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Case Report

A case of a huge mandibular tumor composed of ameloblastoma and high-grade sarcoma

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ABSTRACT

Ameloblastoma is a benign odontogenic tumor of the jaw. Sarcoma is a rare neoplasm that accounts for only about 1% of all tumors. Here, we report a quite rare case of simultaneous occurrence of an ameloblastoma and a high-grade sarcoma in the mandible of a 65-year-old man. He was referred to our hospital because of his difficulties in eating and drinking due to a remarkable swelling of the right mandible. He had first noticed the swelling about six years before and consulted a clinic one year before, but he had refused any examination or treatment. He underwent an incisional biopsy, and it revealed a high-grade spindle cell sarcoma with massive necrosis. Therefore, we suggested surgery, but he had never hoped any operation. His poor nutritional status gradually further declined, and he died due to massive hemorrhage from the tumor. In the autopsy, the histological findings revealed that the tumor was predominantly composed of high-grade spindle cell sarcoma and contained a small amount of epithelial component which was a granular cell variant of ameloblastoma. Metastatic tumors were seen in the bilateral lungs and the liver. They contained only the sarcomatous element.

1. Introduction

Ameloblastoma is one of the most common odontogenic tumors of the jaw. This tumor is histologically benign but locally invasive and can cause severe expansion of the cortical bone and gross anatomical deformities. Clinically, ameloblastoma and related tumors can be classified broadly into five groups: unicystic, extraosseous/peripheral, metastasizing, ameloblastic fibroma, and malignant. There are two types of malignant tumors related to ameloblastoma, namely ameloblastic carcinoma and odontogenic sarcomas, also termed ameloblastic fibrosarcoma.

In the present report, we describe a rare case of a huge tumor in the right mandible, which represents the simultaneous occurrence of an ameloblastoma and a high-grade sarcoma which cannot be classified as ameloblastic fibrosarcoma.

2. Case report

A 65-year-old man was referred to our hospital, in June 2015, because of his difficulties in eating and drinking. He had a huge mass in the right side of the mandible (Fig. 1). He had first noticed a swelling in this region about 6 years before and consulted a clinic one year before, but he refused any examination or treatment. When referred, he was in a poor nutritional state and hardly ate or drank anything, so we had him admitted to our hospital and started to feed him via a nasogastric tube.

He had no noteworthy medical history. Computed tomography (CT) showed that the huge mass lesion had a size of 22 cm and destroyed the mandibular bone irregularly. The lesion included solid areas as well as multilocular cyst-like structures with scattered calcifications (Fig. 2). Furthermore, there were some nodules in both upper lung fields (Fig. 3). On the basis of these clinical and imaging findings, the lesion was suspected to be an ameloblastoma and/or a malignant tumor arising from the right side of the mandibular bone. The lung nodules were suspected multiple metastases from the mandibular tumor.

An incisional biopsy revealed a high-grade spindle cell sarcoma with massive necrosis, but the diagnosis was not established.

We planned to perform a radical surgery after improving his general condition. Though we provided nutrition via a feeding tube, his protein-energy malnutrition did not improve due to repeated hemorrhages from

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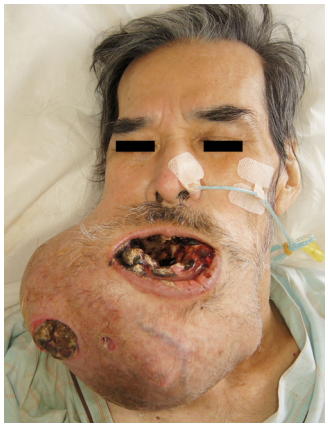

Fig. 1. Patient presenting with a remarkable swelling of the right mandible.

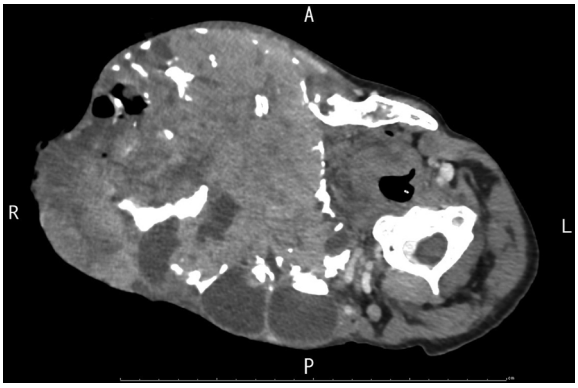

Fig. 2. CT with enhancement (axial plane) demonstrating the huge mass in the right side of the mandible.

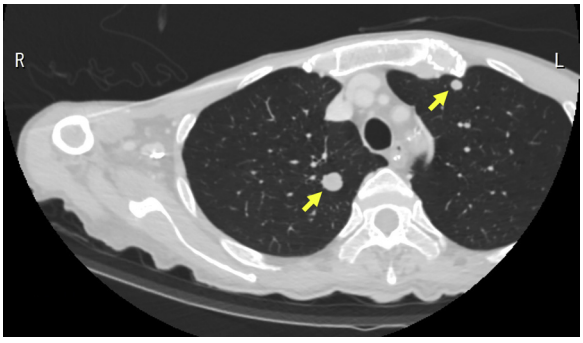

Fig. 3. Chest CT showing some nodules, which indicates suspected metastases of the mandibular tumor (arrow).

the tumor. His general condition deteriorated, and he died.

In the autopsy, the tumor measured 22 cm × 20 cm × 18 cm and weighted 2550 g. It was fragile, and the cut surface showed a solid and cystic appearance with massive necrosis (Fig. 4). The histological examination of the tumor revealed that the tumor was predominantly composed of high-grade spindle cell sarcoma and contained a small amount of epithelial component. The latter was exclusively located in the pericystic areas. The epithelial cells formed follicles supported by fibrous connective tissue stroma. These follicles were lined by tall columnar ameloblast-like cells with reverse nuclear polarity and central stellate reticulum-like cells, both of which had abundant eosinophilic cytoplasmic granules and bland nuclei. These histologic findings indicated that the epithelial component was a granular cell variant of ameloblastoma (Fig. 5). The sarcomatous component, which lacked a specific line of differentiation, showed hypercellularity, marked nuclear

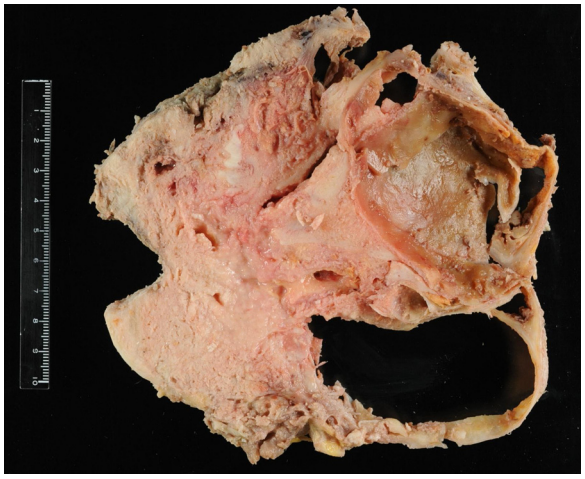

Fig. 4. The cut surface of the specimen. It shows solid and cystic appearance with massive necrosis.

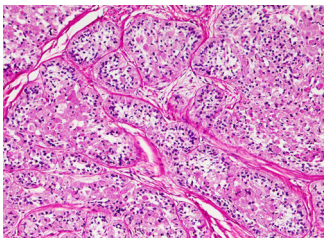

Fig. 5. Large or small epithelial islands made up predominantly of granular cells are rimmed by tall columnar cells, consistent with granular cell variant of ameloblastoma (hematoxylin-eosin, x200).

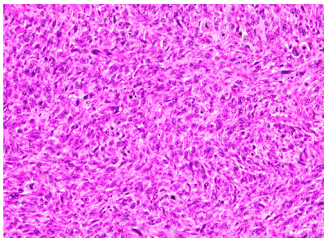

Fig. 6. Most of the tumor is composed of high-grade spindle cells. They lack a specific line of differentiation (hematoxylin-eosin, x200).

pleomorphism, and numerous mitotic figures (Fig. 6). There were only a few foci where the epithelial tumor component was intimately intermingled with the sarcomatous component (Fig. 7A). Immunohistochemically, the epithelial component was positive for Cytokeratin AE1/AE3 and calretinin but negative for vimentin while the cells in the sarcomatous lesion were strongly reactive for vimentin but nonreactive for Cytokeratin AE1/AE3 (Fig. 7B,C). The sarcomatous cells showed a high Ki-67 labeling index (39.3%) (Fig. 7D) and overexpression of p53 antigen (Fig. 7E). There were no reactive cells for desmin, CD31, or S-100 protein in the sarcomatous lesion (Fig.8A–C). Metastatic tumors were seen in the bilateral lungs and the liver. They contained only the sarcomatous element, and both showed overexpression of p53 antigen, too (Fig. 9A,B).

3. Discussion

Malignant odontogenic tumors showing ameloblastic differentiation are classified into ameloblastic carcinoma, and odontogenic sarcoma, also termed ameloblastic fibrosarcoma. Metastasizing ameloblastoma is defined by the World Health Organization classification of 2017 as a

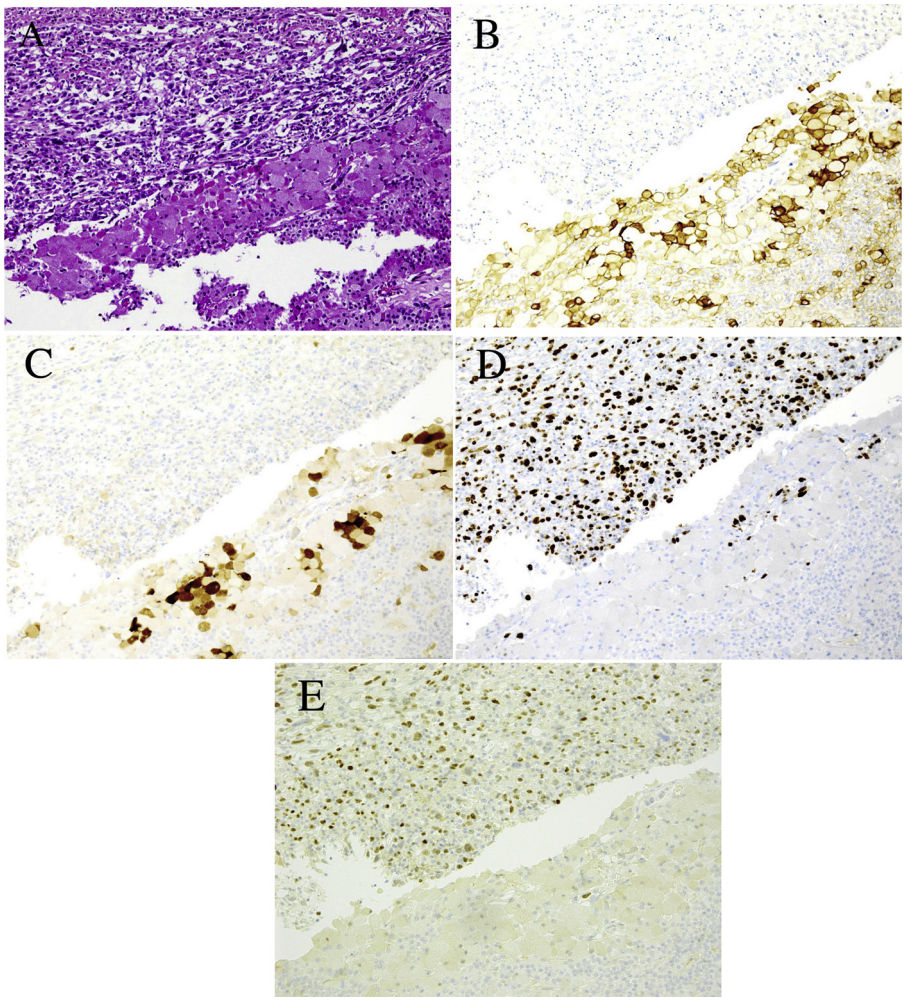

Fig. 7. (A) Only a few epithelial cells are intimately intermingled with the sarcomatous component (hematoxylin-eosin, x200). (B) Epithelial cells express Cytokeratin AE1/AE3 (x200). (C) They are also immunoreactive for Calretinin (x200). (D) The sarcomatous cells show a high Ki-67 labeling index (x200). (E) Overexpression of p53 is diffusely observed in the sarcomatous cells (x200).

metastasizing neoplasm with benign microscopic features of ameloblastoma. The granular cell ameloblastoma is reported to be a locally aggressive tumor with repeated recurrences and the potential to metastasize when compared with other variants [1–4]. Although the epithelial component of the present case was also a granular cell ameloblastoma, the metastatic lesions were composed only of high-grade sarcoma. Therefore, the present tumor is not a metastasizing ameloblastoma. Ameloblastic carcinoma is defined as a tumor showing histologically malignant appearances no matter whether it recurs or metastasizes. They show an increased nuclear to cytoplasmic ratio, nuclear pleomorphism, vascular invasion, and focal tissue necrosis. In contrast to ameloblastic carcinoma, the epithelial component in the present case did not show malignant histological appearances. Massive necrosis was only seen in the sarcoma component. This indicates that the present tumor was not an ameloblastic carcinoma either. Ameloblastic fibrosarcoma is regarded as the malignant counterpart of the benign ameloblastic fibroma [5,6]. The mesenchymal tissue surrounding the epithelial component shows histological evidence of malignancy while the epithelial component appears bland. The present tumor also consisted of unequivocally benign ameloblastoma and high-grade sarcoma. Most of the ameloblastic fibrosarcoma occur through malignant transformation of the mesenchymal component of a pre-existing ameloblastic fibroma, which is a mixed epithelial and mesenchymal neoplasm. However, the present case did not contain any foci with biphasic appearance suggesting an ameloblastic fibroma. Above all, in the present case, most of the epithelial component distributed without a contact with the sarcomatous component although a topographically intimate relationship between the epithelial component and the mesenchymal component should be expected in the case of ameloblastic fibrosarcoma. Furthermore, the sarcomatous component in the present case showed a much more anaplastic and pleomorphic appearance than those described in the textbooks [7] or case reports [5,6,8–10]. Although there are a few reports of anaplastic ameloblastic fibrosarcoma [5], all the cases have ameloblastic fibroma-like lesions. Therefore, we cannot categorize the present tumor in the conventional classification system. Here, we considered the possibility of a collision tumor, which was a cooccurrence of ameloblastoma and high-grade sarcoma. From the frequency of occurrence, ameloblastoma is a relatively rare tumor, and high-grade sarcomas are also very rare tumors. In addition, the sarcomatous component in the present case was undifferentiated of unknown origin. So, it was extremely rare that both rare tumors occurred at the same site simultaneously and came into collision incidentally. Therefore, we postulate that it might have arisen through an accidental malignant transformation of the stromal fibrous connective tissue which supports the follicles of the granular cell ameloblastoma.

In summary, we described an extremely unusual case of a patient with an ameloblastoma and a high-grade sarcoma. Although the history of these tumors cannot be clearly determined, it is important to recognize that such an unusual situation can occur.

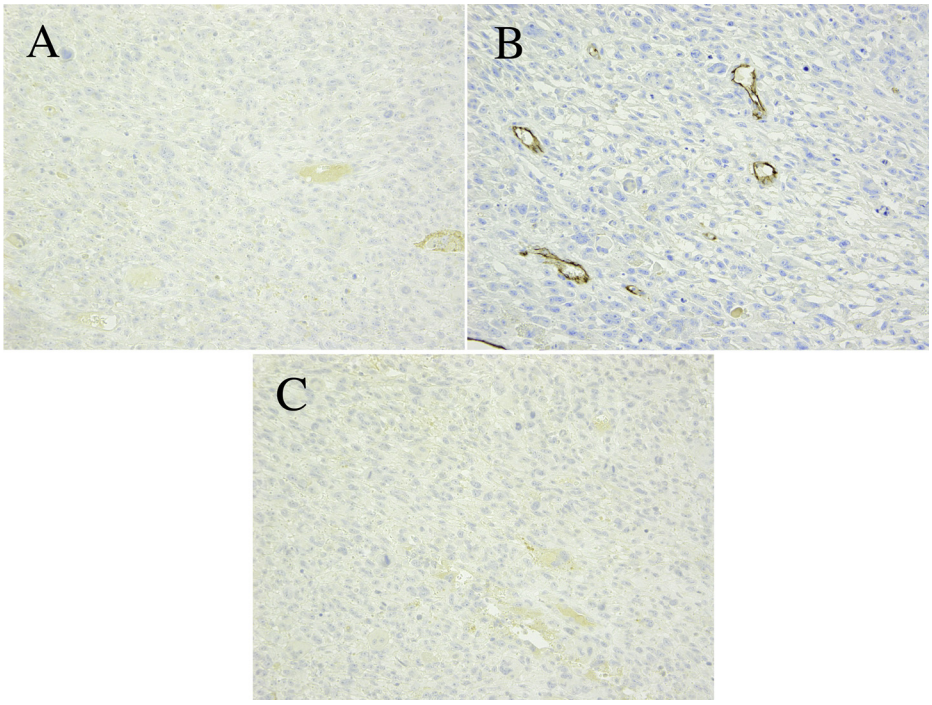

Fig. 8. There are no reactive cells for desmin, CD31, or S-100 protein in the sarcomatous component of the mandibular tumor (x200). (A) Desmin. (B) CD31. (C) S-100 protein.

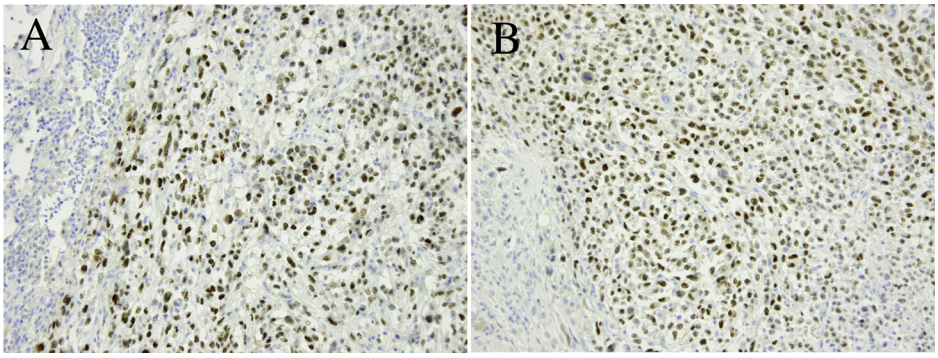

Fig. 9. Overexpression of p53 is seen in the sarcomatous cells of the metastatic tumor in both of the lung and liver (x200). (A) the lung. (B) the liver.

Conflicts of interest

The authors declare no conflicts of interest associated with this manuscript.

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