A Case of a Huge Solitary Neurofibroma Extending from the Tongue to the Floor of the Mouth

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Abstract
We describe a case of huge solitary neurofibroma in the oral cavity. A 59-year-old man with a history of dysphagia presented with swelling of the tongue. Magnetic resonance (MR) images showed a lesion extending from the base of the tongue to the floor of the mouth located above the mylohyoid muscle. The dimensions of the enucleated tumor were 95 mm in length, 65 mm in width, and 15 mm in thickness. Histopathological examination revealed that the lesion was a diffuse neurofibroma.

Introduction
Neurofibroma is a benign nerve sheath neoplasm composed of a heterogeneous population of Schwann, perineural-like, and fibroblastic cells; it often occurs as a local manifestation of neurofibromatosis (von Recklinghausen’s disease) (1, 2). However, the occurrence of solitary neurofibroma in the oromaxillofacial region is relatively rare.

Here, we describe a case of huge solitary neurofibroma extending from the tongue to the floor of the mouth. Its pathological manifestations were consistent with diffuse neurofibroma.

Case Report
A 59-year-old man with a 3-month history of dysphagia presented with swelling of the tongue that had persisted for 1 year.

Oral examination revealed a 30 x 30 mm swelling in the lateral and dorsal regions of the right side of the tongue toward the base (Fig. 1A). The overlying mucosa was normal in color. An increased amount of mucosa in the anterior region of the floor of the

![Fig. 1. Swelling of the right portion of the tongue (A). Increased amount of mucosa in the floor of the mouth (B).](image-url)
mouth was also observed (Fig. 1B). Palpation of the tongue revealed that the swelling was an elastic, soft, smooth, nontender mass with ill-defined margins. Bimanual palpation of the floor of the mouth revealed similar findings.

There were no clinical signs such as multiple cafe-au-lait spots or a family history of neurofibromatosis.

Axial computed tomography (CT) images with soft-tissue windows demonstrated a low-density mass in the tongue and the floor of the mouth. Magnetic resonance (MR) imaging revealed a lesion with low T1-weighted signal intensity and high T2-weighted signal intensity (Fig. 2A, B, and C), extending from the base of the tongue to the floor of the oral cavity. The mass was located above the mylohyoid muscle. A clinical diagnosis of benign tumor was made.

Enucleation of the tumor was performed under general anesthesia via an oral approach.

Macroscopically, the enucleated mass was grayish yellow, solitary, and multicocular, measured 95 in length, 65 mm in width, and 15 mm in thickness, and was encapsulated by a thin fibrous membrane (Fig. 3).

Histopathologically, the lesion was composed of proliferating spindle-shaped cells with ovoid and elongated nuclei and scant cytoplasm that were embedded in a mucous-rich or variably collagenous matrix (Fig. 4A). Differentiation of the tumor cells to those mimicking meissnerian corpuscles was observed (Fig. 4B). A portion of the capsule was not visible. The tumor was surrounded by muscular and fatty tissue and a minor salivary gland, and diffuse infiltration of tumor cells into these tissues was observed. Immunohistochemically, most of the tumor cells were positive for S-100 protein (Fig. 5A), and some were positive for neuron-specific enolase (Fig. 5B) and glial fibrillary acidic protein. The histopath-
ological diagnosis was diffuse neurofibroma. Two-year follow-up data showed no recurrence of the tumor.

**Discussion**

Neurofibromas are usually associated with neurofibromatosis (von Recklinghausen's disease) and are usually found in multiple numbers, although they can occur as solitary tumors (3, 4).

Oral manifestations of neurofibromatosis have been reported in only 4–7% of reported cases (5). Solitary neurofibroma cases occur less frequently than multiple neurofibroma cases, which are manifestations of neurofibromatosis.

Solitary neurofibromas are benign, slow growing, and relatively circumscribed but nonencapsulated neoplasms, originating in a nerve and consisting of Schwann cells, perineural cells, and varying amounts of mature collagen (6). Because the tumor observed in our case showed pseudo-meissnerian corpuscles, it was histopathologically classified as a diffuse neurofibroma. Clinically, diffuse neurofibroma is an uncommon but distinctive form that generally occurs in children and young adults; it is most commonly observed in the head and neck region and presents as a plaque-like elevation of the skin (2).

In the literature, the size of solitary neurofibromas has been reported to vary from $3 \times 4$ mm to $70 \times 50$
mm, with most lesions measuring less than 15–20 mm (2, 7, 8). Thus far, only 2 cases of large lesions have been reported. The first case was a large neurofi-
broma in the oral cavity that measured 70×50 mm (detailed characteristics of this tumor have not been reported) (7), and the second case was reported in the soft palate and measured 50×40 mm (8). The present case of neurofibroma is the largest reported thus far (95×65×15 mm); it appeared as 2 separate lesions, one on the tongue and the other on the floor of the mouth. A neurofibroma with such characteristic size and shape is very rare. In addition, the progression of the tumor seemed to be slow and likely would have gone unnoticed until dysphagia developed. Because the progression of solitary neurofibromas is slow and rarely accompanied by subjective symptoms, most neurofibromas are discovered accidentally.

Recurrence of solitary neurofibromas is extremely rare (7); this lack of recurrence was also observed in the present case, in which no recurrence was noted during the 2–year follow-up period.

References