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Tongue schwannomas associated with neurofibromatosis type 2

Harusachi Kanazawa [a, *], Hirotaka Sawai [b], Akihiro Kita [c], Nao Koide [c], Masanobu Yamatoji [c], Atsusi Kasamatsu [c], Katsuhiro Uzawa [c]

[a] Division of Dentistry and Oral-Maxillofacial Surgery, Sanmu Medical Center, 167 Naruto, Sanmu, Chiba 289-1326, Japan
[b] Department of Oral Science, Graduate School of Medicine, Chiba University, 1-8-1 Inohana, Chuo-ku, Chiba 260-8670, Japan
[c] Department of Dentistry and Oral-Maxillofacial Surgery, Chiba University Hospital, 1-8-1 Inohana, Chuo-ku, Chiba 260-8670, Japan

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ABSTRACT

Schwannomas are typically solitary benign neural tumors; however, multiple lesions associated with the rare genetic disorder neurofibromatosis type 2 (NF2) have been reported in some cases. We present the case of a tongue schwannomas in a 36-year-old woman previously diagnosed with NF2 with bilateral vestibular schwannomas. To alleviate difficulties with swallowing, tongue nodular masses were surgically removed, and schwannoma was histologically diagnosed. Our patient represents the first case of histologically confirmed tongue schwannoma associated with NF2. This indicates that in patient with NF2, schwannoma may be detected in the oral cavity as well as in other parts of the body. Thus, careful clinical and histological examinations are warranted to identify schwannomas associated with NF2 even in the oral cavity.

1. Introduction

Schwannoma or neurilemmoma is a benign tumor arising from the neural sheath Schwann cells of the cranial, spinal, peripheral, and autonomic nerves [1]. Extracranially, approximately 25%–48% of all schwannomas are found in the head and neck regions, of which only 1% appear intraorally, with the tongue being the most common location [2,3]. Schwannomas mostly present as solitary lesions but can present as multiple lesions in some cases when associated with the rare genetic disorder neurofibromatosis type2 (NF2) [3]. Therefore, schwannomas occurring in the oral cavity may also be the oral manifestations of NF2.

NF is a group of genetically distinct disorders characterized by the presence of multiple cutaneous lesions and tumors of the nervous system [4]. NF is categorized by the National Institutes of Health (NIH) diagnostic criteria (1988) into three subtypes. NF1 was previously referred to as the von Recklinghausen disease and accounted for 85%–90% of all NF cases; NF2 was previously referred to as bilateral vestibular or central neurofibromatosis; and schwannomatosis was once considered to be a variation of NF2 [5].

NF1 manifests as café au lait spots on the skin and multiple cutaneous neurofibromas of the peripheral nerves, whereas NF2 is characterized by the presence of characteristic slow-growing bilateral vestibular schwannomas of the eighth cranial

* Corresponding author. Division of Dentistry and Oral-Maxillofacial Surgery, Sanmu Medical Center, 167 Naruto, Sanmu, Chiba 289-1326, Japan.
E-mail addresses: h-kanazawa@sanmu-mc.jp (H. Kanazawa), aefa2771@chiba-u.jp (H. Sawai), akihiro.kita@chiba-u.jp (A. Kita), adsa2719@chiba-u.jp (N. Koide), yamatojim@chiba-u.jp (M. Yamatoji), kasamatsua@faculty.chiba-u.jp (A. Kasamatsu), uzawak@faculty.ciba-u.jp (K. Uzawa).

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nerves [6]. NF1 is known to present as oral manifestations of the soft tissues and jaw in up to 74% of the affected patients, with tongue neurofibromas being the most common [7]. However, no data is available regarding oral schwannomas as a manifestation of NF2 even though they can occur along any peripheral nerve in the body [8]. In this report, we present an extremely rare case of tongue schwannomas occurring in a patient with NF2.

2. Presentation of case

A 36-year-old woman was referred to our clinic with a chief complaint of swallowing difficulties because of painless elevated masses on the dorsal surface of the tongue. She reported that these lesions had slowly increased in size over the last 3 years. Her medical history revealed that she was diagnosed with NF2 with an initial symptom of hearing loss at approximately 30 years of age and had been monitored since then by the neurosurgery department. Her family history revealed that her parents had no disorders, but one of her two daughters was diagnosed with NF2. Magnetic resonance imaging (MRI) scans provided by the neurosurgery department revealed the development of a variety of tumors of the central and peripheral nervous systems: bilateral vestibular schwannoma and right trigeminal schwannoma in the cerebellopontine angle (Fig. 1A), three intracranial meningiomas (Fig. 1B), more than 20 spinal meningiomas (Fig. 1C), and 15 subcutaneous nodules (Fig. 1C) in the body. Intraoral examination revealed two smooth oval-shaped masses in the anterior (small, 8×6 mm) and posterior (large, 30×20) regions on the left dorsal surface of the tongue along the midline (Fig. 2A). On palpation, these masses exhibited elastic-soft consistency and mobility. Further, MRI confirmed some well-circumscribed masses as intracranial and spinal lesions (Fig. 2B). She had a normal oral mucosa and dentition. The clinical diagnosis was tongue neural tumor associated with NF2.

She underwent the surgical enucleation of the tongue lesions. On gross finding, these tumors were globular and well encapsulated yellowish masses and were easily shelled out (Fig. 3). The histological examination of the specimens revealed hypercellular spindle-shaped cell areas (Antoni A) with a predominant microscopic pattern and with only occasional hypocellular round cell areas (Antoni B) within a loose, myxomatous stroma (Fig. 4A). Characteristic histological signs of spindle cells arranged in palisading nuclei were observed in the Antony A areas (Fig. 4B). Immunohistochemical analysis revealed clear positive staining for S-100 in the tumor cells (Fig. 4C). The histological diagnosis was a schwannoma. The patient has had no recurrence during a follow-up period of 2 years.

3. Discussion

The characteristic finding of NF2 is the development of bilateral vestibular schwannomas, which are observed in more than 90% of NF2 cases, leading to hearing loss and subsequent deafness by the age of 30 years [6]. NF2-related schwannomas can occur along any cranial nerve, on spinal nerve roots, and on peripheral nerves in the body, but oral manifestations are rarely observed in NF2 patients [6]. A literature search of the PubMed database yielded only three cases of oral manifestations in patients with NF2 in the English literature over the past three decades, all of which reported a single, well-circumscribed, nodular lesion on the dorsal surface of the tongue [7–9]. No other soft tissue alterations were observed in the oral cavity. Due to the medical history of NF2 in these patients, the tongue lesions were initially suspected to be neural tumors associated with NF2. However, only two of the reported cases were histologically diagnosed as localized amyloidosis and amyloid tumors [7,9]. The third case was clinically diagnosed as neurofibroma or schwannoma but without histological confirmation [8]. None of these cases in this review was histologically confirmed as a schwannoma of the tongue associated with NF2 although the dorsal surface of the tongue is a common intraoral location for a schwannoma [10].

Interestingly, two of the reviewed cases were diagnosed as the localized amyloid lesions of the tongue with no systemic involvement of amyloidosis or multiple myeloma. Amyloidosis is a term used to describe a group of diseases in which an extracellular deposition of amorphous febrile proteins known as amyloids occurs [11]. Amyloid involvement in the tongue is almost always secondary to systemic amyloidosis and localized involvement is extremely rare [12]. However, clinicopathological data regarding NF2 patients with amyloidosis affecting oral tissues remains scarce [7,9]. Andreadis et al. (2011) have posited a random co-existence rather than a direct association for the concurrent presence of tongue amyloidosis and NF2 [9].

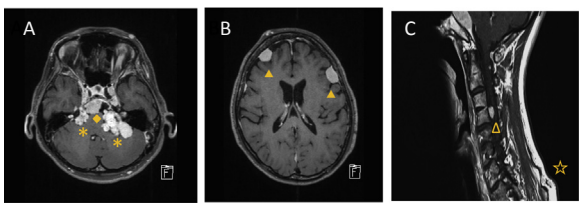

Fig. 1. MRI (A, B: intracranial axial view, C: cervical sagittal view) showing bilateral vestibular schwannomas (*) and right trigeminal schwannomas (▲) in cerebellopontine angle, intracranial meningiomas (◆), spinal meningiomas (Δ), and subcutaneous nodules (☆) with high signal intensity on gadolinium-enhanced T1-weighted images. MRI, magnetic resonance imaging.