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Ancient schwannoma in oral cavity: a report of two cases

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Abstract (J Korean Assoc Oral Maxillofac Surg 2011;37:530-4)

This paper reports two cases of schwannomas arising from the oral cavity. One is an intraoral ancient schwannoma located at the left cheek, which evolved over a period of 13 years. The tumor was a well-demarcated buccal mass, which was located in the left lower first premolar area, with an obliterated the buccal vestibule, leaving the overlying mucosa intact. The second case was a central intraosseous schwannoma located from the left lower 1st molar periapical area to the left 3rd molar periapical area. Pathologically, the first mass was composed of the spindle shaped tumor cells with wavy nuclei beneath the fibroconnective tissue of the gingiva but second case mass was not. Occasional nuclear pleomorphism was observed but mitosis or necrosis was absent. There were Antoni A and B areas along with strong, diffuse staining with the S-100 protein. Ancient schwannomas were diagnosed. Schwannoma is a slow-growing benign tumor, and an ancient schwannoma that shows cellular atypism is a variant of a schwannoma caused by purely degenerative changes. To date, only limited cases of ancient schwannomas in the oral cavity have been reported.

Key words: Neurilemmoma, Mouth, S100 proteins

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I. Introduction

Schwannoma is also named neurilemmoma, neurinoma and Schwann cell tumor, it is a rare benign neural tumor that arises from the neural sheath Schwann cells of the peripheral, cranial or autonomic nerves[1]. Its origin is most commonly associated with a nerve trunk, and most of the time it affects the whole nerve throughout its the course in the peripheral nervous system. The clinical symptoms depend on the nerve of origin[2]. Up to approximately 40% of head and neck tumors are schwannomas, and intraoral schwannomas constitute a mere 1%. The tongue is the commonest site of intraoral schwannomas[3]. The schwannoma occurs centrally within the jaws is rare[4]. Ancient schwannoma is another further rare variant of intraoral schwannomas. To date, only limited cases of ancient schwannomas have been reported in the oral cavity[5-16].

We report here on additional two cases of ancient schwannoma at the buccal mucosa and mandibular body along with discussing its possible pathogenesis.

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II. Cases report

1. Case 1

A 66-year old Korean woman visited to the hospital for evaluation of an asymptomatic protruded intraoral mass that had initially developed 13 years previously. The mass had been slowly growing in size from that time. She had no symptoms or family history of neurofibromatosis 1. No neurological abnormality was noted. Neck computed tomography showed an ill-defined mass like soft tissue density at the left mandible.(Fig. 1) Intraoperatively, a palpable fluctuating 2.0×1.3×1.0 cm-sized firm mass was found in the buccal region of the left lower first premolar area and adjacent to the mental foramen, and the mass had obliterated the mental foramen with leaving the overlying mucosa intact. The protruding mass was firm in consistency, freely mobile and was not tender. It was clearly separated from the adjacent tissue. Intraoperatively, the attachment portion of origin of the nerve could not be visible. There was no carious tooth. Complete excision was done under local anesthesia via an intraoral approach. Grossly, the excised

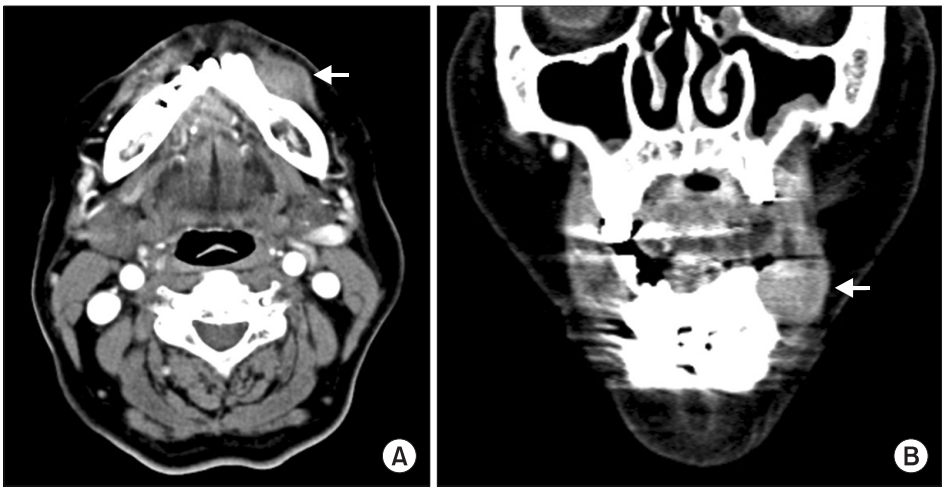

Fig. 1. Axial (A) and coronal (B) computed tomography of Case 1 reveals an ill defined lesion at the left mandibular body (arrows).
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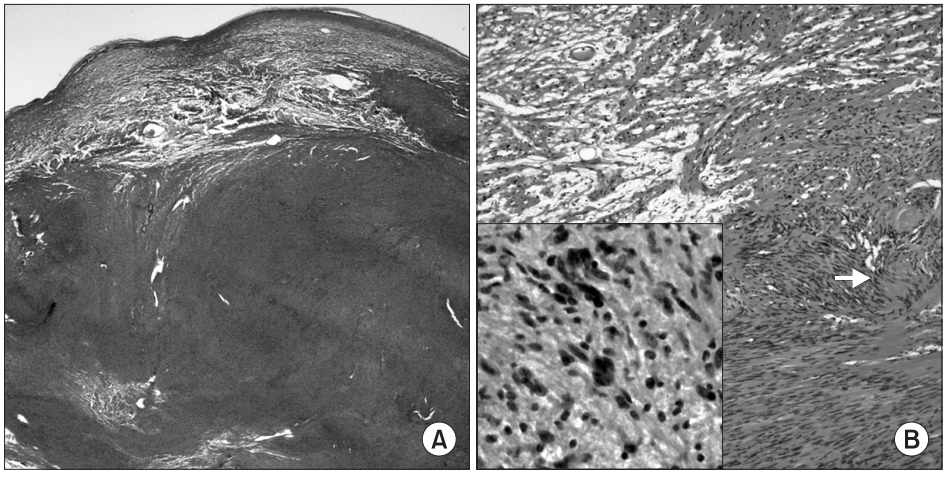

Fig. 3. A. The relatively well marginated mass consists of spindle shaped tumor cells with buckling nuclei (H & E staining, ×40). B. The mass comprises the Antoni A area with the nuclear palisading patterns forming occasional Verocay bodies (arrow). Note the paucicellular myxoid Antoni B area (left upper portion, H & E staining, ×200).
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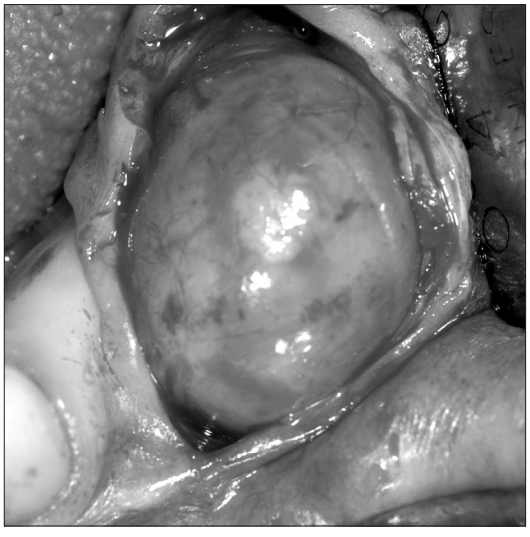

Fig. 2. The well circumscribed protruding mass over the first premolar area near the mental foramen is seen in Case 1.
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mass was pinkish tan colored smooth glistening.(Fig. 2) Pathologically, a well-demarcated, but unencapsulated lesion was found.(Fig. 3. A) The lesion was composed of the spindle cells arranged in fascicles. The tumor cells had a wavy shape, poorly defined cytoplasm and oval nuclei with tapering ends or buckling.(Fig. 3. B) Verocay bodies were seen and thickened hyalinized vessels were also detected. Occasional nuclear pleomorphism was found, but mitosis or necrosis was absent. It was diagnosed as an ancient schwannoma because of lacking necrosis, mitosis, and invasiveness as well as the absence of abrupt transition between the typical schwannoma areas and foci of atypical bizarre cells. The tumor cells were positive for S-100 protein (polyclonal; 1:1,200 dilution, Zymed, San Francisco, CA, USA) and they were negative for pancytokeratin (AE1/AE3, Dako, Glostrup, Denmark, prediluted), desmin (D33; Dako, 1:100 dilution) and smooth muscle actin (1A4; Dako, 1:100 dilution).

Numbness on the left lower lip developed postoperatively.

She has been followed up for 9 months and partial recovery of sensation was achieved. There has been no evidence of recurrence.

2. Case 2

A 35-year old Vietnamese woman was referred by local dental clinic for evaluation radiolucent mass on left mandible body area. She had no symptoms even paraesthesia. Panoramic X-ray showed well-defined homogeneous low density about 3.0×1.5×2.0 cm-sized cystic mass which is including inferior alveolar nerve in left mandible body. Left lower 1st molar distal root and 2nd molar root were resorption state.(Figs. 4, 5) Intraoperatively, bone intrusion did not show and overlying mucosa was intact. Left lower 1st molar was root canal therapy and gold crown state. There was no carious tooth. Lower vestibular incion of left posterior area was performed and incisional biopsy was done after bony grinding of thin buccal wall. Pathologically, the lesion was composed of cellular spindle cells in fascicles.(Fig. 6) The tumor cells showed occasional bizarre shaped enlarged cells with no mitosis or necrosis. It was diagnosed as schwannoma with focal ancient changes. The spindle cells were positive for S-100 protein and they were negative for pancytokeratin, desmin and smooth muscle actin.

Numbness on the left lower lip developed postoperatively. Unfortunately, she was dead by a car accident before complete mass removal surgery.

III. Discussion

Schwannoma is a tumor that arises from Schwann cells, and it occurs throughout the body. The benign schwannoma is a slow-growing encapsulated nodular lesion that is usually solitary. About half of all neurogenic tumors are seen in the head and neck region. Approximately 25% to 45% of all schwannomas are seen in the head and neck region and are

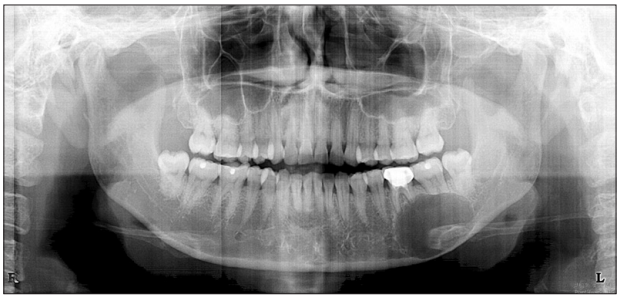

Fig. 4. Panoramic view of Case 2 showed well-defined homogeneous low density about 3.0×1.5×2.0 cm-sized cystic mass which is including inferior alveolar nerve in left mandible body. Left lower 1st molar distal root and 2nd molar root were resorption state.
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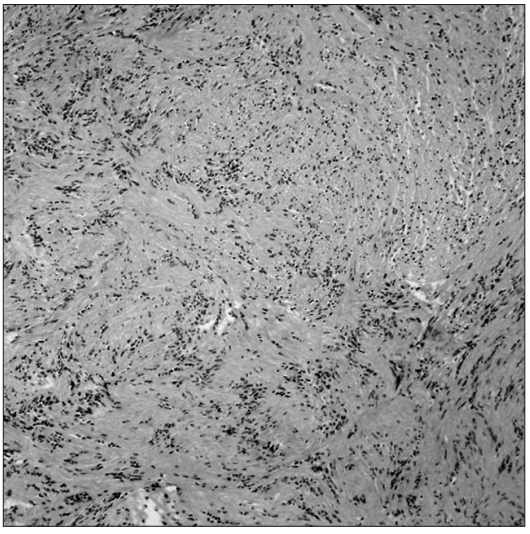

Fig. 6. The biopsied specimen is composed of spindle cells with twisted nuclei (H & E staining, ×100).
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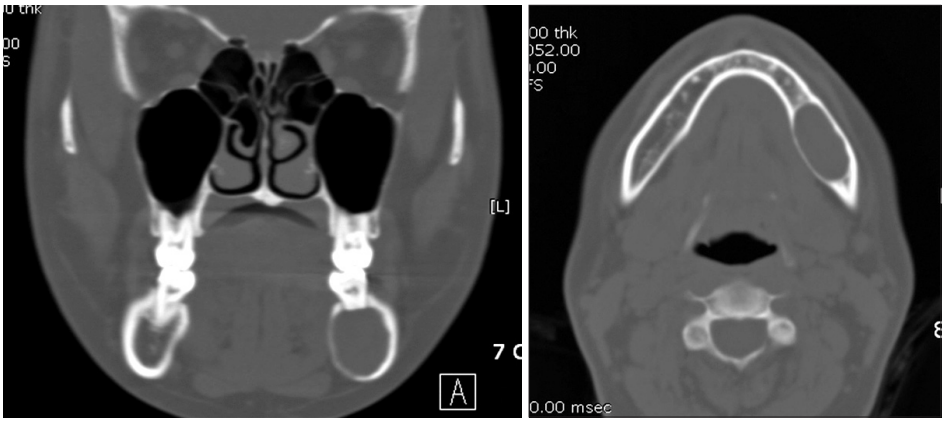

Fig. 5. A well-defined homogenous low density cystic mass in left mandible body. Definite root resorptions of molars were seen in left mandible.
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found rarely in the oral cavity. The most frequently affected site among the head and neck region is the 8th cranial nerve. Other locations include the scalp, face, pharynx, parotid gland, middle ear and external auditory canal. Only 1% of schwannomas demonstrate an intraoral origin; but when they do they occur with greater to lesser frequency in the mobile portion of the tongue, the floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and the mental nerve region[1-3,13,17]. It can develop at any age, but it is more common in the third and fourth decades. Pediatric intraoral schwannoma has been reported[17,18].

Clinically, intraoral schwannomas present with nonspecific symptoms that are indistinguishable from those of other encapsulated lesion that appear as a smooth submucosal swelling such as intraoral mucocele, fibroepithelial polyp, traumatic neuroma, granular cell tumor, solitary neurofibroma, lipoma, fibroma, malignant schwannoma, other intraoral cystic lesions and salivary gland tumors[1,19]. Making the preoperative diagnosis is often difficult, and imaging studies such as computed tomography and magnetic resonance imaging are helpful for the differential diagnosis[18]. Magnetic resonance imaging is superior to other imaging modalities for examining intraoral lesion. A schwannoma is smooth and well-demarcated, and it is isointense compared to muscle on T1-weighted images and it is homogeneously hyperintense on T2-weighted images[20].

Schwannoma is a benign peripheral nerve sheath tumor with the gross presentation with being a gelatinous or cystic long encapsulated or unencapsulated mass with occasional secondary changes such as cystic degeneration, hyalinized vessels and necrosis[21]. The pathologic findings of schwannoma are quite distinct. Microscopically, schwannoma shows a typical biphasic pattern of cellular Antoni A and paucicellular Antoni B areas; the Antoni A areas are composed of spindle cells with twisted, buckled nuclei and occasional intranuclear vacuoles, and the spindle cells are arranged in short bundles or fascicles. Occasional focal areas of nuclear palisading, whirling of the cells and parallel fibers are named Verocay bodies. Degenerative fibrillar myxoid areas in the paucicellular Antoni B areas and

Table 1. Summary of the intraoral ancient schwannomas including the presented two cases

No	Authors	Age, sex	Location	Size (cm)	Symptoms	Follow up
1	Eversole and Howell[5], 1971	58, F	Floor of mouth and ventral tongue	2.5	Nil	No follow up data
2	Marks et al.[6], 1976	65, F	Right floor of mouth	3.5	Nil	No complication, and no follow up data
3	McCoy et al.[7], 1983	36, F	Maxillary posterior vestibule	2.0	Nil	No follow up data
4	Dayan et al.[8], 1989	52, F	Left maxillary vestibule	0.9	Nil	No recurrence during one year of follow up
5	Nakayama et al.[9], 1996	40, F	Floor of mouth and ventral tongue	5.5	Asymptomatic swelling for 2 months	No recurrence during 2 years of follow up
6	Ledesma et al.[10], 1999	21, F	Floor of mouth and ventral tongue	3.0	Asymptomatic swelling for 5 month	Not available
7	Kim et al.[11], 2000	29, F	Origin of the lingual nerve	4.0	Swelling for 4 years	No recurrence during 1 year of follow up
8	Chen et al.[12], 2006	34, M	Floor of mouth	3.0	Asymptomatic swelling for 18 years	No recurrence during 2 years of follow up
9	Subhashraj et al.[13], 2009	18, M	Posterior vestibule of the mandible near the mental foramen	3.1	Asymptomatic swelling for 8 months	Transient neurapraxia for four weeks with no recurrence during 18 months of follow up
10	Amirchaghmaghi, et al.[14], 2010	14, M	Gingiva	1.5	Asymptomatic swelling for one year	Not described
11	Bilici et al.[15], 2011	45, M	Tip of the tongue	3.0	Disturbance in articulation and swallowing	Not available
12	Humber et al.[16], 2011	82, F	Upper lip extending from the midline to the canine eminence	2.0	Firm mass and intermittent mild paresthesia for about 2 years	No recurrence during 5 years of follow up
13	Kim et al, 2011 (the present case)	66, F	Posterior vestibule of the left mandible near the mental foramen	2.0	Asymptomatic swelling for 13 years	Paresthesia and no recurrence during 9 months of follow up
14	Kim et al, 2011 (the present case)	35, F	Left Mandible body	3.0	Asymptomatic	Dead with car accident before excision

(F: female, M: male)

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hyalinized vessels are the characteristic pathologic findings of schwannoma. Immunohistochemically, schwannoma typically shows a strong, diffuse reactivity for S-100 protein, whereas malignant peripheral nerve sheath tumor shows minimal and focal immunoreactivity for S-100 protein. Ancient schwannoma like the present two cases is a neurilemmoma displaying pronounced degenerative changes forming bizarre nuclear features with no mitotic figures. Prominent cellular atypia are regarded as purely degenerative changes because of no increased mitotic figures or necrosis identified. Ancient schwannomas cannot be clinically distinguished from conventional schwannomas, necessitating histologic evaluation. Case 2 could not be entirely examined because of incisional biopsy. However, the schwannoma showing above degenerating changes especially bizarre nuclear changes mimicking malignant counterpart even focal lesion may be the term ancient, i.e., degenerating schwannoma compatible. Intraoral ancient schwannomas behave the same as ordinary schwannomas[5-16]. Although there have been only a small number of reported cases, all except for four occurred in females, while there is no gender predilection for conventional intraoral schwannomas. Clinical features were summarized in Table 1.

The etiology and natural history of intraoral schwannoma have not been properly characterized, but the nerve sheath of origin has been regarded to be the branches of the mental nerve and inferior alveolar nerve[13]. In Case 1, considering its anatomic location near the mental foramen and the postoperative sensory changes of the left lower lip, the present case was also derived from the small branches of the mental nerve. Malignant transformation of intraoral schwannomas is rare, be it the conventional or ancient variant. Complete surgical excision is the treatment of choice for intraoral schwannomas[5-16], and the prognosis is excellent. When the tumor is well-encapsulated it is easily treated with surgery, and the difficulty resides in preserving the associated nerve by performing careful dissection.

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