Ameloblastic carcinoma is a rare malignant odontogenic carcinoma that has metastatic potential, and because of its rare incidence, there are few reports focusing on its radiologic imaging. If it shows aggressive appearances, it can be diagnosed as malignant tumor. But in case of negative appearance, it is difficult to distinguish ameloblastic carcinoma from ameloblastoma. We report a case of ameloblastic carcinoma of the maxilla in a 76-year-old female patient with radiologic images and pathologic features. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011;112:e40-e47)

Ameloblastic carcinoma, a rare odontogenic carcinoma, is divided into primary type; secondary type, intraosseous; and secondary type, peripheral in the World Health Organization (WHO) classification in 2005. 1-5 Ameloblastic carcinoma was classified as malignant ameloblastoma in odontogenic carcinomas in the WHO classification in 1972. 6 Thereafter, Elzay 7 and Slootweg and Müller 8 distinguished ameloblastic carcinoma from malignant ameloblastoma.

Because ameloblastic carcinoma is a rare occurrence, 1-4 many examples are case reports. 9-12,14-35 However, some authors have reported literature reviews. 9-17,30 According to them, ameloblastic carcinoma has different clinical features in the maxilla and in the mandible and has a lower incidence in the maxilla than in the mandible. 9-11 The radiologic features of ameloblastic carcinoma are variously reported, and there are some differential diagnoses, such as cysts, benign tumors, other malignant tumors, and metastatic tumors. 9-24,26,28,30-35 Ameloblastic carcinoma tends to be aggressive and may involve lymph nodes and distant metastasis. 9-20,22,24-28,30-35 Therefore, diagnostic imaging before treatment is very important, but previous reports are mostly related to clinical courses and pathologic features. 9-18,20,30 In particular, there are no reports about 18F-fluorodeoxyglucose–positron emission tomography (FDG-PET) of the primary site, and only a few images with magnetic resonance imaging (MRI) and computerized tomography (CT) have been presented. 9-12,14-17,19,21,22,24,26-28,35

In the present report, we show various imaging characteristics of FDG-PET, MRI, including dynamic study, CT, and conventional radiographs in a case of ameloblastic carcinoma of the maxilla.

CASE REPORT

A 73-year-old woman referred to a general dental practitioner in 2007 complaining of a painless swelling of right cheek. Her primary doctor diagnosed a residual cyst of the maxillary premolar region on the basis of a panoramic radiograph and then followed her. In July 2009, she referred to Okayama University Hospital because she felt a sense of discomfort in the right cheek. Her medical history revealed hypertension, arrhythmia, and cataract. Intraoral examination revealed a painless, bone-like swelling in the right anterior maxilla (Fig. 1).

Conventional radiographs showed a unicocular cystic lesion with well defined margins which elevated the floor of the right anterior maxillary sinus, and a tooth-like radiopaque body in the alveolar process of the right premolar region (Fig. 2). Contrast-enhanced CT with iohexol was performed to differentiate between cystic lesions as a residual cyst and benign tumors as an ameloblastoma. Axial CT (bone window)
image revealed a globular-shaped lesion arising from the inside of maxillary bone with a slight expansion and anterior bone resorption of the anterior maxillary sinus (Fig. 3, A). Contrast-enhanced axial CT image showed a heterogeneous enhancement of the lesion (Fig. 3, B). On reconstructed coronal CT image, the mass showed the diffuse bone partly resorption of alveolar bone and elevated anterior maxillary sinus floor with destruction of a part of the floor and all of the nasal side wall (Fig. 3, C). These findings of marginal bone destruction of the lesion in some parts had the possibility of malignancy, but we assigned the diagnosis of a benign tumor, such as ameloblastoma, because the margin of the lesion was almost well defined. Based on clinical diagnosis of benign tumor, the incisional biopsy was performed a few days later and showed that the lesion was suspected ameloblastic carcinoma. Contrast-enhanced MRI with gadodiamide including dynamic study was performed 12 days later to evaluate soft tissue invasion at the maxillary sinus and nasal cavity. Furthermore, we performed FDG-PET/CT to investigate the presence of regional lymph nodes and distant metastasis 14 days after the biopsy.

The MR images showed the mass consisting of a solid component, which had a predilection for isointensity on T1-weighted image (T1WI), hyperintensity on short TI inversion recovery (STIR) image, and enhancement on contrast-enhanced T1WI, and liquid components, which had a predilection for homogeneous isointensity on T1WI, homogeneous hyperintensity on STIR, and no enhancement on contrast-enhanced T1WI. These findings were similar to ameloblastoma (Fig. 4). Contrast index (CI) curve was created using dynamic images to observe the flow pattern of contrast medium into the tumor mass. The region of interest (ROI) was essentially drawn to include the maximal region of solid portion excluded the cystic portion by free hand drawing using cursor on monitor. The mean signal intensity (SI) of the ROI of each lesion was calculated on a workstation (Synapse Vincent; Fujifilm Medical Co.). The CI was calculated using the following formula: CI = (signal intensity (postcontrast) − signal intensity (precontrast))/signal intensity (precontrast).

Then the CI was plotted on a time course to obtain the CI curve. The CI curve of our case showed gradual increase, reached a plateau at 90-120 seconds, and nearly sustained the plateau to 800 seconds (Fig. 5). This appearance also was similar to ameloblastomas.

On FDG-PET imaging, elevated FDG uptake was found in the right maxilla (maximum standardized uptake value (SUVmax) 28.3; Fig. 6, A and B). No abnormal high uptake suggesting the distant metastasis was observed on FDG-PET images (Fig. 6, C). On the basis of these imaging findings, the patients were diagnosed with ameloblastic carcinoma (stage IV, T4N0M0) and underwent tumor resection and split-thickness skin grafting. After the surgery, the diagnosis was confirmed histopathologically.

Histologic findings revealed that the tumor mass dominated in the maxillary bone with giant cyst formation (Fig. 7, A). The tumor exhibited massive bony destruction and progressed into the maxillary sinus, invading into gingival connective tissue and subcutaneous tissue of the cheek region (Fig. 7, B).
Tumor cells with hyperchromatism and nuclear polymorphism formed anastamosing strands with edematous stroma, exhibiting plexiform pattern in some areas (Fig. 7, C). In another area, the tumor cells presented follicular growth pattern with desmoplastic stroma, which consisted of proliferation of hyalinized collagen fibers. The follicular nests consisted of palisaded columnar cells located in the outer layer and polygonal or loosely arranged stellate reticulum–like cells located in the inner layer. In the tumor nests, focal keratinization and/or comedolike necrosis was observed (Fig. 7, D). Histopathology of the tumor was similar to the plexiform-type or follicular-type ameloblastoma but accompanied by strong cellular atypia, necrosis, apoptosis, and mitoses, indicating its malignant features. Moreover, the tumor lacked the finding of ordinary ameloblastoma. Immunohistochemical stains demonstrated high positive ratios of p53 and Ki-67 in the tumor cells (Fig. 8). The lesion was finally diagnosed as ameloblastic carcinoma “primary type” based on the histopathologic and immunohistochemical findings.

In the follow-up a year after surgery, the patient was free of recurrence and metastasis.

DISCUSSION

Ameloblastic carcinoma is a rare disease and its incidence has been reported as 1%-3%.1-4 Ameloblastic carcinoma has a high incidence in men twice than that in women; maxilla and mandible location differ in average age, the 5th decade for the maxilla, which is ~10 years older than for the mandible.9-11 The mean age for ameloblastic carcinoma tends to be higher than
Fig. 5. Contrast index curve of ameloblastic carcinoma shows the pattern that increases relatively, reaches a plateau at 90-120 seconds, and then nearly sustains the plateau to 800 seconds.

Fig. 4. Magnetic resonance images in the coronal plane. A, T1-weighted image shows hypo- to isointensity with heterogeneous pattern. B, Short TI inversion recovery (STIR) image shows heterogeneous hyperintensity with a marked high signal spot. C, On contrast-enhanced T1-weighted image, the tumor shows well enhancement heterogeneously outside the region which shows marked hyperintensity on STIR image.

Fig. 5. Contrast index curve of ameloblastic carcinoma shows the pattern that increases relatively, reaches a plateau at 90-120 seconds, and then nearly sustains the plateau to 800 seconds.
for ameloblastoma, which affects the genders and jaws almost the same.\textsuperscript{36-39}

The clinical symptoms of ameloblastic carcinoma are variable, such as gingival swelling with or without ulceration, rapid growth of mass, perforation of the cortex, pain, and paresthesia.\textsuperscript{9-16,30} These symptoms of ameloblastic carcinomas are more aggressive than ameloblastomas.\textsuperscript{9,11,36-40}

Radiographic features of ameloblastic carcinomas were similar to those of ameloblastomas, and some authors have reported the presence of the some focal radiopaque body of dystrophic calcifications.\textsuperscript{11,14,15,18,19} The radiographic appearance of our case was almost the same as benign tumors and cysts and had no dystrophic calcification in radiographic and histologic features.

We wrongly diagnosed our case as benign tumor, because the occurrence of ameloblastic carcinoma is rare, it had a normal-appearing mucosal surface without ulceration, and the margin of the lesion was almost well de-
fined although partly destructed on CT images. However, retrospectively, we recognized the bone resorption of alveolar bone and walls of anterior maxillary sinus due to bone invasion of the tumor as a characteristic finding suggestive of malignancy. These findings are not commonly observed in benign tumors and cysts. And such small changes of bone were more clearly seen in the CT images compared with conventional radiographs. On the basis of contrast-enhanced CT images, the lesion could be diagnosed with tumorous lesions because the inner part of the mass was enhanced.

MRI could more clearly show internal state of the lesion, as mentioned by many authors, including us. In the present case, the lesion showed isointensity and slight hyperintensity on T1WI, and slight hypointensity, hyperintensity, and marked hyperintensity on STIR image. Contrast-enhanced T1WI showed inhomogeneous enhancement with nonenhancement area of liquid component which was a markedly hyperintense area on STIR image. These patterns of signal intensities were not specific to ameloblastic carcinoma, although they are consistent with the gross features of the resected tumor and the MRI findings of ameloblastoma. We have previously reported dynamic contrast-enhanced MRI of ameloblastomas. We reported that the CI curves of ameloblastomas show 2 patterns: one increases, reaches a plateau at 100-300 seconds, then sustains the plateau or decreases gradually to 600-900 seconds, and the other increases relatively rapidly, reaches a plateau at 90-120 seconds, then decreases relatively rapidly to 300 seconds and decreases gradually thereafter. This case of ameloblastic carcinoma was consistent with the latter pattern of CI curve, so it was difficult to differentiate ameloblastic carcinoma from ameloblastoma.

Fig. 7. A, Macrographic view of hematoxylin-eosin staining shows that the tumor cells formed an intrabony lesion with giant cyst formation. B, The tumor showed osteolytic growth. The anterior maxillary sinus wall was destroyed and extravasation into submucosal tissue was observed. C, Tumor cells formed plexiform growth with loose edematous stroma and D, follicular tumor nest with cystic degeneration and necrosis accompanied by dense collagenous stroma.
Ameloblastic carcinomas are known to have occurrences of metastasis, with the site of predilection being the lung and other sites including the cervical lymph nodes, brain, bone, soft tissue, and liver.\textsuperscript{9-11,13,15,18-20,24,30-35} FDG-PET is a useful modality for evaluation of malignant tumor as well the primary site as lymph nodes and distant metastasis. However, as far as we ranged extensively over the literature, there was only 1 article about FDG-PET of ameloblastic carcinoma.\textsuperscript{35} Moreover, the article described only the skull and lung metastasis and not the primary site. They reported that the FDG accumulations were observed in the skull (SUV max 6.0) and lung (SUV max 2.0) metastasis. In our case, we observed the strong FDG uptake (SUV max 28.3) in the primary tumor, although there were no abnormal FDG accumulations suggesting the metastasis. Because the ameloblastic carcinoma has a potential of distant metastasis with or without cervical lymph node metastasis, it is useful to use PET as the initial staging method before surgery.

Histologic findings revealed that the tumor exhibited an osteolytic solid mass with multiple cyst formation. The tumor cells demonstrated plexiform growth and/or stellate reticulum–like follicular growth pattern, resembling ameloblastoma. However, it lacked benign ameloblastoma features, and the tumor cells showed strong cellular atypia, hyperchromatism, necrosis, and apoptosis. Moreover, immunohistochemistry revealed high positive ratios for p53 and Ki-67. Taking these together, we diagnosed the tumor as ameloblastic carcinoma (primary type), according to the WHO classification revised in 2005.

Ameloblastic carcinoma is a very rare intrabony neoplasm and includes metastasizing ameloblastoma and ameloblastic carcinoma (primary/secondary).\textsuperscript{5} Differential diagnosis of ameloblastoma and ameloblastic carcinoma is sometimes controversial, because ameloblastoma will show osteolytic grows although it is benign. Therefore, the findings of cellular atypia or necrosis in the tumor nest and immunohistochemical analysis for p53 and Ki-67 might be the key to differentiate the two.

CONCLUSIONS

We have reported a case of ameloblastic carcinoma of the maxilla in a 73-year-old female patient. In our case, it was difficult to distinguish between tumorous lesion and cystic lesion by conventional radiographs. CT and MRI findings suggested it to be tumor but were nonspecific to ameloblastic carcinoma. FDG-PET/CT was useful for ascertaining the presence of regional lymph node and distant metastasis. In reviewing our case, CT was useful for distinguishing ameloblastic carcinoma from ameloblastoma, because it could observe bone resorption clearly.

REFERENCES


