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Giant ameloblastoma with intracranial invasion treated with a pure endoscopic approach

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ABSTRACT

Ameloblastoma is a benign but locally aggressive tumor that can invade the brain. A 75-year-old male patient admitted with massive exophthalmos and a tumor protruding from his nose. Magnetic resonance imaging revealed a giant mass lesion occupying the entire ventral skull base. The tumor was removed with endoscopic endonasal surgery. The patient was stable in the postoperative period, and there was no recurrence during the 5-year follow-up. Ameloblastoma may reach a huge size despite its benign character. Surgical treatment can prevent serious complications, and an endoscopic approach can be considered a treatment option.

Introduction

Ameloblastoma is a benign but locally aggressive odontogenic epithelial neoplasm that presents as a slowly growing painless swelling of the jaws and accounts for approximately 1% of all mandibular tumors and cysts (1). Although histologically benign, ameloblastoma can be locally destructive, spreading to the base of the skull, paranasal sinuses, infratemporal fossa, pterygopalatine fossa, parapharyngeal space, therefore causing severe facial deformity and functional impairment (2,3).

The treatment of ameloblastoma varies based on clinical, histopathologic, and radiographic characteristics (4). Surgery is the first line of treatment recommended to prevent recurrence and metastasis, and the goal should be complete surgical resection (5). Several surgical approaches have been used in its treatment, with varying success.

With the development of technology recently, new approaches have been added to surgical treatment techniques for ameloblastoma, and one of them is the endoscopic endonasal approach (EEA). The main advantage of EEAs is not only providing more direct access to the anterior and central skull base while avoiding craniofacial incisions commonly used in open surgical approaches but also increasing the range of the endoscopic visual surgical field with the angled lenses (6,7). EEA precisely determines the areas of tumor attachment by improving access, allowing complete tumor removal while minimizing the size of the maxillary defect and the associated morbidity as well as allowing brainstem and optic nerve decompression with less risk of damage to nervous and vascular structures (8,9).

The case described in this report is the second patient treated with a pure endoscopic approach. Additionally, it is one of the huge ameloblastomas published in the literature

Case Presentation

A 75-year-old male patient admitted with massive exophthalmos in the left eye and a tumor protruding from his nose (Figure 1A,1B). He had anosmia, headache, tingling sensation in the left half of his face, and swallowing difficulty with frequent choking for two months. His symptoms had rapidly progressed in the last two weeks. Neurological examination revealed a Glasgow Coma scale of 15, low visual acuity in the left eye, left facial hypesthesia, hearing impairment, and absence of vomiting reflex.

Magnetic resonance imaging (MRI) showed a slightly enhanced giant mass occupying the entire ventral skull base extending into cavernous sinuses, left pterygopalatine, and infratemporal fossae (Figure 1C-1E).

Computed tomography (CT) showed that all bony structures of the ventral skull base were invaded by the lesion (Figure 1F-1H).

CT angiography showed that the tumoral tissue was fed by both external carotid artery branches, more prominently on the right. Bilateral internal carotid arteries were surrounded by the tumor and displaced superiorly.

Initially, an excisional biopsy was performed. The endonasal part of the tumor was markedly removed for diagnostic purposes. Histological examination showed ameloblastoma, and the patient was referred to the neurosuregery diviison for further management where tumor excision was planned using the endoscopic skull base approach.

General anesthesia was performed by percutaneous tracheotomy due to the difficult intubation of the patient. After induction of anesthesia, head was fixed in a Mayfield clamp, head position was adapted to the tumor location and then registered with the navigation system (StealthStation System, Medtronic, USA). Initially, the tumor remaining in the nasal cavity was resected. After major decompression, with the help of endoscopic endonasal surgery, the tumor was removed from the clival, sellar, suprasellar, frontobasal regions, both cavernous sinuses, and the pterygopalatine and infratemporal fossa. After the hemostasis was obtained, skull base repair was performed with fascia lata and subcutaneous fat tissue obtained from the anterolateral of the left femoral region. Additionally, a lumbar drain was placed.

Histopathological examination was reported as plexiform type ameloblastoma **(**Figure 2A-2D).

The postoperative course was safe and stable. Neurological examination findings also remained stable. A CT scan on postoperative day 1 showed gross total resection of the tumor (Figure 1I-1K). Lumbar drain was removed on postoperative day five. A permanent dental prosthesis was placed in the dentistry service. During follow-up, annual MRI showed no signs of residual disease or recurrence for five years as of the submission of this report (Figure 1L-1P).

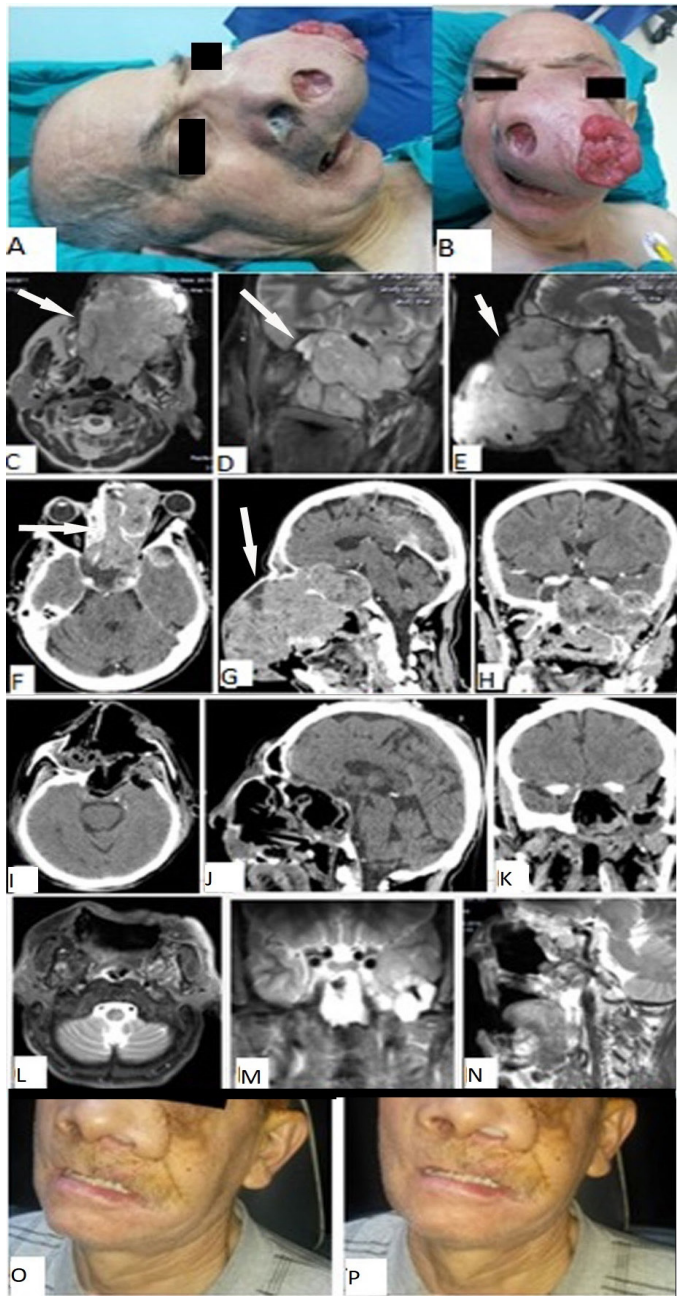

Figure 1. Preoperative (A, B) and postoperative photographs (O, P) of the patient. Preoperative axial (C), coronal (D), and sagittal (E) contrast-enhanced MRI showing a slightly enhancing, large, huge mass occupying the entire ventral skull base. The left side infratemporal fossa, pterygopalatine fossa, and base of the middle cranial fossa have been invaded by the tumor. Both cavernous was also involved by the tumor displacing both intracavernous internal carotid arteries superiorly and laterally. Axial (F), sagittal (G), and coronal (H) CT scans of the patient demonstrated that all ventral skull base bony landmarks were devastated by the expansile, lytic tumor. Axial (I), sagittal (J), and coronal (K) CT scans were obtained on the first postoperative day demonstrating the gross total resection of the tumor including the temporobasal component. Axial (L), coronal (M), and sagittal (N) contrast-enhanced MRI five years after the intervention, showing the continuous integrity of the skull base and no signs of recurrence. Both internal carotid arteries are in their normal anatomical location and are free from compression (white arrows=tumor)

MRI: Magnetic resonance imaging, CT: Computed tomography

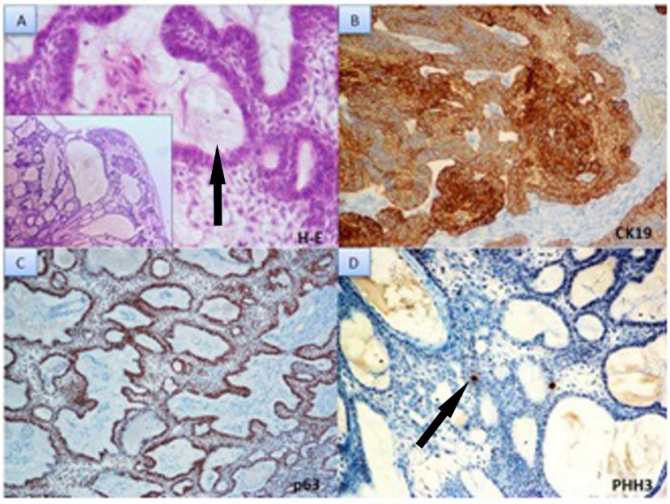

Figure 2. Solid/multicystic type, infiltrative ameloblastoma of the sinonasal tract: (A) cysts and follicular islands (black arrow) of odontogenic epithelium in the myxoid and edematous stroma, protruding to the nasal cavity as a polypoid mass (inlet) [hematoxylin-eosin (H-E), x20], (B) immunohistochemically, strong cytokeratin (CK) 19 expression (x40), (C) p63 expression of all cells (x20), and (D) a few mitoses (black arrow) with Phosphohistone H3 (PHH3) at the ameloblastic epithelium (x40)

Discussion

Ameloblastoma is a locally invasive benign epithelial origin tumor that may originate from the rest of the dental lamina, enamel apparatus, the epithelial lining of an odontogenic (dentigerous) cyst, or from the basal epithelial cells of the oral mucosa (10). They are rare neoplasms with a global incidence of 0.5 cases per million person-years most common between 30 to 60 years of age with an average of 36 years and a peak around the fifth decade (11).

The main approved treatment for ameloblastoma is surgery, and overall aim is complete resection. The literature describes two surgical therapy strategies to achieve wide surgical resection; 1) conservative surgical methods like decompression, enucleation, or curettage, and 2) radical procedures like marginal or segmental resection (4,12).

Recently, EEA has emerged as a new option for treating ameloblastoma, as the optics of endoscopes have improved, along with angled scope options. Its main advantages are direct visualization, enhanced magnification, avoidance of skin incisions, no external deformity, as well as less brain retraction, and less soft tissue damage resulting in cranial nerve deficits (13,14). Another advantage of the endoscopic surgical approach is that it reduces the operation time and has significant cosmetic benefits (15).

Conclusion

Ameloblastoma with intracranial invasion is very rare. To the best of our knowledge, the current patient is one of the few reported cases of ameloblastoma with intracranial involvement and was treated by pure endoscopic approach despite its hugeness. Endoscopic intervention may be an alternative to the conventional treatment of ameloblastoma.

Ethics

Informed Consent: Written informed consent was obtained from the patient to publish this case report anonymously.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: G.D., A.M.K., Concept: G.D., M.O.D., A.M.K., Design: M.O.D., M.C.E., A.M.K., Data Collection or Processing: D.E., Analysis or Interpretation: D.E., M.O.D., M.C.E., Literature Search: D.E., Writing: D.E., M.O.D., M.C.E.

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