Ameloblastic Carcinoma of the Mandible Metastatic to Skull

Christie L. Morgan, MS1; Scharukh Jalsi, MD, FACS2; Osamu Sakai, MD, PhD2; Brooke Denvenney-Cakir, MD; Shevta Arya, MD; Sarah Seo, MD2; Jeffrey H. Spiegel, MD, FACS2

1Boston University School of Medicine, Boston, MA
2Department of Otolaryngology - Head and Neck Surgery, Boston University School of Medicine, Boston, MA

Abstract

Educational Objectives: After reviewing this poster presentation, participants should be able to discuss the clinical manifestations of the rare tumor, ameloblastic carcinoma of the mandible, and describe its metastatic patterns. Study Design: Case Report. Methods: We report a very rare case of ameloblastic carcinoma of the mandible in a 16-year-old boy, who presented four years after definitive treatment with metastasis to the anterior skull. We review the presenting history, pathology, radiology, and management of a very rare tumor with an unusual metastatic pattern. Results: There are very few reported cases of ameloblastic carcinoma of the mandible. Metastases, when present, have most often been reported in the lung. Conclusions: Aggressive surgical intervention provides the best chance for survival. Metastases are known to occur in lung, brain, and distant bone. Routine, long-term surveillance is mandatory and should include physical examinations and serial imaging.

Background

Odontogenic tumors are an uncommon, complex group of benign and malignant lesions derived from odontogenic epithelium, ameloblastoma, or both, that exhibit diverse histological features and clinical behavior. Ameloblastoma is a common form of odontogenic tumor arising from odontogenic epithelium that is commonly cystic, slow growing, locally invasive, and recurrent with a typically benign course. In contrast, ameloblastic carcinoma is an extremely rare form of malignant odontogenic tumor arising from the odontogenic epithelium de novo (primary type) or by malignant transformation of a preexisting ameloblastoma (secondary type) with a typically aggressive course. Prior publications on ameloblastic carcinoma have encouraged continued reporting of this disorder as relatively little is known about the clinical nature of the tumor. We contribute a rare case of bony metastasis to the anterior skull.

Case

A 20-year-old Hispanic male with a past medical history significant for T4N0M0 ameloblastic carcinoma of the right mandible, treated with resection and reconstruction followed by radiation therapy in 2004, presented four years after definitive treatment with a two-week history of painless, rapidly enlarging, right-sided forehead swelling. He was otherwise asymptomatic, and denied fever, chills, weight loss, headache, and visual changes. Physical examination revealed a firm, nontender, nonpurulent, forehead mass just slightly to the right of the midline, measuring 5 cm in diameter. The overlying skin was mobile. Cranial nerves II-XII were intact with the exception of the right marginal mandibular nerve.

CT Sinus without contrast in axial (Figure 1) and coronal (Figure 2) views revealed an expansile, osteolytic lesion, eroding the anterior and posterior tables of the frontal bone in the midline. PET/CT demonstrates moderate FDG uptake (SUV 6.0), suggestive of malignancy. Fine-needle aspiration biopsy of the forehead mass was consistent with ameloblastic carcinoma.

Surgical excision of the forehead mass was planned after a multidisciplinary evaluation. The patient underwent a bifrontal craniectomy for resection of metastatic tumor via a bicoronal approach to the anterior cranial fossa. The tumor extended externally to involve the overlying subcutaneous tissue, and internally through the bone to involve the dura (Figure 3). Reconstruction of the dura was accomplished with the use of DuraMatrix (Figure 4) and a pericranial pedicled flap. A 9x9cm cranial defect was reconstructed with Leibinger mesh. A lumbar drain was placed.

Examination of the gross surgical specimen obtained in 2004 revealed a single, multilobulated firm white mass (Figure 5). Microscopic examination demonstrated sheets of atypical spindle epithelial cells separated by areas of hyalinized fibrous stroma, exhibiting nuclear pleomorphism with increased mitotic figures (4 Mitotic Figures/10 HPF) in the areas of increased cellularity. Immunohistochemical staining of the mass demonstrated positive staining of cells for cytokeratin AE1/3 and vimentin (Figure 6).

Examination of the gross surgical specimen obtained in 2008 revealed a circular segment of frontal bone with a tumor on the outer surface of the specimen measuring 6.9 x 5.9cm. Immunohistochemical studies performed on Paraffin embedded cell block tissue demonstrated positive staining of tumor cells for cytokeratin AE1/3 and vimentin (Figure 7). Tumor cells stained negative for synaptophysin, chromogranin, CD99, and S-100.

Discussion

The most recent WHO classification (2005) defines ameloblastic carcinoma as a rare odontogenic malignancy that combines the histological features of ameloblastoma with cytologic atypia, with or without metastasis. The differential diagnosis of ameloblastic carcinoma includes primary intra-alveolar epidermoid carcinoma, kerato-ameloblastoma, acanthomatous ameloblastoma, squamous cell carcinoma, and metastasis by tumor from adjacent tissue or distant visceral organs.

A review of the literature from 1927 to 2006 reveals 66 cases of ameloblastic carcinoma, 20 of which reported distant metastases, primarily to the lung. The literature is also notable for one case of ameloblastic carcinoma arising directly from the anterior skull base.

Histological features such as nuclear pleomorphism, hyperchromasia, increased mitotic figures, and the presence of a spindle-cell population suggest the diagnosis of ameloblastic carcinoma.

A complete understanding of the biological activity of ameloblastic carcinoma is limited by the scarcity of well-documented cases. Ameloblastic carcinoma is known to occur primarily in the mandible and to a lesser degree in the maxilla, in both sexes, all races, and over a wide range of age groups.

The clinical course of ameloblastic carcinoma is typically aggressive with extensive local destruction, lymph node involvement, and both early and late patterns of metastasis to sites including the lung, brain, and distant bone.

Conclusions

Management of ameloblastic carcinoma requires a multidisciplinary approach to determine proper treatment.

Aggressive surgical intervention provides the best chance for survival.

Wide local excision is the treatment of choice for ameloblastic carcinoma, and cervical lymph node dissection should be considered when there is obvious lymphadenopathy.

Metastases are known to occur in lung, brain, and distant bone.

Radiotherapy and chemotherapy has been used in cases of metastatic disease that are not amenable to surgical resection, but have limited value as primary modalities for the treatment of ameloblastic carcinomas.

Routine, long-term surveillance is mandatory and should include physical examinations and serial imaging.

Advanced imaging techniques play an essential role in the diagnosis, surgical planning, treatment, and surveillance of ameloblastic carcinoma.

References