

# Case of Desmoplastic Ameloblastoma Arising in the Anterior Maxilla

Minako Aiki<sup>1</sup>, Seigo Ohba<sup>1,2</sup>, Kyoko Ishimaru<sup>1</sup>, Shinpei Matsuda<sup>1</sup>, Hitoshi Yoshimura<sup>1</sup>, Shuichi Fujita<sup>3</sup>, Yoshiaki Imamura<sup>4</sup>, Kazuo Sano<sup>1</sup>

<sup>1</sup>Division of Dentistry and Oral Surgery, Department of Sensory and Locomotor Medicine, Faculty of Medical Sciences, University of Fukui, Japan.<sup>2</sup>Department of Regenerative Oral Surgery, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan.<sup>3</sup>Department of Oral Pathology and Bone Metabolism, Unit of Basic Medical Sciences, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan.<sup>4</sup>Division of Surgical Pathology, University of Fukui Hospital, Japan.

## Abstract

Desmoplastic ameloblastoma is a relatively rare subtype of ameloblastoma. Tumor resection with curettage is considered the most reliable treatment for desmoplastic ameloblastoma because of its aggressive behavior with bone invasion. A 43-year-old Japanese man presented with a swelling on the left side of his anterior maxilla. A painless, bone-like hard swelling was observed in the anterior maxillary region. An orthopantomograph revealed a diffuse radiolucent area containing the root apices of the left maxillary lateral incisor and canine tooth. Computed tomography showed a well-margined radiolucent area. Magnetic resonance imaging showed a well-circumscribed lesion with a signal intensity close to that of muscle tissue in the T1-weighted image and slightly lower than that of adipose tissue in the T2-weighted image. Tumor resection with curettage was performed under general anesthesia. Histology of biopsy and resected samples showed scattered epithelial follicles resembling enamel organ and strands of epithelial cells in a coarse, collagen-rich fibrous stroma. Acanthomatous or microcystic changes were also observed in the parenchyma. These findings were consistent with those of desmoplastic ameloblastoma. No recurrence has been observed in the 3-year follow-up period. Curettage of the surrounding bone after tumor resection with sufficient margins prevents recurrence of desmoplastic ameloblastoma. DA is a subtype of ameloblastoma and grows with bone invasion. The risk of recurrence is high. However, early diagnosis and treatment of DA is possible because its preferred site is the anterior jaw.

*Key words: Desmoplastic ameloblastoma, Bone invasion, Surgical treatment, Recurrence*

## Introduction

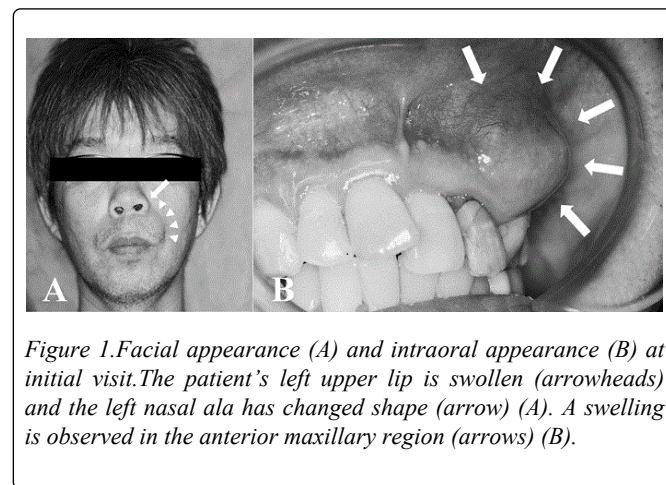
Ameloblastomas are well-known odontogenic tumors that may be classified into several subtypes, including solid/multicystic, extraosseous/peripheral, desmoplastic, and unicystic types [1]. Desmoplastic ameloblastoma (DA) was first described by Eversole et al. [2] and is characterized by the presence of scattered small islands and strands of epithelial cells, and by cystic structures in a coarse, collagen-rich fibrous stroma [3]. DA is extremely uncommon, accounting for only 4%–13% of all ameloblastomas [4]. DA usually creates a unicystic lesion. While other types of ameloblastoma occur predominantly in the posterior mandible, the site of predilection for DA is the anterior maxilla and mandible, especially the alveolar bone or dentulous areas [5].

Tumor resection with curettage is considered the most reliable treatment for DA, because the tumor grows with bone invasion [6]. Marginal resection is performed when lesions are small and located in the alveolar bone [5,7]. The postoperative prognosis usually good; however, the recurrence rate after DA resection is reportedly the same or higher than other types of ameloblastoma, because of bone invasion [8]. We herein report a case of DA arising at the anterior maxilla.

## Case Report

This study received ethical approval from the institution. A 43-year-old Japanese man was referred to the Division of Dentistry and Oral Surgery at the University of Fukui Hospital for swelling on the left side of the anterior maxilla. His medical and family histories were unremarkable. He had first noticed a painless swelling on the left side of the anterior

maxilla 6 months previously. The left side of his upper lip was swollen, changing the shape of the left nasal ala and resulting in facial asymmetry (Figure 1A). A bone-like hard swelling was observed at the anterior maxilla without spontaneous pain or tenderness. The swelling was observed only on the labial side and not on the palatal side of the maxilla (Figure 1B). The left maxillary lateral incisor with root canal filling moved pathologically on palpation. The adjacent teeth were vital and showed physiological movement.



*Figure 1. Facial appearance (A) and intraoral appearance (B) at initial visit. The patient's left upper lip is swollen (arrowheads) and the left nasal ala has changed shape (arrow) (A). A swelling is observed in the anterior maxillary region (arrows) (B).*

## Imaging Findings

An orthopantomograph revealed a diffuse radiolucent area (25 × 24 mm) containing the root apices of the left maxillary lateral incisor and canine tooth, with root divergence between these teeth (Figure 2).

Corresponding author: Seigo Ohba, DDS, PhD, Department of Regenerative Oral Surgery, Nagasaki University Graduate School of Biomedical Sciences, 1-7-1 Sakamoto, Nagasaki 852-8588, Japan, Tel: +81 95 819 7704; Fax: +81 95 819 7705; e-mail: seigoohba@gmail.com /sohba@nagasaki-u.ac.jp

## Progress

Based on clinical and imaging findings, the lesion was diagnosed as a benign tumor of the left anterior maxilla. To make a definitive diagnosis, an incisional biopsy was performed.

Histological examination of the biopsy specimen revealed scattered small islands and strands of epithelial cells, with cystic structures in a coarse, collagen-rich fibrous stroma (Figure 5A). The epithelial islands showed peripheral arrangement of cells, hyperchromatic nuclei, and inner polyhedral cells with abundant cytoplasm. This histological architecture resembled the enamel organ of the dental germ (Figure 5B). Some epithelial strands were arranged in double-stranded trabeculae, and these constituted the lining cells of the cystic structure and the peripheral cells of the epithelial islands. On the basis of these findings, a histopathological diagnosis of ameloblastoma was made.

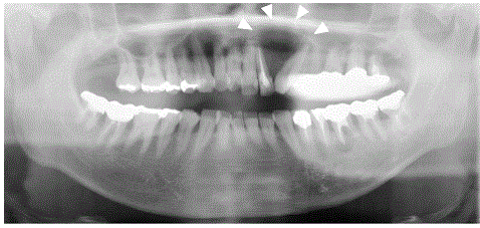


Figure 2. Panoramic radiograph reveals a diffuse radiolucent area (arrowheads).

Computed tomography (CT) showed a well-margined radiolucent area ( $22 \times 21$  mm) in the left anterior maxilla. The alveolar bone in the region had expanded on the labial side but not on the palatal side. Bone resorption was observed, with border irregularity around the left maxillary lateral incisor and canine tooth; root divergence between these teeth was also found (Figure 3).

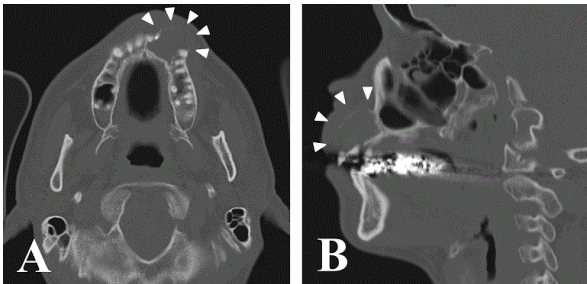


Figure 3. Axial (A) and sagittal (B) sections of CT images. Computed tomography (CT) shows a well-margined radiolucent area ( $22 \times 21$  mm) in the left maxillary anterior region (arrowheads).

Magnetic resonance image (MRI) showed a  $17 \times 8$ -mm well-circumscribed lesion on the left side of the anterior maxilla, with a signal intensity close to that of muscle tissue in the T1-weighted image (Figure 4A) and slightly lower than that of adipose tissue in the T2-weighted image (Figure 4B). The lesion is enhanced slowly by contrast medium. Rim enhancement was observed around the lesion and the interior of the lesion was homogeneous (Figure 4C).

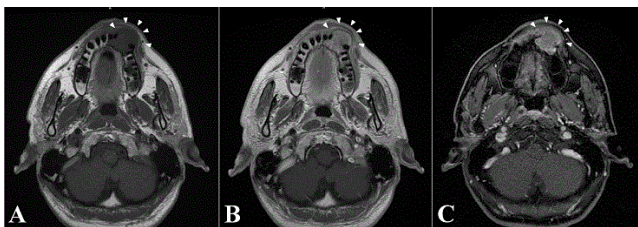


Figure 4. MRI findings. Magnetic resonance image (MRI) shows a lesion on the left side of the anterior maxillary region (arrowheads) in the T1-weighted image (A) T2-weighted image (B)

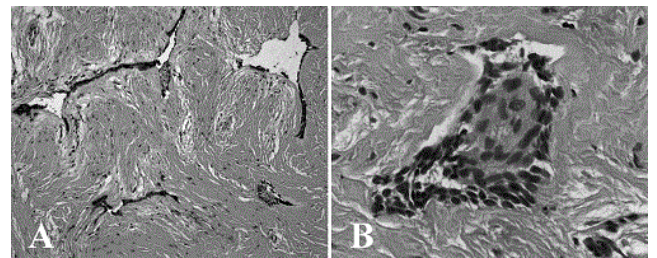


Figure 5. Biopsy findings. Scattered small islands and strands of epithelial cells, and small cysts in the abundant fibrous stroma are seen. The stroma consists of dense collagen bundles including spindle cells (A; H&E,  $\times 10$ ). The small epithelial islands have a peripheral arrangement of cells with hyperchromatic nuclei and inner polyhedral cells with abundant cytoplasm (B; H&E,  $\times 40$ ).

After histopathological diagnosis, tumor resection with extraction of the left maxillary lateral incisor and canine tooth was performed under general anesthesia. A Wassmund incision was made between the right maxillary central incisor and the left maxillary first premolar, with additional longitudinal incisions at these teeth. After extraction of the left maxillary central incisor and first premolar, the tumor was resected en bloc with associated teeth (Figure 6). The exposed bone was curettaged after tumor resection. Because the tumor reached the anterior wall of the left maxillary sinus, this region was also removed without damage to the mucosa of the maxillary sinus. The resection site was covered with collagen membrane (Terudermis, Terumo, Tokyo, Japan) with tie-over.

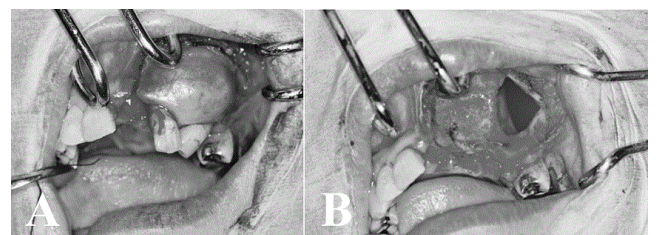


Figure 6. Intraoperative findings. anterior wall of the maxillary sinus was also resected without sinus (B).



The resected specimen was approximately 30 mm in diameter. Histologically, the tumor included irregularly shaped epithelial islands scattered throughout abundant dense fibrous connective tissue (Figure 7A). The tumor had caused resorption of the maxillary bone and extended into the oral submucosal layer. Although there was no obvious fibrous capsule, the tumor was demarcated. No continuity between the ameloblastoma and the mucosal epithelium was observed. Some epithelial islands had a follicular pattern, while most nests had an acanthomatous pattern with keratinization of the inner cells. Narrow zones of myxoid loose connective tissue were seen around some of these follicular and acanthomatous nests. In addition, central cystic degeneration was noted (Figure 7B). The final histopathological diagnosis of the lesion was DA. There has been no recurrence during the 3-year follow-up period (Figures 8 and 9).

## Discussion

Histopathological features of DA include tumor stroma with abundant connective tissue and irregularly shaped islands and cords of odontogenic epithelium of varying sizes embedded in a desmoplastic connective tissue stroma [1]. In this case, scattered small islands, strands of epithelial cells, and microcystic structures were observed in the coarse, collagen-rich fibrous stroma. These histopathological findings satisfied the criteria of DA.

DA occurs predominantly in the anterior maxilla and mandible, whereas other types of ameloblastoma typically occur in the posterior mandible [6]. The location of the tumor in the present case aided the diagnosis. According to previous reports, DA lesions tend to be relatively small (2 cm or less) [8], with the exception of one unusual case that expanded into the maxillary sinus [6]. This tendency makes DA different from conventional ameloblastoma [8]. In this case, the tumor appeared to originate around the left maxillary lateral incisor and canine tooth, because the roots of these teeth diverged, as has been described in a previous report.<sup>5</sup> Growth of the DA resulted in a change in facial appearance, and thus the patient noticed the lesion early in its development.

Histopathologically, ameloblastoma lesions arising in the alveolar bone often contain abundant stroma, resulting in the formation of copious collagen tissue. This may be one reason that DA occurs in the alveolar bone [9].

While root divergence between the left maxillary lateral incisor and canine tooth was observed, there was no periapical resorption of these teeth in the present case. Based on these findings, a possible diagnosis of keratocystic odontogenic tumor was also considered. Root resorption of adjacent teeth is commonly observed in conventional ameloblastoma [5], while it is seldom observed in DA [9]. The reason that DA does not induce root resorption but does induce tooth movement may be its indolent growth pattern. The left maxillary anterior alveolar bone was swollen on the labial side in the present case. This suggests that the tumor grew gradually.

The appearance of DA on X-rays is variable, including radiolucent cystic lesions and lesions with a combination of radiolucent and radiopaque areas similar to fibro-osseous lesions [6]. The combination of radiolucent and radiopaque areas suggests the invasive growth of DA into bone, with bone trabeculae preserved within the lesion. Thus, the typical X-ray finding for DA is a poorly-demarcated lesion with a honeycomb appearance. There have been no reports of DA combined with impacted teeth, and DA usually occurs after the second decade of life. This timing may be relevant, because tumors commonly occur after eruption of the anterior teeth and premolars [4].

DA grows indolently with bone invasion. Therefore, it is necessary to resect the tumor with sufficient scraping of the surrounding bone [5,8,10]. Tumors resected without scraping of the surrounding bone and tumors resected by curettage alone have high recurrence rates [5,8,10]. Total maxillectomy or partial maxillectomy is recommended for good outcomes when DA occurs in the maxilla [6]. The dredging method is often used to treat DA, depending on patient age and

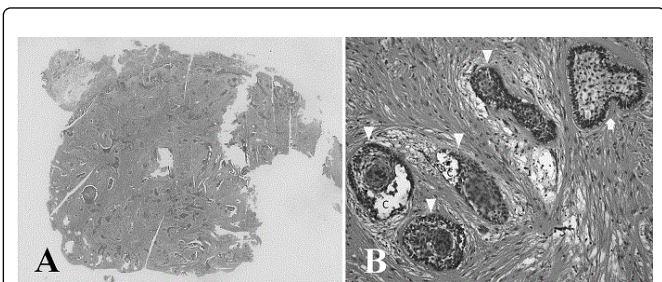


Figure 7. Histopathological findings of the resected tumor. Scattered epithelial islands throughout copious dense fibrous connective tissue was observed. (A; H&E, loupe image). Some epithelial islands had an enamel organ-like follicular pattern composed of a peripheral palisading arrangement of columnar cells and inner satellite cells (arrow); the other acanthomatous pattern is the presence of keratinization of the inner cells (arrowheads). Narrow zones of myxoid loose connective tissue were seen around some nests. (B). (B; H&E,  $\times 10$ ).

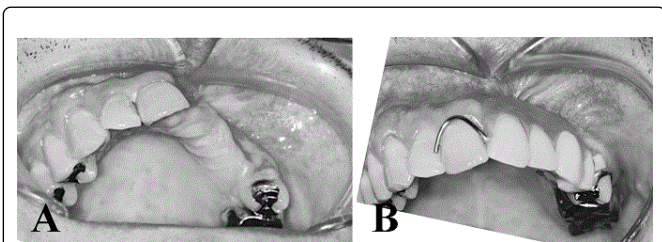


Figure 8. Intraoral findings 3 years after surgery. No mobility or inclination of teeth adjacent to the tumor site are observed. No recurrence was found.

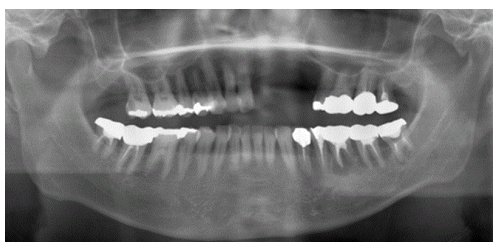


Figure 9. Orthopantomography after surgery. No radiolucent area, which would indicate tumor recurrence, is observed in the region.

characteristics [4]. When DA occurs in the mandible, tumor resection with sufficient margins and reconstruction are considered the best treatment strategy to reduce recurrence [11]. In the present case, orthopantomographic images showed a poorly-marginated lesion, and the lesion showed aggressive invasion into the bone. Thus, the tumor was resected en bloc with the surrounding tissue, including the left maxillary lateral incisor and canine tooth. Curettage of the exposed bone was performed after resection. No recurrence has been found in 3-year follow-up.

### Conclusion

DA is a subtype of ameloblastoma and grows with bone invasion. The risk of recurrence is high. However, early diagnosis and treatment of DA is possible because its preferred site is the anterior jaw. Ongoing follow-up and consideration of facial appearance, occlusion, and recurrence are necessary.

### Conflict of Interest and Source of funding

The authors have no conflicts of interest or sources of funding to declare.

### References

1. Gardner DG, Heikinheimo K, Shear M, et al. Ameloblastomas. In: Barnes L, Eveson JW, Reichart P, et al. (Editors). World Health Organization Classification of Tumors. Pathology and Genetics. *Head and Neck Tumours*, Lyon, France: IARC Press, 2005: 296-300.
2. Eversole LR, Leider AS, Hansen LS. Ameloblastomas with pronounced desmoplasia. *J Oral Maxillofac Surg* 1984; **42**: 735-740.
3. Iida S, Kogo M, Kishino M, et al. Desmoplastic ameloblastoma with large cystic change in the maxillary sinus: report of a case. *J Oral Maxillofac Surg* 2002; **60**: 1995-98.
4. Waldron CA, El-Mofty SK. A histopathologic study of 116 ameloblastomas with special reference to the desmoplastic variant. *Oral Surg* 1987; **63**: 441-451.
5. Durmus E, Kalayci A, Ozturk A, et al. Desmoplastic ameloblastoma in the mandible. *J Craniofac Surg* 2003; **14**: 873-875.
6. Kishino M, Murakami S, Fukada Y, et al. Pathology of the desmoplastic ameloblastoma. *J Oral Pathol Med* 2001; **30**: 35-40.
7. Hatada K, Noma H, Katakura A, et al. Clinicostatistical study of ameloblastoma treatment. *Bull Tokyo Dent Coll* 2001; **42**: 87-95.
8. Higuchi Y, Nakamura N, Ohishi M, et al. Unusual ameloblastoma with extensive stromal desmoplasia. *J Craniomaxillofac Surg* 1991; **19**: 296-300.
9. Kato H, Nomura J, Matsumura Y, et al. A case of desmoplastic ameloblastoma occupying maxillary sinus. *Contemp Clin Dent* 2011; **2**: 234-236.
10. Kaffe I, Buchner A, Taicher S. Radiologic features of desmoplastic variant of ameloblastoma. *Oral Surg Oral Med Oral Pathol* 1993; **76**: 525-529.
11. Reichart PA, Philipsen HP, Sonner S. Ameloblastoma biological profile of 3677 cases. *Eur J Cancer B Oral Oncol* 1995; **31**: 86-99.