few cases reported in the literature. Our case is unique due to the presentation in a very young patient of a large ameloblastoma with a probable origin in the sphenoid bone (witch, to the best of our knowledge, has never been reported before under these conditions) presenting an important brainstem distortion that was treated with a pure endoscopic approach.

The surgical approach with total resection of the lesion is the treatment of choice in these cases. Several surgical approaches can be used for treatment aiming at maximum resection. In this article, we present the possibility of a pure endoscopic treatment for ameloblastoma with an intracranial

invasion of a patient with more than 2 years of follow-up who still free of the disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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