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Case report

Ghost cell odontogenic carcinoma transformed from dentinogenic ghost cell tumor of the maxilla after recurrences

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ABSTRACT

Ghost cell odontogenic carcinoma (GCOC) is a rare malignant odontogenic tumor with aggressive growth characteristics. We report a case of GCOC of the maxilla in a 65-year-old Japanese man who was referred to our hospital with painful swelling of the left maxilla. Computed tomography showed a bone defect in the left upper jaw and the border with the surrounding tissue was indistinct in one area. Tumor resection was performed under general anesthesia, and the histopathological diagnosis was dentinogenic ghost cell tumor (DGCT). Ten months postoperatively, the tumor recurred in the same area, and partial resection of the maxilla was performed. Twelve months after reoperation, the tumor recurred a second time. The histopathological diagnosis this time was GCOC, as a secondary malignant manifestation of the benign DGCT.

1. Introduction

Ghost cell odontogenic carcinoma (GCOC) is a malignant odontogenic epithelial tumor with features of calcifying cystic odontogenic tumor (CCOT) and/or dentinogenic ghost cell tumor (DGCT) [1]. Despite aggressive surgical resection, GCOC demonstrates a high recurrence rate. Because GCOC is very rare, no standard treatment has yet been established. We describe here in a case of GCOC arising from recurrent DGCT of the maxilla, and treated with surgery and adjuvant carbon ion therapy.

2. Case report

A 65-year-old Japanese man visited our hospital with painful swelling of the left maxilla. The patient was otherwise in good health. No enlarged cervical lymph nodes were found. Clinical examination revealed an undefined, hard mass from the left maxillary first premolar to the first molar with no ulceration of the surface mucosa. Panoramic X-ray revealed a well-defined radiolucent lesion in the left upper molar, with no root resorption. Computed tomography (CT) revealed a 40-mm soft tissue lesion with resorption of the adjacent maxillary bone and sinus, without neck or distant metastases. The border with the surrounding tissue was indistinct in one area. Calcific substances weren' t ditected (Fig. 1A). Magnetic resonance imaging (MRI) showed a lesion with cystic and solid portions (Fig. 1B). The cystic portion showed homogeneous intermediate signal on T1-weighted imaging (WI), signal hyperintensity on T2WI, and no enhancement on Gd-enhanced T1WI. The solid portion showed intermediate-low signal on T1WI, high signal in T2WI, and enhancement on Gd-enhanced T1WI.

Puncture was performed before biopsy, and 10 ml of fluid was found in the lesion. When intraoral incisional biopsy was performed, the cystic wall-like tissue in contact with the oral mucosa was resected. Pathological examination suggested CCOT. The maxillary neoplasm was resected under general anesthesia. The lesion, surrounding soft tissues, alveolar bone, and front wall of the maxillary sinus were removed surgically using an oral approach. For the part in contact with bone, the lesion was resected with 3-mm margin from the bone resorption. The part in contact with soft tissue was resected including the induration. The pathological diagnosis was DGCT, and the tumor was excised with clear margins.

Ten months after the first operation, local recurrence was suspected. CT showed an ill-defined, radiolucent lesion (Fig. 1C). Maxillary segmental resection was performed using an extra-oral approach (modified Weber-Fergusson incision) with a 5-mm safety

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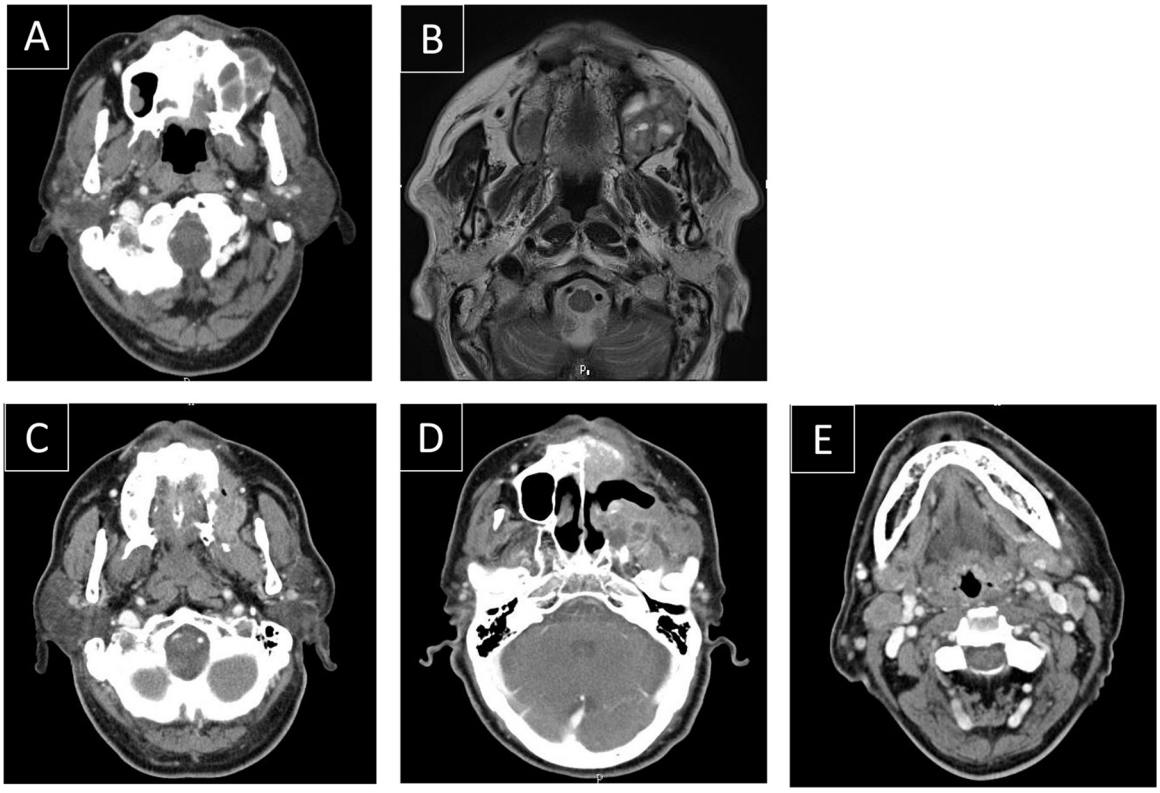

Fig. 1. A) At initial diagnosis, CT shows a well-defined mass in the left maxilla with resorption of the maxillary bone and sinus. B) T2-weighted MRI shows the cystic and solid portions of the lesion. C) At the time of recurrence, CT shows an ill-defined mass demonstrating variable densities and bony erosion of the maxilla. D) At the second recurrence, CT shows two poorly demarcated masses extending into the left maxillectomy cavity. E) Bilateral cervical regions show a large number of lymph node metastases.

margin. The tumor mass, left front wall of the maxillary sinus, palatal bone, surrounding tissues, and nasal mucosa were removed surgically. The histopathological diagnosis was DGCT, and no malignant characteristics were evident.

Twelve months after the reoperation, the tumor recurred a second time. The lesion mass was in the left maxillary midline in the front, and at the processus pterygoideus and fossa pterygopalatina in the rear (Fig. 1D). The safety margin was set at ≥5 mm from the identified margin of forward tumor advance. However, in the area extending backward, resection with adequate safety margins was difficult, the soft-tissue lesion was resected and the bone surface was shaved off. Tumor resection was planned to be performed in combination with adjuvant chemoradiotherapy. The histopathological diagnosis was transformation to GCOC from DGCT. The patient received 64 Gy/16 F/4 weeks of carbon ion therapy for local control. Because the effects of chemotherapy on GCOC were unclear, adjuvant chemotherapy was not provided.

Six months after the second operation, CT showed lymph node metastases to bilateral cervical regions (Fig. 1E). Right-sided extended supraomohyoid neck dissection and left-sided radical neck dissection were performed. The patient received 50.4 Gy/28 F/6 weeks of external-beam radiotherapy to both sides of the neck.

The primary tumor and cervical lymph node metastases were controllable, but 6 months after neck dissection the patient showed distant metastases to the left chest, left ilium, thoracic vertebrae and lumbar vertebrae. He died 3 years and 10 months after initial diagnosis.

The primary resection specimen (Fig. 2) showed solid growth of ameloblastoma-like islands, which had locally invaded into surrounding bone tissue. Part of the lesion was a cystic structure. The basal cells were columnar and arranged in a palisaded fashion. On the luminal side, tumor cells and ghost cells showed colonization. The tumor also showed dysplastic dentin. Immunohistochemistry showed a Ki-67 labeling index (LI) of 4.1% and a p53-positive rate of 1.6%. Based on these findings, DGCT was diagnosed. The specimen resected at reoperation resembled that from the primary operation. Ki-67 LI was 4.4% and p53-positive rate was 1.1%. The specimen resected at the second recurrence (Fig. 3A) showed a solid tumor with severe atypical epithelial cells and ghost cell proliferation. The tumor comprised columnar cells, ghost cells, and acidophilic tumor cells. Tumor cells showed severe atypia, such as atypical mitosis, hyperchromasia, and anisonucleosis (Fig. 3B). The entirety of the resected tissue was carefully examined, but no DGCT tissue was apparent. Based on these findings, GCOC due to malignant change in the DGCT was diagnosed. Ki-67 LI was 2.9%. GCOC was identified within lymph nodes. The metastatic GCOC showed features similar to those of the maxillary region, a 22.8% Ki-67 LI and a 36.2% p53-positive rate.

3. Discussion

GCOC and DGCT were diagnosed based on the 2005 World Health Organization (WHO) guidelines [1]. DGCT is a rare odontogenic tumor classified as a neoplastic variant of CCOT. GCOC is a particularly rare malignant counterpart of CCOT or DGCT. The characteristic of GCOC is aggressive and destructive behavior.

Only 40 cases of GCOC have been reported in the literature to date, including this case [2–31]. The results of a systematic analysis of the reported cases of GCOC are summarized in Table 1. GCOC occurs more commonly in men than in women (male:female = 31:9), and patients have ranged in age from 10 to 89 years. The maxilla is affected more often than the mandible, with 27 cases in the maxilla, and 13 cases in the mandible. The most common complaint of patients was swelling, with or without pain.

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