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Extragnathic Sinonasal Ameloblastoma: A Rare Benign Intranasal Tumor with Malignant Features

ABSTRACT

Objective: To report a case of extragnathic sinonasal ameloblastoma and discuss its clinical features, approach to diagnosis, pathology and management.

Methods:

Design: Case Report

Setting: Tertiary Government University Hospital

Patient: One

Results: A 40-year-old female consulted for a rapidly enlarging right intranasal mass of four months duration associated with recurrent profuse epistaxis and nasal obstruction. Previous specimens of the mass were histopathologically interpreted as ameloblastoma versus craniopharyngioma. Examination revealed a pink, fleshy, smooth right intranasal mass with associated nasomaxillary bulge and supero-lateral displacement of the right eye. Computed tomography (CT) scan and magnetic resonance imaging (MRI) of the nasal cavity and paranasal sinuses demonstrated a soft-tissue density occupying the entire nasal cavity with erosion but no invasion of the maxillary sinus and no intracranial extension despite erosion of the skull base. The mass was completely excised via lateral rhinotomy and the final histopathologic diagnosis was ameloblastoma.

Conclusion: Extragnathic sinonasal ameloblastoma is a benign but locally aggressive variant of ameloblastoma involving the nasal cavity and/or paranasal sinuses often mimicking malignant tumors. Diagnosis is primarily based on histopathology but radiologic and intraoperative findings help distinguish it from differentials. Complete surgical excision remains the treatment of choice, and coupled with good follow up, may improve the prognosis of patients.

Keywords: *sinonasal ameloblastoma, extragnathic, craniopharyngioma*

Signs and symptoms of a recurrent rapidly enlarging intranasal mass, epistaxis, nasal obstruction and displacement of the ipsilateral globe lead one to suspect a possible malignancy. A physician who performs a biopsy may be surprised by a histopathologic diagnosis of

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ameloblastoma – not only because it is benign but also due to its unusual location in the nasal cavity (being odontogenic). Although benign, ameloblastoma is a locally aggressive tumor predominantly involving tooth-bearing regions of the oral cavity including the mandible and the maxilla. It is relatively rare comprising only 1% of all head and neck tumors despite being the most common true odontogenic neoplasm with an incidence of 11%.¹

Extragnathic ameloblastoma is a variant of ameloblastoma that appears to elude its pathogenesis as it arises outside the boundaries of the odontogenic apparatus.² Extragnathic ameloblastoma primarily from the nasal cavity is extremely rare with only few documented reports in the literature.²⁻⁶ Its unusual location and highly aggressive behavior make it a worthy consideration among the differential diagnosis of nasal masses that should be of interest not only to ENT surgeons with special interest in rhinology, but to maxillofacial surgeons, oral surgeons and pathologists.

Here is one such case of extragnathic sinonasal ameloblastoma and a discussion of its clinical features, approach to diagnosis, pathology and management.

CASE REPORT

A 40-year old female presented with a 4-month history of persistent mucoid, non-foul smelling, occasionally blood-tinged right rhinorrhea and recurrent nasal congestion. A previous intranasal mass punch biopsy by an otorhinolaryngologist revealed ameloblastoma; the final histopathologic diagnosis following undisclosed nasal surgery by another otorhinolaryngologist was craniopharyngioma. One month after surgery, the patient experienced recurrence of nasal obstruction and rhinorrhea, with bulging of the right nasal bridge, an enlarging right intranasal mass and spontaneous recurrent profuse epistaxis. This prompted emergency consult at our hospital.

On examination a right nasomaxillary bulge and supero-lateral displacement of the right eye were evident. Nasal endoscopy revealed a pink, fleshy, smooth mass with foul-smelling, mucoid discharge in the right nasal vestibule and contralateral septal deviation. The nasopharyngeal mass was appreciated on posterior rhinoscopy. The rest of the head and neck examination findings were unremarkable. Plain and contrast-enhanced axial and coronal computed tomography images of the nasal cavity and paranasal sinuses showed a large, lobulated, heterogeneously enhancing intranasal mass measuring 4.15 x 4.95 x 8.01 cm (transverse, craniocaudal, anteroposterior) occupying the entire nasal cavity. (Figures 1 and 2) The mass extended to the ethmoid sinuses with suspicious extension into the right orbit, sphenoid sinus, nasopharynx and partially to the right maxillary sinus with associated thinning of the medial maxillary wall.



Figure 1. Contrast-enhanced CT scan of the nasal cavity and paranasal sinuses, coronal view showing a heterogeneously enhancing mass occupying the entire nasal cavity with thinning of the right medial maxillary wall and lamina papyracea. There is opacification of the right maxillary sinus.



Figure 2. Contrast-enhanced CT scan of the nasal cavity and paranasal sinuses, axial view showing additional opacification of the right sphenoid sinus.

Due to the extensive involvement of adjacent structures, MRI was requested to rule out intracranial extension or origin (excluding craniopharyngioma) and paranasal origin (excluding gnathic maxillary ameloblastoma) of the tumor. MRI revealed fluid accumulation--possibly from obstruction without invasion of the maxillary sinus.

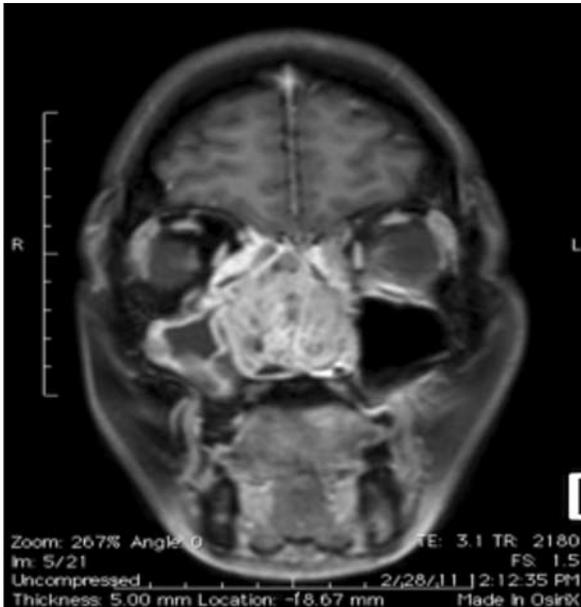


Figure 3. MRI of the nasal cavity and paranasal sinuses with gadolinium, coronal view showing a heterogeneously contrast-enhancing mass in the nasal cavities with fluid accumulation in the right maxillary sinus.



Figure 4. MRI of the nasal cavity and paranasal sinuses with gadolinium, sagittal view showing a heterogeneously contrast-enhancing mass in the nasal cavities with extension to the right sphenoid sinus.

(Figure 3) The sphenoid sinus was only partially occupied by the tumor alongside fluid accumulation. (Figure 4) Likewise, there was no evidence of intracranial extension or origin.

A slide review of the specimen from the previous surgery was interpreted as consistent with ameloblastoma versus

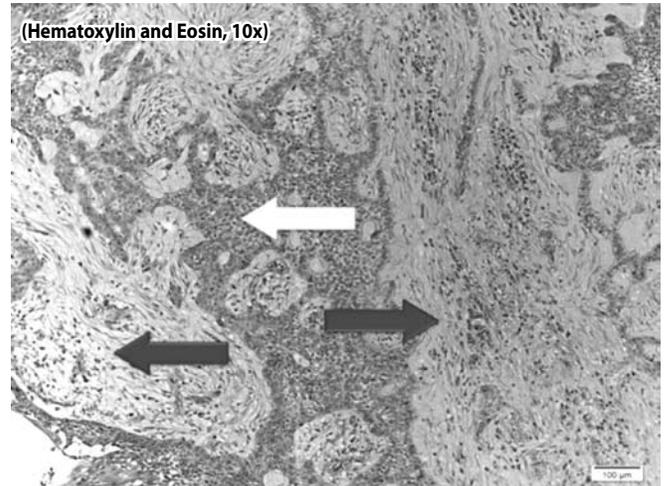


Figure 5. Histopathologic slide, Hematoxylin and Eosin stain, scanning view (10x) shows blue-gray relatively less cellular areas (blue arrow) and dark blue cellular areas (yellow arrow). The blue-gray areas correspond to the loose, fibrous stroma while the dark blue cellular areas correspond to the interspersed cords and islands of epithelial cells.

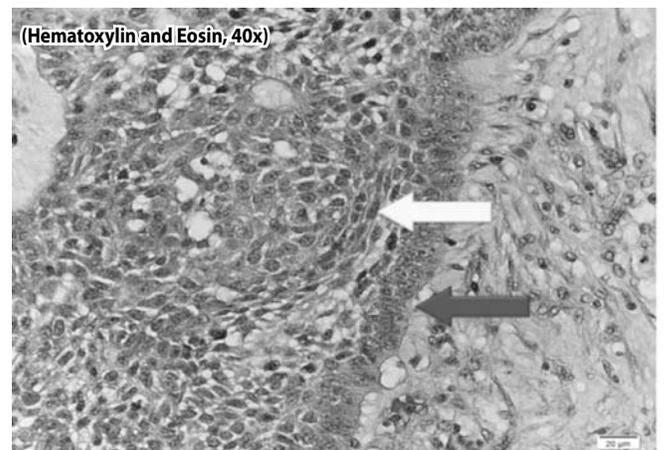


Figure 6. Histopathologic section, Hematoxylin and Eosin stain, higher magnification (40x) of the epithelial component shows two cell types. In the periphery, there are columnar cells that show palisading with a reversed polarity arrangement (dark arrow). Medial to it, the cells are arranged more loosely, corresponding to the stellate reticulum. Higher magnification of the stroma (light arrow) shows fibroblasts arranged in loose connective tissue.

craniopharyngioma. Excision of the mass via right lateral rhinotomy and partial medial maxillectomy yielded a grayish, friable, fungating mass occupying the right nasal cavity attached to the right postero-medial choana extending to the sphenoid sinus pushing against (but not involving) the septum with no involvement of the right maxillary sinus or attachment to the skull base. Final histopathology was signed out as ameloblastoma. No recurrence was noted after 11 months of postoperative follow up.

DISCUSSION

Ameloblastoma is a locally aggressive benign tumor of odontogenic tissues with a high rate of recurrence if not adequately excised. It represents 1% of all oral cavity tumors, generally appearing in the

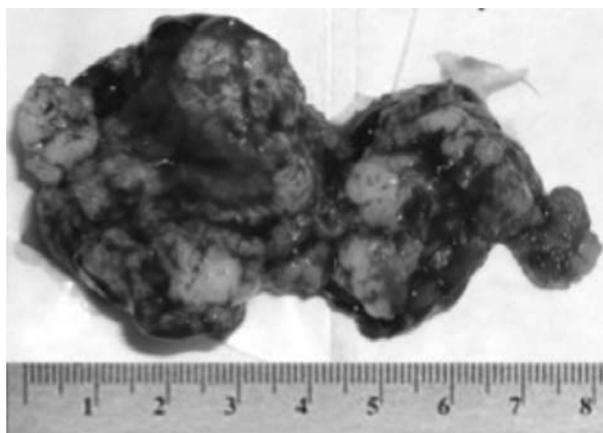


Figure 7. Gross specimen consisting of cream tan, irregular, soft to rubbery tissue fragments with an aggregate diameter of 8 cm.

mandible in 80% of cases and 15-20% in the maxilla.²

Differential diagnoses of an intranasal ameloblastoma include 1) gnathic ameloblastoma from the maxilla with the nasal cavity only secondarily involved; 2) extragnathic ameloblastoma, a variant of ameloblastoma that arises primarily in the sinonasal mucosa known as primary sinonasal ameloblastoma⁷; and 3) infrasellar craniopharyngioma extending down to the nasal cavity. The majority of intranasal ameloblastomas in the literature are actually of maxillary origin with extension into the nasal cavity.⁸ Ameloblastoma exclusively arising in the sinonasal tract is extremely rare, with very few reported cases in the literature²⁻⁶ and its presence should be well established before making a definitive diagnosis. There are even fewer reports of infrasellar craniopharyngioma with intranasal extension in the literature.^{9,10} The difficulty in distinguishing these two entities (ameloblastoma and craniopharyngioma) as evidenced by the histopathologic reports in this case stems from the very similar histopathologic features reflective of their odontogenic origins. In addition, the possibility of malignant ameloblastic carcinoma (although rare) should never be discounted, especially in aggressive recurrent cases¹¹ nor should malignant transformation of ameloblastoma be overlooked (diagnosed with specific staining like CK AE1/AE3).¹²

The histopathologic sections (*Figures 5 and 6*) showing odontogenic epithelium arranged in long strands and cords that appear to surround central areas of supporting stroma with palisading columnar cells exhibiting reverse polarization surrounding loosely arranged stellate reticulum-like epithelium are compatible with a diagnosis of ameloblastoma, both for gnathic and extragnathic variants. Although the same features are found in craniopharyngioma, the absence of cystic formation, degenerative changes, calcifications and cholesterol clefts⁶ characteristic of the latter favor a diagnosis of ameloblastoma.

As evidenced by the histomorphologic, radiologic and intraoperative findings in the patient, a diagnosis of extragnathic sinonasal ameloblastoma was established.

Extragnathic ameloblastomas comprise only 2–10% of all ameloblastomas.¹⁰ Extragnathic sinonasal ameloblastomas are even less common. Schafer *et al.* reviewed nearly 20,000 sinonasal tumors over a 40-year period and reported only 24 cases of ameloblastoma exclusively arising in the sinonasal tract.⁸ To date, only five additional case reports have been published, based on a PubMed and Google search using the keywords “ameloblastoma,” “sinonasal,” and “extragnathic.”²⁻⁶ The overall mean age at presentation is 59.7 years and more males are affected than females with a ratio of 3.8:1.⁴ In contrast, this case involved a relatively young 40-year-old female.

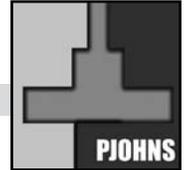
Usual presenting signs and symptoms mimic those of malignant tumors which include intranasal mass, nasal obstruction, sinusitis, epistaxis, facial swelling, dizziness, and headache.⁸ In this case, the intranasal mass, epistaxis and nasal obstruction are consistent with the usual signs and symptoms. The additional supero-lateral displacement of the right globe can be attributed to tumor mass effect.

Sinonasal ameloblastomas are described as polypoid, predominantly solid masses with glistening gray-white, pink or yellow-tan color, ranging from a few millimeters to 9.0 cm with consistency varying from rubbery to granular⁸ consistent with the cream to tan, fleshy, rubbery mass in this case. (*Figure 7*)

CT scans of the nasal cavity and PNS were the primary imaging modality in previous reports. The appearance of sinonasal ameloblastoma depends on tumor extent, generally as a solid mass, soft tissue density or opacification occupying the nasal cavity and/or the paranasal sinuses with occasional bony erosion as seen in this case.^{3,8} The additional use of MRI in this case was beneficial in delineating tumor extent.

The reported sites of origin of primary sinonasal ameloblastoma confined to the nasal cavity include the nasal septum, lateral nasal wall and turbinates.^{4,8,10} Among the paranasal sinuses, the maxillary sinus was most commonly affected, followed by the ethmoid, frontal and sphenoid sinuses (the latter with only one reported case).⁸ In this case, the mass was surgically confirmed to arise from the postero-medial choana with extension into the sphenoid sinus, an occurrence not previously reported.

Gnathic ameloblastoma arises most frequently from rests of primitive dental lamina in the gingiva, alveolar bone above the level of tooth apices, follicular walls of unerupted teeth, lining of odontogenic cysts, and even gingival surface epithelium.¹⁰ Controversy surrounds the origin of extragnathic ameloblastoma particularly those arising in the nasal cavity. This entity is believed to arise from the pluripotential



basal layer of the surface epithelium or from ectopic epithelial rests.^{8,10}

Management of extragnathic sinonasal ameloblastoma as with ameloblastoma is surgical with good prognosis following complete tumor excision. The approach depends on the extent of the tumor and experience of the surgeon. The goal is to completely remove the entire tumor to reduce the risk of recurrence with preservation of as much normal tissue as possible to reduce morbidity. In the current case, a lateral rhinotomy approach was employed due to the extensive involvement of adjacent structures by tumor. Recurrences have been documented even after adequate surgery, and close follow up should be emphasized.

Extragnathic sinonasal ameloblastoma, an extremely rare variant of ameloblastoma, is benign but may present as a locally aggressive entity, mimicking malignant tumors involving the nasal cavity and/or the paranasal sinuses. Diagnosis is primarily based on histopathology but radiologic and intraoperative findings aid in its distinction from the closest differentials. Apart from CT scan as a primary imaging modality, MRI plays a crucial role in extensive cases where involvement of vital structures needs to be assessed. Complete surgical excision remains the treatment of choice, and coupled with good follow up should serve to improve the prognosis of patients.

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