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Neuroradiology/Head and Neck Imaging *Case Report*

Ameloblastic carcinoma with hepatic metastases: A case report and review of ameloblastomic carcinoma

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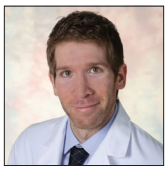

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ABSTRACT

Ameloblastic carcinoma is a locally aggressive odontogenic tumor that most commonly affects young and middle-aged adults. Metastatic disease may develop insidiously and manifest months or years after the initial diagnosis. Herein, we describe the clinical, imaging, and pathologic findings of a 31-year-old male who presented to the emergency department with headache and vision loss of 3 months duration and was subsequently found to have ameloblastic carcinoma with hepatic metastases. Initial computed tomography (CT) and magnetic resonance imaging revealed a multilocular cystic mass with avidly-enhancing nodular soft-tissue components associated with the right temporal fossa. Histologic examination of a tissue sample showed findings consistent with ameloblastic carcinoma. An initial staging CT scan showed several small hepatic cystic lesions. Follow-up surveillance imaging showed interval growth. A subsequent biopsy of a hepatic lesion showed findings compatible with metastatic ameloblastic carcinoma. The patient was started on systemic chemotherapy with evidence of disease progression at 1-year follow-up.

Keywords: Ameloblastoma, Ameloblastic carcinoma, Metastatic ameloblastoma, Metastatic ameloblastic carcinoma, Odontogenic tumors

INTRODUCTION

Ameloblastic carcinoma is an uncommon odontogenic tumor that typically arises within the mandibular molars or along the mandibular angle. Early middle-aged individuals are most commonly affected, with a median age at diagnosis of 30.5 years.[1] Tumors are often locally aggressive and require prompt surgical resection with wide margins. Metastatic ameloblastic carcinoma is exceedingly rare and almost exclusively involves the locoregional lymph nodes or lungs. However, osseous, intracranial, and hepatic metastases may occur at later stages of disseminated disease.[2] Herein, we report a case of malignant ameloblastic carcinoma with isolated hepatic metastasis. In addition, we review the clinical, imaging, and histologic features of ameloblastic carcinoma and summarize other cases of ameloblastic carcinoma with hepatic metastases.

CASE REPORT

A 31-year-old male with a reported history of a recurrent jaw tumor, status postadjuvant chemoradiation, and multiple resections, presented to the emergency department complaining

of a persistent right-sided headache and vision loss of 3 months duration. The patient reported that he had first been diagnosed with a tumor under his right wisdom tooth 6 years earlier and had undergone multiple resections with hardware placement and reconstructive surgery. Maintenance chemotherapy had been initiated following the most recent resection, but the patient self-discontinued after several months of treatment.

Vital signs on presentation were within normal limits. Physical examination was significant for a mass encompassing the right aspect of the forehead, orbit, and temple with deformity of the right aspect of the hemimandible and overlying scarring and volume loss. Vision and extraocular movement was grossly intact in all fields. Laboratory studies were normal.

A non-contrast computed tomography (CT) scan of the head was obtained and showed a large, heterogeneous mass involving the right masticator space and extending superolaterally along the right temporal fossa with destruction of the right middle cranial fossa floor [Figure 1]. The patient was admitted to the hospital for further evaluation. Magnetic resonance imaging (MRI) of the face showed a large, multilocular cystic mass with avidly-enhancing nodular soft-tissue components associated with the right temporal fossa and extending medially to involve the clivus, sphenoid buttress, and greater wing of the sphenoid. There were additional pericentimeter, avidly-enhancing masses within the posterolateral aspect of the right globe, right frontal calvarium, and right optic strut [Figure 2]. An ultrasound-guided percutaneous biopsy was performed. Histologic examination of a tissue sample revealed numerous singly dispersed, small and large groups of cells with hyperchromatic nuclei, high nuclear: cytoplasmic (N: C) ratio, prominent nucleoli, and irregular nuclear contour and scant cytoplasm with palisading on a background of histiocytes. A diagnosis of ameloblastic carcinoma was established.

Resection of the tumor was subsequently performed using a neurosurgery-otolaryngologist collaborative approach. Intraoperative visualization revealed that the mass had eroded through the floor of the middle cranial fossa and extended extracranially. In addition, an expansile component of the mass had invaded the ipsilateral infratemporal fossa. Resection of the middle cranial fossa and extracranial elements was performed by the neurosurgery team in an en bloc fashion using a combination of dissection, bipolar electrocautery, and microscissors with a margin of 1 cm. The infratemporal components of the tumor were resected with the assistance of otolaryngologists, who carefully dissected around the facial nerve and temporal artery. Histologic examination of frozen sections confirmed negative margins. Reconstruction was performed using a radial forearm free flap.

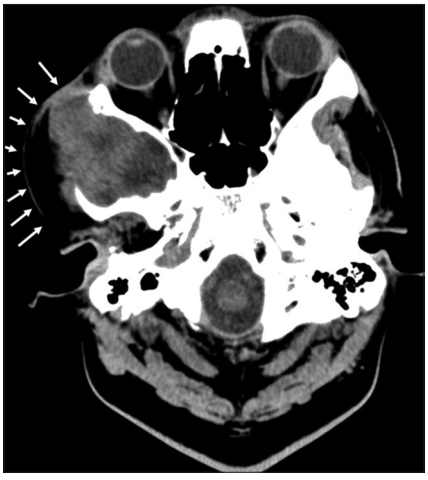

Figure 1: A 31-year-old male with a history of a jaw tumor presented with the right-sided headache and vision loss. Axial view of a non-contrast CT scan of the head demonstrates a large, heterogeneous mass involving the right masticator space and extending superolaterally along the right temporal fossa (arrows).

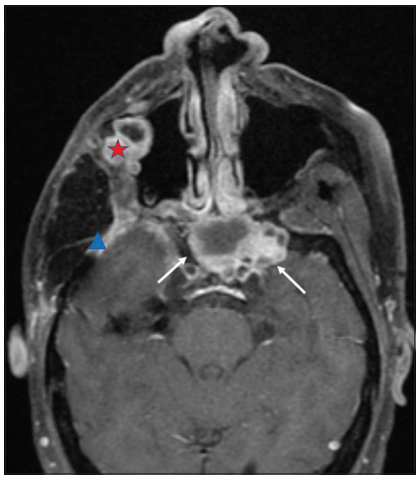

Figure 2: A 31-year-old male with a history of a jaw tumor presented with the right-sided headache and vision loss and subsequently found to have a large, heterogeneous mass involving the right masticator space on CT scan. Axial view of a contrast-enhanced MRI of the face performed for further characterization shows a large, multilocular cystic mass with avidly-enhancing nodular soft-tissue components associated with the right temporal fossa and extending medially to involve the clivus (white arrows), sphenoid buttress (blue triangle), and greater wing of the sphenoid (red star).

A subsequent staging CT scan of the chest showed no evidence of pulmonary nodules, osseous lesions, or locoregional lymphadenopathy. However, there were several small cystic lesions within the imaged portion of the liver, one of which demonstrated attenuation slightly greater than simple fluid [Figure 3]. MRI of the abdomen was performed for further characterization and showed multiple scattered cystic lesions throughout the liver, several of which exhibited irregular margins and peripheral enhancement concerning for hepatic metastases.

The patient was started on carboplatin and paclitaxel for suspected malignant ameloblastoma. A follow-up CT scan of the chest, abdomen, and pelvis obtained 3 months later showed a marked increase in size of the previously demonstrated hepatic lesions [Figure 4a and b]; the lesions demonstrated avid peripheral enhancement on MRI [Figure 5a and b]. Tissue sampling of a lesion within the left lateral hepatic lobe showed crowded sheets and singly dispersed atypical cells with a high N: C ratio admixed with another population of atypical cells showing clear cytoplasm and eccentric hyperchromatic nuclei in a pattern consistent with metastatic ameloblastoma. Samples were sent for further tumor profiling, including programmed death ligand-1, tumor mutational burden, and neurotrophic tyrosine receptor kinase. However, no immunotherapeutic targets were identified. The patient remains on carboplatin and paclitaxel therapy with recurrence of the primary tumor and progression of metastatic disease at 1-year follow-up [Figure 6].

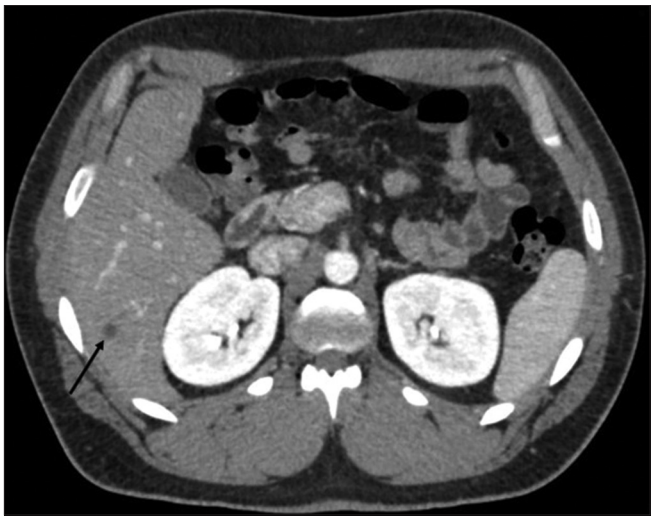

Figure 3: A 31-year-old male presented with a jaw tumor and was subsequently diagnosed with ameloblastic carcinoma. Axial view of a CT scan of the chest performed for staging reveals several small cystic lesions within the imaged portion of the liver, including a lesion in the posterior right hepatic lobe with irregular margins and attenuation slightly greater than simple fluid (black arrow).

DISCUSSION

Ameloblastic carcinoma is rare malignant epithelial odontogenic tumor with a predilection for the maxilla and mandible. Neoplasms share several clinicopathologic features with benign and malignant ameloblastoma. However, ameloblastic carcinoma is far more aggressive and demonstrates a distinct pattern of cytologic atypia and squamous metaplasia. In addition, cross-sectional imaging of ameloblastoma typically reveals a lucent lesion with well-defined borders and an intact lamina dura, whereas ameloblastic carcinoma is characterized by ill-defined borders and destruction of the lamina dura.[3] The imaging differential diagnosis for ameloblastic carcinoma is broad and includes both benign and malignant odontogenic tumors, including dentigerous cysts, central giant cell granuloma, and calcifying epithelial odontogenic tumor.

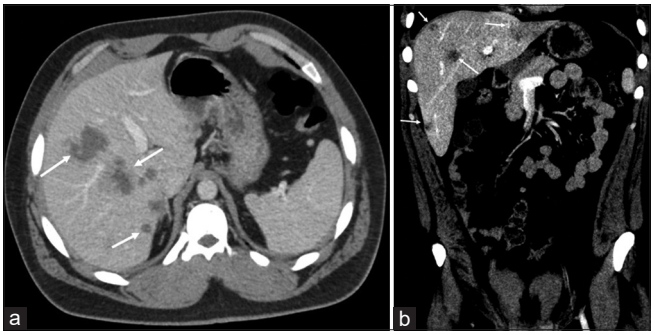

Figure 4: A 31-year-old male presented with a jaw tumor and was subsequently diagnosed with ameloblastic carcinoma. Initial staging CT scan of the chest showed several small cystic hepatic lesions, including a suspicious cystic hepatic lesion in the posterior right hepatic lobe. A CT scan of the abdomen and pelvis was performed 3 months later. Axial view (a) shows rapid interval growth of the cystic hepatic lesions (white arrows). Coronal view (b) illustrates the extent of disease, with hepatic lesions in segments 2, 5, and 8 (white arrows).

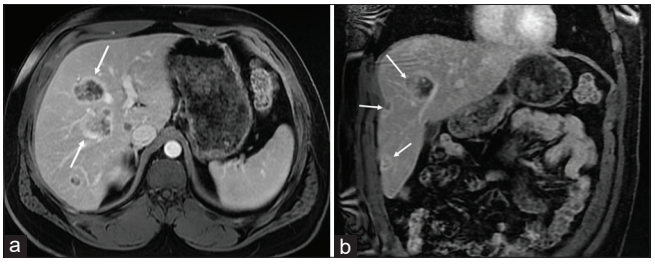

Figure 5: A 31-year-old male presented with a jaw tumor and was subsequently diagnosed with ameloblastic carcinoma with suspected hepatic metastases. Axial view (a) of a contrast-enhanced MRI of the abdomen demonstrates multiple cystic hepatic lesions with irregular morphology and avid peripheral enhancement (white arrows). Coronal view (b) shows extensive disease involving multiple hepatic segments and abutting branches of the hepatic vein (white arrows).

Ameloblastic carcinoma metastases are rare and typically involve the locoregional lymph nodes or lungs. However, visceral metastases have also been described. In a 1974 report by Sedhdev *et al.*, authors describe a 36-year-old male who presented with mandibular ameloblastoma and later developed isolated hepatic metastases. In a more recent review of 65 cases of metastatic ameloblastoma, Dissanayake *et al.* report five cases of hepatic metastases; isolated hepatic metastases were present in only one patient.[4] Additional cases of ameloblastoma and ameloblastic carcinoma with hepatic metastases are summarized in [Table 1].[5-10] Notably, although amelobastic carcinoma may arise within the maxilla or mandible, all cases of ameloblastic carcinoma with hepatic metastases have been reported to originate in the mandible.

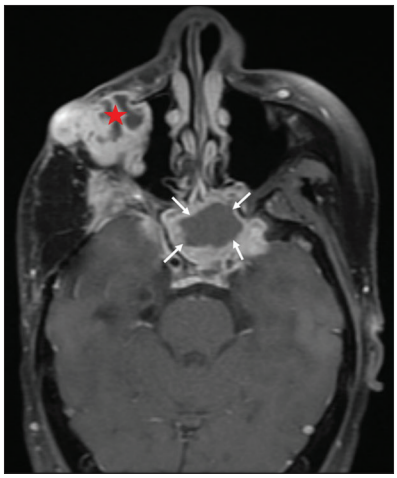

Figure 6: A 31-year-old male presented with a jaw tumor and was subsequently diagnosed with ameloblastic carcinoma with suspected hepatic metastases. Contrast-enhanced MRI obtained at 1 year follow-up shows recurrence of the primary tumor with an increased size of an irregular central cystic component involving the clivus (white arrows) and invasion of the right maxillary sinus (red star). Hepatic metastases had also increased in size (not shown).

There is a paucity of data describing the imaging features of visceral ameloblastic carcinoma metastases. The existing medical literature suggests that visceral metastases are highly variable in appearance: Heterogeneously enhancing solid masses, mixed cystic and solid lesions, and complex cysts have all been described.[5-8] In a recent case by Collins *et al.*, the patient presented with a large complex cyst with septations, initially thought to represent bacterial or parasitic infection; a definitive diagnosis was only established by cytology. In our patient, hepatic cystic lesions appeared similar to simple cysts, but measured greater than fluid attenuation on CT scan and demonstrated slightly irregular borders on MRI. Given the high degree of morphologic variability, we propose that hepatic lesions in a patient with a known diagnosis of ameloblastoma or ameloblastic carcinoma be regarded with a high degree of suspicion for metastatic disease. Positron emission tomography (PET)/CT may be of value to establish a diagnosis.[10] The presence of metastatic disease may influence treatment and affect patient candidacy for aggressive resection, hemimandibulectomy, or hemimaxillectomy.

Table 1: Summary of patients presenting with ameloblastic carcinoma or ameloblastoma with liver metastases.[5-10]

Age*	Sex	Primary site	Metastases	Treatment	Outcome	Ref
9	M	Mandible	Lung, hilar lymph nodes, diaphragm, and liver	Not reported	Death at 10 years after initial diagnosis	5
23	M	Mandible	Lung and liver	Adjuvant cisplatin and adriamycin followed by resection of primary tumor and metastases	Disease-free at 15-month follow-up	6
25	M	Mandible	Liver	Resection of the primary tumor followed by carboplatin and paclitaxel for metastatic disease	Increasing number of metastases at 12-month follow-up	CR †
36	M	Mandible	Liver	Curettage followed by hemimandibulectomy	Death at 17 years after initial diagnosis	7
38	F	Mandible	Locoregional lymph nodes and liver	Multiple resections of the primary tumor	Death 6 years after initial diagnosis	8
39	F	Mandible	Lung and liver	Declined treatment	Death 13 years after initial diagnosis	9
74	M	Mandible	Bones, lung, and liver	Enucleation of primary tumor; no systemic treatment administered for metastases	Not reported	10

*Age at initial diagnosis of the primary tumor. † Current report

Surgical resection represents the mainstay of the management for localized ameloblastic carcinoma. However, there are no clinical practice guidelines for the management of metastatic disease. Multiple attempts have been made to establish a safe and effective chemotherapy regimen, including regimens comprised of paclitaxel, cyclophosphamide, doxorubicin, 5-fluorouracil, adriamycin, methotrexate, and/or platinum-based agents. However, as of this writing, chemotherapy appears to have limited value for both local and metastatic disease.

CONCLUSION

Ameloblastic carcinoma is an uncommon, locally aggressive odontogenic tumor. Metastatic disease is exceedingly rare and typically affects the locoregional lymph nodes and lungs. However, as in our patient, isolated visceral metastases can occur. Hepatic lesions in patients with a known diagnosis of ameloblastic carcinoma should be regarded with a high index of suspicion for metastatic disease. Short interval follow-up and PET/CT may be of value for further characterization in equivocal cases. Prompt recognition and management of metastases is essential to guide management and optimize patient outcomes.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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