Central Schwannoma of the mandible: report of a case and review of the literature

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Abstract: We report a rare case of central Schwannoma, a type of nerve sheath tumor, arising in the mandibular bone of a 34-year-old man. A periapical lesion was discovered during routine dental treatment. Radiographically, the lesion was a large round-shaped and well-defined radiolucent lesion with a radiopaque rim in the anterior apical region, which extended from the right canine to the left lateral incisor in the mandible. The surgically removed mass was a relatively well-encapsulated tumor, measuring 1.5 cm in diameter. Histologically, the tumor showed a typical histology of Antoni type A of Schwannoma, focally interspersed with myxoid areas in which tumor cells had nuclear atypia. A review of the literature revealed that there have been only a total of 54 reported cases of central Schwannoma of the jaw bones including the present case, most of which were located in the posterior part of the mandible with a female/male ratio of 1.6 to 1 and a mean age of 33.9 years.

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Introduction

Schwannoma (also called neurilemmoma or neurilemmoma), is a benign nerve sheath soft tissue tumor which occurs more frequently in the head and neck regions than in other parts of the body (1-3). Its origin is thought to be from neural crest-derived Schwann cells of the nerve sheath (2, 4). In the head and neck soft parts, this tumor has been considered to arise along the 5th, 7-9th, or 11-12th cranial nerves (2-3), while its occurrence within bone tissues is extremely rare. In a large scale clinicopathologic study of bone tumors, intraosseous Schwannomas were reported to account for less than 1% of bone tumors of the whole body (5). Schwannomas have been documented to affect such bones as the fibula, tibia, occipital, sternum, vertebrae, and jaw (5-12). Among them, the mandible is more affected than the other bones (5, 12-40, 49, 50, 53-58), while the maxilla is much less frequently involved than the mandible (41-48). Schwannoma in the mandible is usually considered to originate from the 5th cranial nerve, the third branch of

which penetrates the mandibular body (2-3). This particular intraosseous tumor tends to persist for years without the patient's knowledge and is usually discovered incidentally. Such a clinical course seems to reflect the characteristic histology of central Schwannomas with degenerative changes. This report describes a case of intra-osseous Schwannoma arising in the mandible, and it emphasizes its histopathological characteristics and reviews the English literature to characterize clinicopathologic features of intra-osseous Schwannoma.

Case report

A 34-year-old male patient was referred to Niigata University Dental Hospital for treatment of a broken bridge of the anterior part of the maxilla, which was related to the cleft palate. On clinical examination, there was a slight swelling, hard and painless on palpation, in the right incisor apical area of the mandible with slight mobility of the incisor teeth. The covering labial and gingival mucosa were normal

Fig. 1. Radiographic images of central Schwannoma arising in the mandibular bone of a 34-year-old man. (a) Panoramic X-ray; (b) CT scan, axial, bone window. A well-demarcated radiolucent lesion with a smooth radiopaque rim, involving tooth roots of the right canine to the left incisors, was found in the mandibular bone (a). CT scan images showed that the round-shaped osteolytic lesion was continuous with the mandibular canal (b).

in color, and there was no paresthesia in the lower lip. On a panoramic radiograph, there was a well-defined radiolucent lesion with a smooth radiopaque rim, which involved the interior aspect of the mandible extended from the right canine to the left incisor tooth roots down to the lower cortex of the mandibular bone (Fig 1a). Axial CT images in the bone window revealed that both the lingual and buccal cortical plates of the mandible were thinned and partially interrupted. The lesion was also continuous in part with the mandibular canal, which was focally enlarged with a sclerotic rim (Fig 1b). The radiographic findings suggested a benign tumor, not of odontogenic or osteogenic origin. A biopsy was performed from the intraosseous lesion, and a histopathologic diagnosis of intraosseous Schwannoma of the mandible was made. Partial resection of the anterior part of the mandible was done under general anesthesia.

Pathological findings

The surgically removed lesion was ovoid in shape, elastic firm in consistency, and brownish white in color. There was thinning of the lingual plate of bone and perforation of the buccal plate. Its fibrous capsule was attached to the muscle tissue from the buccal surface. A mandibular nerve bundle was found to be attached to a thin fibrous capsule of the tumor.

Histologically, the tumor was well circumscribed with a fibrous capsule which was partially and directly adhered to

the muscular tissue, indicating that the bone cortex was resorbed by the tumor expansion. The tumor was composed of a dense proliferation of spindle-shaped cells with a characteristic nuclear palisading arrangement in the background of thick collagenous bundles, which was a typical feature of the Antoni type A Schwannoma. This tissue pattern of Antoni type A was interspersed with stromal spaces with loose-textured and finely fibrillar appearances, in which tumor cells with irregular-shaped and hyperchromatic nuclei were scattered and hemorrhage with hemosiderosis was associated (Fig 2a). Immunohistochemically, S-100 protein was labeled strongly in most of the nuclei of spindle-shaped cells, especially within the Antoni type A areas (Fig. 2b).

Review of the literature

Our review of the English literature from 1945 to 2009 revealed a total of 54 documented cases with complete clinical data of central Schwannoma of the jaw bone including presented case. Forty-six cases (85.2%) were in the mandible and 8 cases (14.8%) in the maxilla (Table 1). Among the 46 mandibular cases, 24 (52.1%) were located in the molar-premolar region, 9 (20.0%) in the ramus-molar region, and 9 cases (20.0%) in the incisor region. In contrast to the mandibular cases, six of the eight maxillary cases involved the incisor-canine region and only two cases involved the molar region. Regarding sex, there were 21

Fig. 2. Histological findings of central Schwannoma of the mandible. (a) Hematoxylin and eosin (HE) stain; (b) immunoperoxidase stain for S-100 protein, hematoxylin counterstain, $\times 100$. The tumor was mainly composed of a dense proliferation of spindle-shaped cells with a characteristic nuclear palisading arrangement in the background of thick collagenous bundles. However, there were also foci with loose-textured and finely fibrillar stromal space with hemorrhage or hemosiderosis, in which spindle cells with irregular-shaped hyperchromatic nuclei were scattered (a). Tumor cell nuclei in the cellular area were immunopositive for S-100 protein (b).

male and 33 female patients, a ratio of 1:1.6. The mean age of the patients was 33.9 years, with a wide range from 7 to 84 years. Among them, there were 12 teenage patients, constituting 22.2% of the total cases. There was no difference in age distribution between male and female patients as a whole. However, mandibular tumors were more frequently found in older (mean age 38.2 years) females (male:female = 1:1.7), while maxillary ones were in younger (21.4 years) males (male:female = 1:1) (Table 1).

Central Schwannomas present no such apparent clinical signs, other than swelling of jaw bones (58), pain or paresthesia, which are very often noticed in soft tissue Schwannomas (11). Therefore, central Schwannomas tend to be discovered incidentally after long durations. Radiographically, there seem to be no obvious characteristics for central Schwannomas other than lytic defects with sclerotic margins and cortical erosion with neither periosteal new bone formation nor multiloculation (6-10, 12, 13). The presence of central calcification/ossification and root resorption has been documented in a small number of cases (8, 25-27). Thus, benign appearances on radiogram are at least recognizable in central Schwannomas, but it seems to be difficult to differentiate them from other fibro-osseous lesions or odontogenic tumors when central Schwannomas have central spot calcification.

Histologically, the tumor is usually encapsulated, and nerve fibers may be found eccentric to the lesion. It has two characteristic histological patterns: Antoni types A and B, the same as soft tissue Schwannomas (6, 12-13, 49). However, there seem to be some histological differences between intraosseous Schwannomas and their soft tissue counterparts, such as higher cellularities with subtle palisading and poorly-formed Verocay bodies (6) and increased numbers of cells with large, hyperchromatic nuclei in Antoni type B foci (10). There have been two case reports of cellular Schwannoma in the mandible (49-50), which histologically showed nuclear pleomorphism and hyperchromatism of tumor cells with frequent mitotic figures but no Verocay bodies.

Discussion

Although Schwannoma can occur in any part of the body, the head and neck region is the most commonly affected site. Head and neck Schwannomas constitute about 45% of the reported cases from the entire body (1, 3, 28, 54). In the oral cavity, the tongue is most frequently affected by the tumor (1, 3). However, its intraosseous counterparts are extremely rare, representing less than 1% of bone tumors (5-8, 11). Among all intraosseous Schwannomas, those arising in the mandible are more frequent than in any other bones (5, 12, 13, 24-26, 37, 45).

Based on our review of the literature, the clinical characteristics of central Schwannomas of the jaw bones can be summarized as follows: central Schwannomas are more frequent in the mandible, with approximately half of the mandibular cases found in the posterior region, and mandibular tumors are more frequently found in aged

Table 1. Reported cases of Schwannomas of the jaws (1945-2009)

case #	age & sex	location	ref #
1	18 F	maxilla	incisor-canine 41
2	11 F		incisor-canine 42
3	27 M		incisor-canine 43
4	7 M		incisor-canine 44
5	9 M		incisor-canine 47
6	14 F		incisor-canine 48
7	21 M		molar 45
8	64 F		molar 46
9	16 M	mandible	incisor 15
10	28 M		incisor 39
11	60 M		incisor 16
12	22 F		incisor 21
13	11 M		incisor 57
14	64 F		incisor-molar 5
15	12 M		incisor-premolar 26
16	23 F		incisor-canine 35
17	34 M		incisor-canine present case
18	16 F		canine 24
19	19 F		canine-molar 5
20	51 F		canine-molar 27
21	11 M		canine-premolar 38
22	28 M		premolar 13
23	19 F		premolar 23
24	72 F		premolar-molar 33
25	27 F		premolar-molar 19
26	23 F		premolar-molar 53
27	48 F		molar 29
28	43 F		molar 30
29	39 M		molar 31
30	64 F		molar 32
31	20 F		molar 11
32	49 M		molar 36
33	31 F		molar 28
34	30 F		molar 25
35	36 F		molar 25
36	84 F		molar 12
37	50 F		molar 12
38	16 F		molar 40
39	55 F		molar 17
40	40 M		molar 49
41	8 M		molar 20
42	29 F		molar 22
43	34 F		molar 58
44	76 F		molar 56
45	23 F		molar 54
46	25 M		molar-ramus 5
47	65 M		molar-ramus 5
48	44 F		molar-ramus 37
49	72 M		molar-ramus 18
50	50 F		molar-ramus 50
51	12 F		ramus 34
52	20 M		ramus 14
53	66 F		ramus 14
54	60 M		ramus 55

females, while maxillary ones are more frequent in younger males. The patients with jaw bone Schwannoma were younger than those with soft tissue Schwannoma. In terms of sex, Williams *et al.* found an apparent male predilection (83%) in their series of head and neck soft tissue Schwannomas, a tendency opposite to that of the central counterpart (1). Unlike soft tissue counterparts, in which pain is one of the major clinical symptoms, central Schwannomas include only swelling of bones (14).

The radiographic features of central Schwannoma are non-specific, and the lesion can resemble other intrabony lesions such as odontogenic tumors or odontogenic cysts (5, 11-18, 55).

Histopathological features of central Schwannoma, including immunohistochemical findings, seem to be basically the same as those of soft tissue Schwannoma. However, central Schwannoma tends to present the Antoni type B histology because of its longstanding clinical courses (26). De la Monte *et al.* also pointed out this characteristic feature by referring to the subtle palisading tendencies of tumor cells, poorly formed Verocay bodies, and broad areas of dense collagenous stroma in central Schwannomas (6). Increased numbers of cells with large, hyperchromatic nuclei, which were indicative of a degenerative nature, were also reported in tibia cases (10) and cellular variants in the mandible (49-50). All of these characteristics were found in our present case. Thus, the central Schwannoma-specific histology mentioned above should be recognized.

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