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Sinonasal Ameloblastic Carcinoma in a 50-year-old Filipino Female: Continuing Tale of the Unexpected

ABSTRACT

Objective: To report the possible malignant transformation of primary sinonasal ameloblastoma into sinonasal ameloblastic carcinoma.

Methods:

Design: Case Report

Setting: Tertiary Public University Hospital

Patient: One

Result: A 50-year-old woman with a previous diagnosis of sinonasal ameloblastoma reported recurrence of symptoms of right-sided nasal obstruction and epistaxis two years after endoscopic sinus surgery. Clinical examination, CT scans and subsequent total maxillectomy with orbital exenteration revealed a left intranasal mass with maxillary, ethmoid and orbital floor extension and pulmonary and hepatic metastases. Histopathologic findings of palisading columnar epithelium with reverse polarity with malignant features were consistent with ameloblastic carcinoma. Despite subsequent cycles of chemotherapy, the patient died two years after surgery. To the best of our knowledge, there have been no published reports of a primary sinonasal ameloblastoma with malignant transformation in the English literature.

Conclusion: Ameloblastic carcinoma is a rare neoplasm which may arise *de novo* or from malignant transformation of an ameloblastoma. Because ameloblastoma is commonly encountered in our setting, clinicians should be aware of this possibility and closely follow their patients accordingly.

Keywords: *sinonasal, maxillary, ameloblastic carcinoma, malignant transformation*

An endoscopically excised exophytic papilloma turned out to be a primary sinonasal ameloblastoma for which “no deaths, metastases and malignant transformation has been reported.”¹ Unaware that she was the subject of this prior publication, we encountered the same patient with recurrence of symptoms of right-sided nasal obstruction and epistaxis two years later. Unfortunately, we now have to report possible malignant transformation, metastasis and death. Here is her continuing story.

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CASE REPORT

A 50-year-old woman initially presented at the age of 46 in July 2009 with recurrent right-sided nasal obstruction and ipsilateral “spontaneous epistaxis”; “thin brown” rhinorrhea and frontonasal throbbing headache.¹ Examination then revealed a “pale, pink irregularly shaped polypoid mass attached to the lateral nasal wall almost completely obstructing the nasal cavity.”¹ Plain paranasal sinus CT scans “showed opacification of the right nasal chamber by soft tissue densities with obstruction of the ipsilateral ostiomeatal unit and sphenothmoidal recess.”¹ (Figure 1) An intranasal biopsy obtained an aggregate of 2.5 cm soft, friable, fleshy tissue that was histopathologically diagnosed as “sinonasal exophytic papilloma.”¹

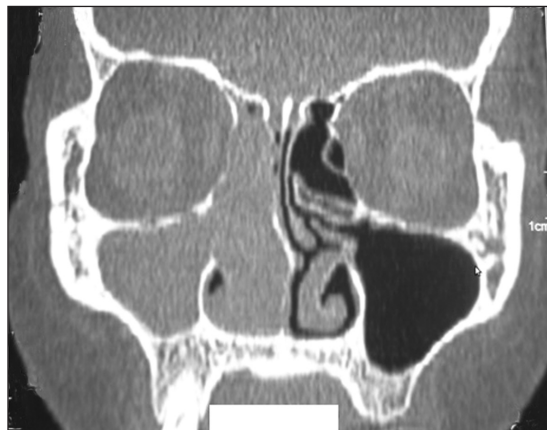


Figure 1. Pre-operative paranasal sinus screening CT Scan, 2010, representative coronal cut (a similar figure has been previously published in this journal).¹

Endoscopic sinus surgery was performed in July 2010. The mass was followed up to the lateral nasal wall, the uncinate process was opened and a cuff of normal tissue was excised around the root of the papilloma. There were no remnant masses noted in the maxillary sinus, anterior ethmoids and frontal sinus (which only contained mucus). Histopathologic diagnosis was extragnathic soft tissue ameloblastoma¹ with positive tumor tissue in the specimen labelled “ethmoid.” (Figure 2) Despite being advised on the possibility of recurrence and importance of regular monitoring, the patient only followed up for a month.

Unilateral, right-sided epistaxis recurred 2 years later in October 2012 and by December that year, she experienced right-sided gradually worsening nasal obstruction with occasional difficulty of breathing. She finally consulted in March 2013 with a fleshy mass in the right nostril. Paranasal sinus CT Scan revealed an expansile lytic lesion with osseous matrix and sunburst periostitis involving the medial wall of the right maxillary sinus extending superiorly to involve the anterior portion of the inferior orbital wall and anteriorly to involve the medial portion of the anterior maxillary wall and nasolacrimal duct opening; with a

heterogeneously enhancing soft tissue component filling the right nasal antrum, maxillary, ethmoid and sphenoid sinuses and obstructing the ostiomeatal unit. (Figure 3) Histopathology of an intranasal punch biopsy specimen revealed odontogenic carcinoma.

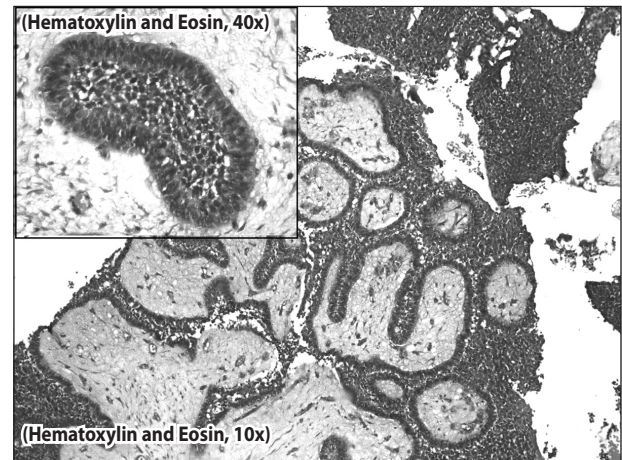


Figure 2. Histopathology, s/p Endoscopic Sinus Surgery, July 2010, Hematoxylin-Eosin, 10x, showing interconnecting trabecula and islands of benign odontogenic epithelium in an edematous, myxoid, hypocellular stroma. Inset (Hematoxylin-Eosin 40x): an epithelial island showing peripheral palisading columnar or cuboidal cells with reverse polarity consistent with Ameloblastoma (a similar figure has been previously published in this journal).¹

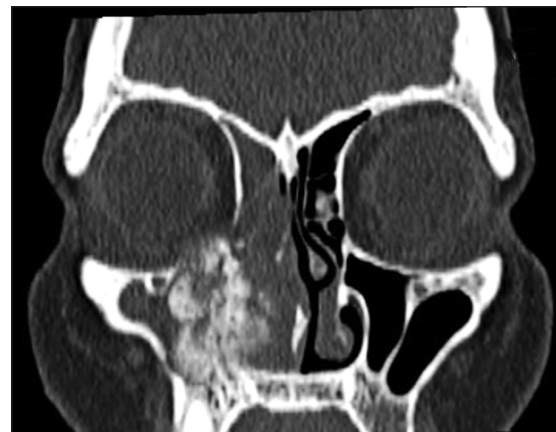


Figure 3. Contrast-enhanced paranasal sinus CT scan, 2013, representative coronal cut, taken after recurrence of symptoms two years after endoscopic sinus surgery.

The patient underwent total maxillectomy with right orbital exenteration and prosthetic reconstruction under general anesthesia in July 2013. Intraoperative tumor extension to the anterior maxillary wall, orbital floor and beyond the orbital periosteum was seen. (Figure 4) Histopathologic diagnosis was “odontogenic carcinoma consistent with ameloblastic carcinoma, maxilla, 5 cm in greatest dimension (specimen consists of a right maxilla with mass and scanty soft tissue measuring 7 x 5 x 2 cm, with the irregularly shaped mass measuring 5 x 3 x 3 cm in the center of the bone); positive for tumor at its superior

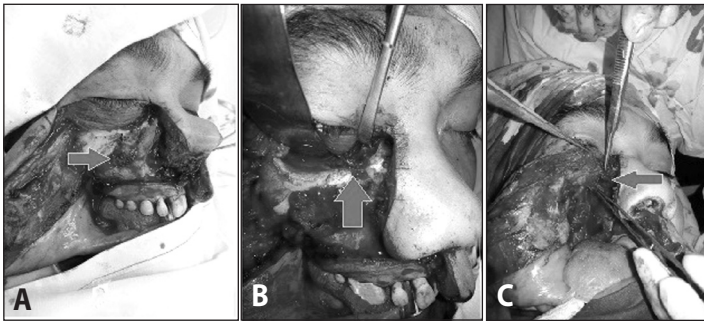


Figure 4. Intra-operative photos, 2013, showing extensions **A.** to the anterior maxillary wall (arrow), **B.** beyond orbital floor and periosteum (arrow), and **C.** to the ethmoids (arrow).

and inferior margins; also positive for tumor in the specimen labeled 'ethmoid' (consists of a 2 x 1.5 x 0.3cm soft to rubbery, tan brown, irregularly-shaped tissue fragment) and 'inferior orbital wall' (consists of brown, gritty, irregularly-shaped tissue fragments with an aggregate diameter of 1 cm)." (Figure 5A-C)

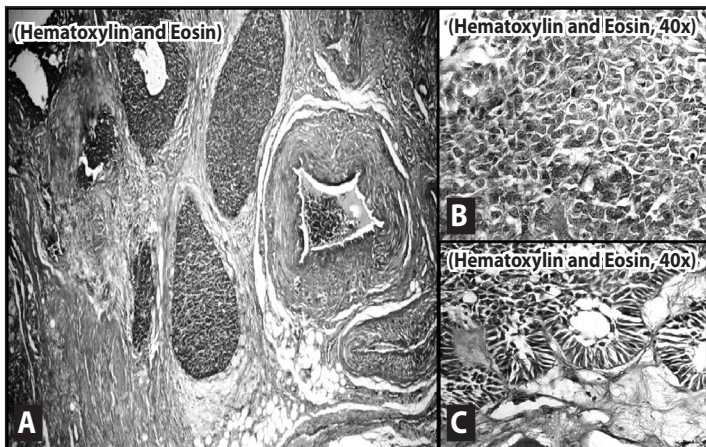


Figure 5. Final histopathology, ameloblastic carcinoma. **A.** Hematoxylin-Eosin, low power showing epithelial islands and trabecula and a myxoid stroma **B.** Hematoxylin-Eosin, 40x, an area showing malignant features including variability of nuclear staining, prominent nucleoli, and mitoses; **C.** Hematoxylin-Eosin, 40x, an area showing palisading columnar epithelium with reverse polarity consistent with Ameloblastoma.

Further surgery was performed in October 2013 to address the positive margins. Intra-operatively, tumor in the superior ethmoid area was positive for ameloblastic carcinoma on frozen section. Bare bone excision was attempted with tumor noted in the area of the cribriform plate. CT Scans revealed pulmonary (Figure 6A) and hepatic (Figure 6B) metastases, and several cycles of chemotherapy were administered. The patient died after two years of treatment.

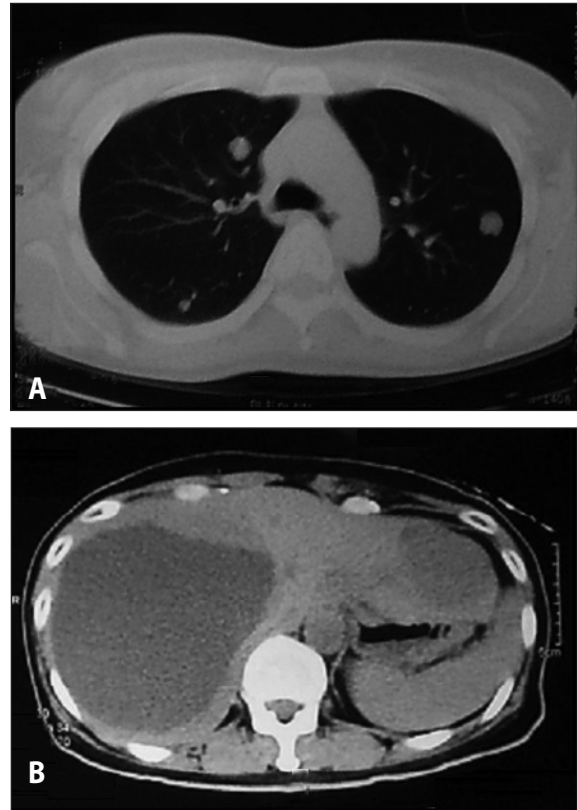
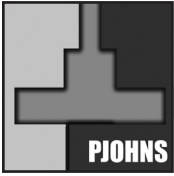


Figure 6. Contrast-enhanced CT Scan, representative axial cuts, 2013 October, showing **A.** multiple bilateral pulmonary nodules, and **B.** a hypodense liver focus.

DISCUSSION

We only learned that our patient was the same woman described in the previous report' while reviewing the literature for this paper. The initial diagnosis of exophytic papilloma and subsequent postoperative histopathologic diagnosis of extragnathic soft tissue ameloblastic carcinoma, metastasis and death need to be reported and the literature reviewed.

Ameloblastic carcinoma designates lesions that exhibit histologic features of both ameloblastoma and carcinoma with or without metastasis. As shown in the representative slides of the patient, while the classic stellate reticulum-like interior of the enamel organ consistent with ameloblastoma is not as obvious (Figure 5A), features of malignancy including variability of nuclear staining, prominent nucleoli and mitoses were demonstrated (Figure 5B). The peripheral layer of palisading columnar or cuboidal cells with hyperchromatic small nuclei oriented away from the basement membrane— so called reverse polarity can also be seen. (Figure 5C).



Swelling of the involved site is the primary clinical manifestation of maxillary involvement of an ameloblastic carcinoma.²⁻⁴ Our patient unusually presented with an intranasal mass, right-sided nasal obstruction and epistaxis but not swelling. Although reports of nasal obstruction pertain to ameloblastoma rather than maxillary ameloblastoma,^{5,6} there are reports of sinonasal ameloblastoma presenting with complete nasal obstruction.^{7,8} Indeed, an uncommon neoplasm such as ameloblastic carcinoma may present unusually and mimic more common disease processes or a benign counterpart.

The relationship between the histopathologic diagnoses from exophytic papilloma to extragnathic soft tissue ameloblastoma may be explained by the ectodermally-derived ciliated respiratory mucosa that lines the nasal cavity and paranasal sinuses, called the Schneiderian membrane which may give rise to Schneiderian papillomas—exophytic papillomas being one morphological type.^{9,10} Ameloblastomatous epithelial proliferations are also often seen in continuity with native sinonasal schneiderian epithelium.^{6,9,10}

The largest comprehensive study of primary sinonasal ameloblastoma by Schafer *et al.* reported no malignant transformation published in the literature¹¹ and only one case report by Karakida, *et al.* subsequently discussed a secondary type which may have arisen from an untreated ameloblastoma of the maxilla.¹²

To the best of our knowledge, no published reports of a primary sinonasal ameloblastoma with malignant transformation are available in the English literature and this may be the first reported death from a sinonasal ameloblastic carcinoma two years after diagnosis.

The advanced disease of our patient manifested by tumor in the area of the cribriform plate and possible pulmonary and hepatic metastases contributed to the difficulty in management. After total maxillectomy with orbital extenteration, subsequent cycles of chemotherapy were given despite the limited available data for the best treatment option.¹³⁻¹⁶

This case has demonstrated that malignant transformation and death previously unreported for ameloblastoma of the maxilla is possible. Ameloblastic carcinoma is a rare neoplasm that may arise *de novo* or from malignant transformation of an ameloblastoma. Because ameloblastoma is commonly encountered in our setting, clinicians should be aware of this possibility and closely follow their patients accordingly.

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