Desmoplastic Fibroma Primarily Originating in the Maxillary Alveolus: A Case Report

Hiroshi Takeuchi¹, Motohiko Nagayama¹, Masaru Sato¹, Kuniteru Nagahara², Yasunori Muramatsu³ and Katsuyuki Ohtomo³

¹Department of Oral Pathology, Asahi University School of Dentistry, ²Department of Oral Surgery, Asahi University School of Dentistry, ³Department of Surgery, Murakami Memorial Hospital, Asahi University School of Dentistry, Mizuho-city, Gifu, Japan.

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We report a rare case of desmoplastic fibroma that originated in the maxillary alveolus of an 84-year-old woman. CT demonstrated a large osteolytic intraosseous mass extending to the maxilla and nasal cavity. The curettaged mass, which was $3 \times 3 \times 5$ cm in size and had an oval shape, consisted of spindle cells separated by abundant collagen fibers. Immunohistochemical analysis revealed that the spindle cells had a fibroblastic origin. These features may be confused with well-differentiated fibrosarcoma, another type of intraosseous fibrous lesion. However, the cells were not atypic and were negative for Ki-67. Thus, the present case was diagnosed as a desmoplastic fibroma. Desmoplastic fibroma originating in the maxillary alveolus is extremely rare (3 reported cases, including the present case). The present case is characterized by the age of the patient, 84 years, which is higher than in previous reports.

Key words: desmoplastic fibroma, maxillary alveolus, spindle-shaped cells

Correspondence: Hiroshi Takeuchi, Department of Oral Pathology, Asahi University School of Dentistry, 1851 Hozumi, Mizuho-city, Gifu, Japan
Phone: +81-58-329-1427, Fax: +81-58-329-1427, E-mail: take@dent.asahi-u.ac.jp

Introduction

Desmoplastic fibroma is a rare, locally-aggressive benign lesion or border-line tumor. The mandible and pelvis are the most common sites of the disease, followed by the metaphysis of the femur, tibia and humerus (1). The age of patients has ranged from 15 months to 75 years, with a reported mean age of 23 years (2-4). Desmoplastic fibroma in the maxilla is very rare, and it is stated that almost all cases of maxillary desmoplastic fibroma originated in the orbital floor and sinus (5, 6).

We report an extremely rare case of desmoplastic fibroma that primarily originated in the maxillary alveolus of a woman (84-year-old), who was older than the patients in previous reports, in which the highest patient age was 75 years (2-4).

Case report

An 84-year-old Japanese woman consulted Murakami Memorial Hospital due to a swelling involving the maxilla that interfered with the insertion of her denture. On examination a prominent expansion of the

maxilla was noted, but the mucosa covering the swelling was normal (Fig. 1). There were no symptoms of pain, tenderness, or numbness. The past history of the lesion was uncertain due to the patient's high age.

Radiographic examination, except computed tomography (CT), could not be performed, because the patient

Fig. 1: Macro appearance of swelling of the maxillary alveolus.

Fig. 2: Computed tomography image of lesion in the maxilla.

was unable to assume the postures for other types of radiographic examinations. The CT revealed a well-defined radiolucent unilocular lesion, extending from all sides of the maxillary alveolus. The rear portion of the lesion had resorbed the maxillary bone over the incisive canal, and the upper portion of the lesion perforated to the nasal cavity. The palatine bone and sinus were not affected (Fig. 2).

An initial incisional biopsy of the intrabony lesion was performed, and it was diagnosed as a fibroma of the soft palate at the Murakami Memorial Hospital. The lesion was curettaged easily from the bone with no apparent involvement of either periosteum or floor of the maxillary sinus. The curettaged mass was $3 \times 3 \times 5$ cm in size, with an irregular globular or disc shape (Fig. 3A). The consistency of the mass was elastic firm and/or rubbery. The cut surface was pearly gray in color and did not show other changes such as hemorrhage or cyst formation (Fig. 3B).

The information obtained from the clinical examinations and surgical treatment suggested that the mass

did not extend or infiltrate from other areas to the maxillary alveolus but originated primarily in the maxillary alveolus.

Histopathological findings

The mass was poorly encapsulated by fibrous tissue, and consisted of fibroma-like components, which were dominated by spindle cells. These proliferating cells were separated by abundant collagen fibers forming delicate strands or broad fibrous bands (Fig. 4A). The nuclei of the proliferating cells varied from plump and ovoid to elongated shape. Mitosis was absent in the lesion (Fig. 4B). Myxoid tissue was present in a part of the lesion. Scattered infiltration of lymphocytes and mast cells was also present.

Immunohistochemical findings

For immunohistochemical analysis, the specimens were prepared from the curettaged sample which was fixed in 10% buffered formalin for 48h and then embedded in paraffin. Immunostaining for vimentin, NSE,

Fig. 3: Surgical specimen of tumor showing disc shape (A), and cut surface of tumor showing pearly gray color (B).

Fig. 4: Histological finding of lesion consisting of a large amount of spindle cells (Orig. mag. $\times 100$)(A) and spindle cells without atypia or mitosis (Orig. mag. $\times 200$)(B).

Fig. 5: Immunohistochemical detection of spindle cells positive for vimentin (Orig. mag. $\times 200$).

Fig. 6: Ultrastructural image of spindle cells characterized by very few mitochondria in the cytoplasm (Bar = $5\mu\text{m}$) (A), and spindle cells with formation of abundant collagenous fibers (Bar = 500nm) (B).

desmin, S-100 protein, smooth muscle actin, and Ki-67 (MIB1) was performed by the streptavidin biotinylated immunoperoxidase method using antibodies. We also attempted to detect basic fibroblast growth factor (FGF) and its receptor (FGF-R) immunohistochemically in order to identify the spindle cells (7). Prior to the immunostaining, some antibodies were pre-treated, as shown in Table 1.

Vimentin was positive for most of tumor cells (Fig. 5). NSE, using both non-treated and the autoclave treated antibodies, showed negative findings. The reactions for S-100 protein, using both antibodies treated and non-treated with 0.1% trypsin or autoclave smooth muscle actin, FGF and FGF-R, were negative for all cells. Ki-67

was also not detected.

Ultrastructural findings

Ultrastructurally, cells varied in shape from fusiform to stellate and exhibited a discontinuous membrane and rough endoplasmic reticulum. Only a few mitochondria were present in the cells (Fig. 6A). Conversely, collagenous fibers appeared in and outside of the cells (Fig. 6B). However, myofibroblastic cells containing actin fibers were not present in the lesion.

Discussion

From the clinical data, including radiographic examination and appearance at surgery, the present case

Table 1: Immunohistochemistry

Primary antibodies (animal, clone)	Source	Pre-treatment	Findings
anti-vimentin (M, V9)	†	autoclave	+
anti-desmin (M, D33)	†	none	–
anti-S-100 protein (R)	†	trypsin/none	–/–
anti-smooth muscle actin (M, 1A4)	†	autoclave	–
anti-Ki-67 (M, MIB-1)	†	autoclave	–
anti-NSE (M, BBS/NC/VI-H14)	‡	autoclave/none	+/–
anti-basic FGF (R)	¶	none	–
anti-FGF-R, Bek (R)	¶	none	–

†, DAKOCytomation A/S, Glostrup, Denmark

‡, DAKOCytomation Inc., California, US

¶, Santa Cruz Biotechnology, Inc., California, US

M, mouse monoclonal; R, rabbit polyclonal

+, positive finding; –, negative finding

evidently originated primarily in the maxillary alveolus. The lesion was histologically composed of spindle cells separated by abundant collagen fibers and myxoid tissue without appearance of histiocytic cells, mono- or multinucleated giant cells, chondroid, osteoid and mineralized bone which are characteristic of some intraosseous fibrous lesions such as fibrous dysplasia, osteofibrous dysplasia, and non-ossifying fibroma. The histological pattern of the lesion was similar to desmoplastic fibroma, fibromyxoma and well-differentiated fibrosarcoma. However, fibromyxoma was excluded from a differential diagnosis because the myxomatous tissue of the present case was not the main component of the lesion, as seen in fibromyxoma, and was considered to be myxomatous changes of the stromal tissue. While well-differentiated fibrosarcoma with considerable collagen production and minimal nuclear atypia often has enlarged nuclei with irregular chromatin clumping, the nuclei of the neoplastic cells of the present case varied from somewhat plump and ovoid to elongated, thin and hyperchromatic; mitotic figures were usually absent. Such a lack of mitotic figures and nuclear atypia may distinguish desmoplastic fibroma from malignant bone tumors (8). Proliferation indexes from flow cytometric DNA analysis for locally-infiltrating desmoplastic fibromas were, however, reported to be between 21.5 and 24% (4). Very few tumor cells of the cranial desmoplastic fibromas were reported to be positive for MIB-1 staining in high-power fields (9). The present case also exhibited a lack of mitotic figures and nuclear atypia, and was negative with Ki-67 (MIB-1), indicating that the present case was a benign tumor with cortical destruction. Ultrastructural features of desmoplastic fibromas

have characterized the principal tumor cell as a fibroblast (10), myofibroblast (11) or abnormal fibroblast showing myoid differentiation (12). In the present case, the myofibroblast was not found by ultrastructural and immunohistochemical analyses. Furthermore, the tumor cells were positive for anti-desmin, and negative for anti-S-100 protein and anti-NSE antibodies, indicating that the cells originated not in neurogenic cells but in fibroblastic cells. Since the present case primarily arose in the jaw bone, an odontogenic tumor, for example, a variant of odontogenic fibroma, would also have to be considered in a differential diagnosis, as previously proposed by Slootweg & Müller (13), Drebber *et al.* (14) and Gardner (15). The present case did not resemble any of the mesenchymal tumors, such as hyperplastic dental follicles, odontogenic fibroma into simple and WHO types, odontogenic fibroma exhibiting pleomorphic fibroblasts or central odontogenic fibroma, which were proposed by Gardner (15). An immunohistochemical discrimination between the present case and odontogenic tumors was attempted using the odontogenic fibrous cell-specific antisera, such as anti-FGF and anti-FGF-R (7). The analysis also revealed that the present case did not originate in odontogenic mesenchymal cells.

Based on the various examinations mentioned above, the present case was diagnosed as a desmoplastic fibroma, primarily originating in the maxillary alveolus. The definition of desmoplastic fibroma, accepted by WHO, is that of a benign tumor characterized by the formation of abundant collagen fibers by the tumor cells and lacking the cellularity, pleomorphism and mitotic activity that are features of fibrosarcoma (16), which were all in accor-

dance with the present case.

Desmoplastic fibroma is a rare tumor of bone that occurs in one case out of 6,221 bone tumors, according to a statistical analysis by Dahlin (3). It constitutes 0.06% of all bone neoplasms. The lesion is most common in young people, as is evident from epidemiological research, with a mean age of 23 years (2, 4). The preferred sites of the lesion are the metaphyses of the long bones and the mandible (17). Since the initial description of a desmoplastic fibroma by Jaffe (19) in 1958, about 200 cases of desmoplastic fibroma have been reported, but only several cases were reported to arise in the maxilla. In particular, only 2 cases of desmoplastic fibromas that primarily originated in the maxillary alveolus have been reported (20). Therefore, the present case can be considered to be an extremely rare case. Further, the present case is also characterized by the age of patient, 84 years, who had the highest age of any reported desmoplastic fibroma patient.

References

1. Inwards CY, Unni KK and Beabout JW. Desmoplastic fibroma of bone. *Cancer* 1991; **68**: 1978-83.
2. Dahlin DC. Bone tumors. In General aspects and data of 6,221 cases, 3rd edi. Thomas, Springfield, 1978; 3325-6.
3. Goldberg AN, Janecka IP and Sekhar LN. Desmoplastic fibroma of the skull: a case report. *Otolaryngol Head Neck Surg* 1995; **12**: 589-91.
4. Böhm P, Kröber S, Greschiok A, *et al.* Desmoplastic fibroma of the bone: a report of two patients, review of the literature and therapeutic implications. *Cancer* 1996; **78**: 1011-23.
5. Osguthorpe JD, Adkins WH and Rawe SE. Combined extra-cranial-intracranial resection of maxillary desmoid tumor. *Otolaryngol Head Neck Surg* 1981; **89**: 392-7.
6. Bertrand JC, Plautier D, Chanterelle A, *et al.* Fibromes desmoids maxillo-mandibularis. *Rev Stomatol Chir Maxillofac* 1981; **82**: 127-31.
7. So F, Daley TD, Jackson L, *et al.* Immunohistochemical localization of fibroblast growth factors FGF-1 and FGF-2, and receptors FGFR2 and FGFR3 in the epithelium of human odontogenic cysts and tumors. *J Oral Pathol Med* 2001; **30**: 428-33.
8. Dutt SN, Mirza S, Irving RM, *et al.* Desmoplastic fibroma of the temporal bone. *J Laryngol Otol* 2000; **114**: 314-7.
9. Yoon SH, Kim SH, Shin YS, *et al.* Case report: Desmoplastic fibroma of the skull in an infant. *Childs Nerv Syst* 2006; **22**: 176-81.
10. Suguira I. Desmoplastic fibroma: case report and review of the literature. *J Bone Joint Surg Am* 1976; **58**: 126-30.
11. Lagace R, Delage C, Bouchard HL, *et al.* Desmoplastic fibroma of bone. An ultrastructural study. *Am J Surg Pathol* 1979; **3**: 423-30.
12. Chomette G, Tranbaloc P, Auriol M, *et al.* Desmoid fibroma of bone. An histoenzymological and ultrastructural study of the mandible in one case. *Ann Pathol* 1981; **1**: 147-51.
13. Slootweg PJ and Müller H. Differential diagnosis of fibro-osseous jaw lesions. A histological investigation on 30 cases. *J Cranio-Max Fac Surg* 1990; **18**: 210-4.
14. Drebber U, Scheer M, Zöller JE, *et al.* Zentrales odontogenes Fibrom. Ein seltener Tumor. *Pathologe* 2003; 136-40.
15. Gardner DG. Central odontogenic fibroma current concepts. *J Oral Pathol Med* 1996; **25**: 556-1.
16. Schajowicz F, Sissons HA and Sobin LH. The World Health Organization's histologic classification of bone tumors. A commentary on the second edition. *Cancer* 1995; **75**: 1208-14.
17. Destout JM, Kyriakos M and Gilula LA. Fibrous histiocytoma with a review of the literature. *Skeletal Radiol* 1980; **5**: 241-6.
18. George DI, Gould AR, Miller RL, *et al.* Desmoplastic fibroma of the maxilla. *J Oral Maxillofac Surg* 1985; **43**: 718-25.
19. Jaffe HL. Tumors and tumorous conditions of the bones and joints. *Lea & Febiger*, Philadelphia. 1958; 98-303.
20. Sleeman DJ, Paterson A and Eveson JW. Desmoplastic fibroma of the maxillary alveolus. *Oral Oncol Eur J Cancer* 1993; **29B**: 151-2.

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