Ameloblastic Carcinoma Arising from Anterior Skull Base

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ABSTRACT

Ameloblastic carcinoma (AC) is an aggressive malignant epithelial odontogenic tumor. It may appear de novo or originate from a pre-existing ameloblastoma or odontogenic cyst. To our knowledge, an AC that originates from the anterior skull base has not been reported before in the English literature. We report a case of an AC that originated from the anterior skull base and invaded the dura of the anterior fossa and discuss its clinical course and treatment.

KEYWORDS: Ameloblastoma, ameloblastic carcinoma, odontogenic carcinoma, skull base

Ameloblastic carcinoma (AC) is an extremely rare, aggressive malignant epithelial odontogenic tumor with a poor prognosis. Two thirds of these tumors arise from the mandible while one third originate in the maxilla.¹ The most common symptom is a rapidly progressing painful swelling. There is no consensus on the treatment of ACs; however, wide surgical excision with or without radiotherapy is the most common treatment modality.^{2,3}

We report an aggressive AC that originated from the anterior skull base and invaded the dura of the anterior fossa. As far as we know, this is the first

case of an AC in the English literature reported to originate from the anterior skull base without a primary tumor in the mandible or maxilla.

CASE REPORT

A 23-year-old woman was admitted to our otorhinolaryngology outpatient clinic complaining of a severe headache and a rapidly progressing swelling on her nose. On inspection, a 2 × 3-cm swelling on her nasal root was most prominent on the right side. The skin overlying the swelling seemed normal. On

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nasal endoscopy, bilateral reddish polypoid masses hung from the anterior skull base medial to both middle turbinates. There were no palpable lymphadenopathies or masses in the neck. The biopsy obtained from the nasal mass was identified as "squamous cell carcinoma."

Paranasal computerized tomography (CT) revealed that the mass occupied the superior part of the nasal cavity to the level of the posterior ethmoidal cells. The overlying nasal bones and subcutaneous tissues appeared to have been invaded. Neither the ethmoid nor the sphenoid sinuses were involved (Fig. 1). On cranial magnetic resonance imaging (MRI) the mass seemed adjacent to the dura of the anterior fossa without any significant evidence of dural invasion. There was no evidence of distant metastasis on CT of the thorax and abdominopelvic region or on Tc99m bone scintigraphy. An anterior craniofacial resection was planned.

The mass was excised en bloc with both lamina papyracea, nasal bones, and the skin. The posterior border of the resection was the anterior wall of the sphenoid sinus. The cribriform plate and anterior fossa dura were invaded by the tumor. The invaded portion of the dura was also excised and duraplasty was performed. The anterior skin defect was reconstructed with a radial forearm free flap. There were no major complications during the surgery or postoperative period.

The histopathological changes were characteristic of AC. The neoplastic cells formed nests of cells with the distinctive features of ameloblastic differentiation: peripheral pallisading of basaloid cells coupled with the dyscohesiveness of the cells in the middle of the nests created the typical stellate reticulum arrangement (Figs. 2, 3). Focal cystic degeneration in the middle of the cellular clusters had left a rim of basaloid cells clinging to the stroma, also characteristic of ameloblastic tumor.

Figure 1 (A) Coronal and (B) axial CTs show the mass between the two middle turbinates. Neither the ethmoid nor the sphenoid sinuses are involved.

Figure 2 The tumor was composed of variably sized nests of basaloid cells that formed a jigsaw puzzle arrangement. Some of the nests showed cystic degeneration, with a layer of basaloid cells clinging to the stroma.

The degree of mitotic activity, presence of necrotic foci, and infiltrative characteristics of the tumor supported the malignant nature of this ameloblastic neoplasm. After the main specimen was analyzed, the punch biopsy specimens were re-examined. The final pathological analysis was that those biopsy materials were in fact ACs.

Postoperative panoramic radiography of the mandible was normal.

Figure 3 Despite the relative maturation of the cells, no keratinization was evident. Instead, the dyscohesiveness of the cells at the center of the nests created the stellate reticulum formation, characteristic of ameloblastic neoplasia.

Seven months after surgery, a fragile mass was seen on nasal endoscopy on the rest of the nasal septum immediately anterior to the choanae. A punch biopsy was obtained and identified as AC. Paranasal CT revealed multifocal masses with amorphous calcifications anterior to the choanae and adjacent to the left orbit. Cranial MRI showed no intracranial involvement. The patient received 5000 cGy of radiotherapy. No mass was evident on nasal endoscopy performed 6 weeks after the end of radiotherapy. The patient's follow-up clinical examination and radiological imaging performed 18 months later showed no local recurrence or distant metastasis.

DISCUSSION

Histologically, ameloblastoma is a benign neoplasm that arises from the odontogenic apparatus and constitutes only ~1% of tumors and cysts in the jaw.⁴ The malignant form of ameloblastoma has been controversial for many years. The term "malignant ameloblastoma" implies that lesions metastasize despite their benign histology. The term "AC" is reserved for an ameloblastoma with a malignant morphologic appearance, regardless of the presence of metastasis.

ACs are extremely rare malignant odontogenic epithelial neoplasms and may arise *de novo* or from a pre-existing odontogenic lesion.^{5,6} They typically involve the mandible and less often the maxilla. Involvement of the nasal cavity is usually related to local invasion of the maxillary ACs. In our case, the maxilla was free of tumor. The tumor primarily involved the anterior skull base, suggesting a *de novo* neoplasm of this region.

The embryology of the sinonasal tract and odontogenic apparatus accounts for the growth of a *de novo* odontogenic tumor in the anterior skull base. The embryological development of the sinonasal tract and the odontogenic apparatus are closely related to each other. The oral cavity and sinonasal tract communicate until the 10th intrauterine

week.⁷ These two cavities are separated by the development of the palatine shelves.⁸ During this period, the odontogenic epithelium may be trapped in the sinonasal mucosa, or the sinonasal cells may acquire the capability of odontogenesis.⁸ It has also been suggested that odontogenic tumors may originate from the pluripotent cells of the basal layer of the oral and sinonasal epithelium.^{9,10} Ectopic teeth in the nasal cavity may also be a potential source for odontogenic neoplasms.⁷

The main differential diagnosis for this tumor was squamous cell carcinoma, in particular, the basaloid variant. In this case the features that distinguished the AC from squamous cell carcinoma included the jigsaw puzzle-type nesting of the tumor cells, the absence of frank keratinization (despite the relatively bland cytologic findings), the presence of stellate reticulum, and the distinctive cystic degeneration of the nests. The diagnosis of craniopharyngioma was also considered in the differential diagnosis, primarily because of its well-known similarities to odontogenic neoplasia and partially because of its location in the cranial base. However, this possibility was ruled out because the findings were characteristic of AC.

Because ACs are rare, there is no consensus on their treatment. Despite the lack of adequate clinical data, surgery followed by radiotherapy seems to be the treatment of choice in most reported cases.^{2,3,11} Preoperative radiotherapy has been suggested to decrease the tumor size and may be used to treat some rapidly growing tumors before radical surgery.¹² The role of chemotherapy is not yet proven.¹³ We performed an anterior craniofacial resection and administered postoperative radiotherapy. Unfortunately, because of financial issues, the radiotherapy was administered late in the postoperative period, after the tumor recurred locally. However, the recurrent tumor disappeared after radiotherapy.

ACs can recur locally 0.5 to 11 years after definitive therapy.^{2,14} Distant metastasis is usually fatal and may appear as early as 4 months or as late as 12 years postoperatively.² The most common site for a distant metastasis is the lung, followed by

bone, liver, and brain.^{2,3,11,13} Distant metastasis can occur in the absence of a local or regional recurrence.³

Although ACs primarily involve the mandible and maxilla, they may appear in the anterior skull base and nasal cavity. Larger clinical series and longer periods of follow-up are needed to establish the best therapeutic option for these tumors.

REFERENCES

1. Lolachi CM, Madan SK, Jacobs JR. Ameloblastic carcinoma of the maxilla. *J Laryngol Otol* 1995;109:1019-1022
2. Dhir K, Sciubba J, Tufano RP. Ameloblastic carcinoma of the maxilla. *Oral Oncol* 2003;39:736-741
3. Simko EJ, Brannon RB, Eibling DE. Ameloblastic carcinoma of the mandible. *Head Neck* 1998;20:654-659
4. Calcaterra TC, Wang MB, Sercarz JA. Unusual tumors. In: Myers EN, Suen JY, eds. *Cancer of the Head and Neck*. Philadelphia, PA: Saunders; 1996:644-668
5. Slootweg PJ, Muller H. Malignant ameloblastoma or ameloblastic carcinoma. *Oral Surg Oral Med Oral Pathol* 1984;57:168-176
6. Cizmeci O, Aslan A, Onel D, Demiryont M. Ameloblastic carcinoma ex ameloblastoma of the mandible: case report. *Otolaryngol Head Neck Surg* 2004;130:633-634
7. Weber BP, Kempf HG, Mayer R, Braunschweig R. Ectopic teeth in the area of the paranasal sinuses [in German]. *HNO* 1993;41:317-320
8. Guilemany JM, Ballesteros F, Alos L, et al. Plexiform ameloblastoma presenting as a sinonasal tumor. *Eur Arch Otorhinolaryngol* 2004;261:304-306
9. Schafer DR, Thompson LD, Smith BC, Wenig BM. Primary ameloblastoma of the sinonasal tract: a clinicopathologic study of 24 cases. *Cancer* 1998;82:667-674
10. Woo SB, Smith-Williams JE, Sciubba JJ, Lipper S. Peripheral ameloblastoma of the buccal mucosa: case report and review of the English literature. *Oral Surg Oral Med Oral Pathol* 1987;63:78-84
11. Datta R, Winston JS, Diaz-Reyes G, et al. Ameloblastic carcinoma: report of an aggressive case with multiple bony metastases. *Am J Otolaryngol* 2003;24:64-69
12. Andersen E, Bang G. Ameloblastic carcinoma of the maxilla. *J Maxillofac Surg* 1986;14:338-340
13. Bruce RA, Jackson IT. Ameloblastic carcinoma. Report of an aggressive case and review of the literature. *J Craniomaxillofac Surg* 1991;19:267-271
14. Lee L, Maxymiw WG, Wood RE. Ameloblastic carcinoma of the maxilla metastatic to the mandible. *J Craniomaxillofac Surg* 1990;18:247-250

Commentary

This is a well-written, well-illustrated, thorough yet concise case report of a very rare histology involving the anterior cranial base. As such, our

differential diagnosis for lesions in this region needs to be expanded to include this entity. This case will serve as a nidus for future systematic reviews and complications of the topic.

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