



# A Case of 27-Year-Old Localized Solitary Neurofibroma of the Base Tongue; A Case Report

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Received January 24, 2022; Accepted April 26, 2022; Online Published May 18, 2022

## Abstract

**Introduction:** Neurofibromas, benign peripheral nerve sheath tumors, are the most common among neural lesion. Neurofibromas are rare in head and neck location, although it may involve any part of oral cavity. However, tongue is the most common region.

**Case Presentation:** A 27 year-old man was admitted to Otorhinolaryngology Department due to a swelling in the base of his tongue, since two weeks before. This mass was painless, and without any associated symptoms. He was suffered from a mild eating disorder, too. He had no any positive past medical history, and only a large soft 6 × 5 cm mass was found at the base of his tongue without bleeding in the oral cavity. Excisional biopsy was performed for him under general anesthesia and the tumor was easily enucleated. The final diagnosis was neurofibroma. After the diagnosis of neurofibroma, the physical reexamination revealed a series of café au lait spots on his skin.

**Conclusion:** the neurofibroma should be considered by otorhinolaryngologist, as a differential diagnosis of tongue mass, and more accurate physical examination are recommended for better management.

**Keywords:** Neurofibroma, Neurofibromatoses, Tongue

## 1. Introduction

Neurofibromas are benign peripheral nerve sheath tumors, which are the most common among neural lesion, with unknown etiology. They may present either as solitary lesions or as part of the generalized syndrome of neurofibromatosis or skin von Recklinghausen's disease. Mostly, they are considered as hallmark of a generalized syndrome of neurofibromatosis, a rare inherited autosomal dominant genetic disorder.<sup>1-5</sup> It was reported that only 10% of neurofibromas are associated with neurofibromatosis.<sup>6</sup> Neurofibromatosis can involve almost any organ system. Thus, the signs and symptoms presented may vary widely.<sup>7</sup>

The most common manifestations of this disorder are café au lait spots, retinal tumors, freckles in axillary region, and iris involvement, and its diagnosis should be considered when three or more of them are present.<sup>1,5</sup> Neurofibromas are rare in head and neck location, although they are most common among neural lesion in this region and they may involve any part of oral cavity.<sup>6</sup> Tongue is the most common region, which is presenting as a large mass in an isolated setting.<sup>4,7</sup> The prevalence of oral manifestation of neurofibromatosis was reported 4%-7%.<sup>8</sup> They can be in different forms in clinicopathology: localized (sporadic or solitary), diffuse, plexiform (less common), and epithelioid.<sup>5,6,9</sup> Localized form develops

along a peripheral nerve as a focal mass with well-defined margins without any encapsulated<sup>5</sup>. Solitary form is more frequent (90%).<sup>9</sup>

In this case report, a patient with painless swelling at the base of his tongue will be present, which was diagnosed as localized solitary neurofibroma after excisional biopsy. The physical reexamination revealed a series of café au lait spots on his skin.

## 2. Case Presentation

A 27 year-old man was admitted to Otorhinolaryngology Department of Imam Khomeini Hospital, one of the biggest teaching hospital in Kermanshah, west of Iran, because of a swelling in the base of his tongue, since two weeks before. This mass was painless, and without any associated symptoms. He was suffered from a mild eating disorder (dysphagia), too. He had no any positive past medical history, and his vital signs were stable at the time of admission. Otorhinolaryngology examination was normal, and only a large soft 6 × 5 cm non-tender and firm mass was found at the base of his tongue without bleeding (Figure 1).

Laboratory data showed a white blood cell of 5.3 10<sup>9</sup>/L, hemoglobin of 15.5 g/dL, platelet of 311 (10<sup>3</sup>/μL), PT of 12 seconds, and INR of 1. With the initial diagnosis of lingual



**Figure 1.** A 6 × 5 cm Non-tender Firm and Soft Mass in the Base of Tongue Without Bleeding In Intraoral Clinical Examination.

thyroid, thyroid function tests were performed, which were normal (Table 1).

Thyroid ultrasonography showed a few small cervical lymph node, as well as sublingual lymph node with preserved hilum and a picture of inflammatory lymph node. Moreover, spiral neck computed tomography (CT) scan with and without intravenous (IV) contrast showed a soft tissue density mass measuring 4.5×3.5 cm in right upper lobe near his superior mediastinum. Magnetic resonance imaging (MRI) of neck with IV contrast was done which showed a well-defined, enhancing cyst lesion in the base of tongue measuring about 2.6×2.1 cm. A related ultrasound showed internal debris and cystic nature. Also, a well-defined 4.9×3.3 cm right side extra spinal cyst was reported at the root neck causing compression of lower cords of bronchial plexus. It was suggestive for infected thyroglossal duct remnant incidental meningeal cyst causing right side bronchial plexus compression.

Excisional biopsy was performed for him under general anesthesia, and the tumor was easily enucleated (Figure 2). Pathology report indicated a creamy soft piece measuring 3×2.5×2 cm covered by a piece of tongue measuring 2×2×0.5 cm. The final diagnosis was localized solitary neurofibroma.

After the diagnosis of neurofibroma, the physical reexamination revealed a series of café au lait spots on his skin that he did not talk about them in the taking of medical history. He was discharged with good condition.

### 3. Discussion

In the cases of tongue masses, some differential diagnosis should be considered such as; neurofibroma, schwannoma, malignant neoplasm, lingual thyroid, lymphangioma, hemangioma, hamartoma, teratoma, pyogenic granuloma, lipoma, nerve sheath myxoma, vascular malformations, tongue infections, and cystic lesions such as mucoid, foregut duplication and dermoid cysts. Neurofibromas are a benign nerve sheath neoplasms, which are uncommon in head and neck region.<sup>2,5,6</sup> However, tongue involvement is the most common in this region for neurofibroma, which is presenting as a large mass in an isolated

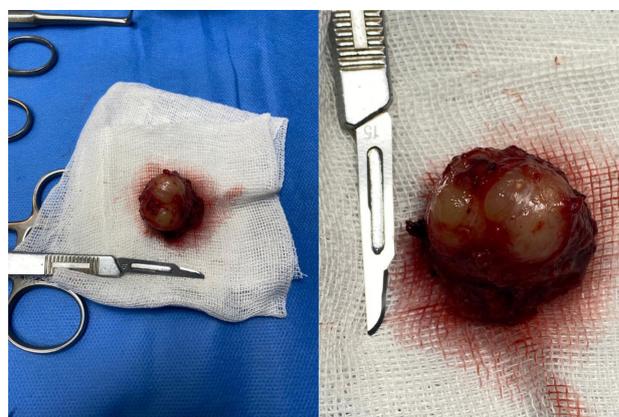
setting.<sup>4,7</sup> Neurofibroma classically originates from one of the terminal branches of the lingual and hypoglossal nerves, however, it is regularly difficult to determine the specific origin of the involved nerves.<sup>1</sup> Head and neck neurofibroma presents with upper airway obstruction, swallowing difficulty, mastication deficits, or cosmetic distortion of the face.<sup>5</sup> Since neurofibromas are usually multiple lesions, the whole body must be investigated, especially larynx and trachea due to its effect on the upper airway and respiratory obstruction.<sup>3</sup>

CT scan is not a sensitive imaging modality for small tumors of the soft tissue, but it should be considered, especially to determine the size and vascularity of the tumor before surgery, since the surgeon needs to obtain clear surgical margins to avoid recurrence.<sup>10</sup> However, MRI is a choice modality for diagnosis of neurofibromas lesions, that low to intermediate signal intensity on T1-weighted (T1W1) and high signal intensity on T2-

**Table 1.** The Results of Laboratory Tests

Laboratory Tests	Results
RBC count (10 <sup>6</sup> /μL)	5.4
WBC count (10 <sup>9</sup> /L)	5.3
Lymphocyte (%)	35
Hb (g/dL)	15.4
Hct (%)	46.6
Platelet (103/μL)	311
PT	12
PTT	35
INR	1
Blood sugar (mg/dL)	85
T4 (μg/dL)	9.1
T3 UP (%)	27
T3 (ng/dL)	0.9
TSH (μIU/mL)	1.2

RBC: red blood cell; WBC: white blood cell; Hb: hemoglobin; Hct: hematocrit; INR: international normalized ratio; PT: prothrombin time; PTT: partial thromboplastin time; TSH: Thyroid-stimulating hormone.



**Figure 2.** Excisional biopsy of Mass in the Base of Tongue Showing a Multilobular Mass Partly Encapsulated By a Thin Membrane, Measuring 3 × 2.5 × 2 cm with a Covered by a Piece of Tongue Measuring 2 × 2 × 0.5.

weighted (T2W1) images are suggestive. It is worth mentioning that neurofibroma and Schwannoma appear similar in MRI, but degenerative changes in nerve can be recognized in Schwannoma.<sup>4,6</sup> The definite diagnosis can only be confirmed by histological examination.<sup>6</sup> Most of the neurofibromas are without encapsulated like what we presented in this report. Also, neurofibroma is immunopositive for the S-100 protein in most of the cases which indicate its neural origin.<sup>5</sup> Complete surgical excision is a standard treatment for neurofibroma regardless of whether the tumor is single or multiple. It can be performed with laser, electrocautery, harmonic scalpel or coblation depending on the surgeon's decision. Neurofibroma has extensive vascularity and tend to bleed during surgical removal.<sup>2,3,6</sup> Recurrence of neurofibroma is extremely rare, but patient' follow-up have been recommended.<sup>8</sup>

In this report, we described a patient with localized solitary neurofibroma in the base of his tongue, which is a rare case. Güneri et al explained a 3 year-old girl with a gradually growing painless mass in the tongue presented since birth. She suffered from disturbance of mastication and phonation. Finally, lingual plexiform neurofibroma was diagnosed, and it was treated by total excision.<sup>3</sup>

Moreover, a 59 year-old man with swelling of the tongue from 1 year and dysphagia from 3 months was reported by Tanaka et al. MRI revealed a lesion extending from the base of the tongue to the floor of the mouth, which was located above the mylohyoid muscle. After excisional biopsy, histopathological examination confirmed it as diffused neurofibroma.<sup>9</sup> Similarly, Sharma et al. presented an 11 year-old girl with a painless enlarging mass of the tongue, which was diagnosed as isolated plexiform neurofibroma, and it was treated with intraoral surgery.<sup>9</sup> Lykke et al also reported a 62 year-old woman with irritation and dysaesthesia of the lateral aspect of her tongue. Only a slightly red area was found in oral examination. The histopathological findings of biopsy showed a small, rounded tumor closely approximated to an invagination of the surface epithelium and with a small lymphatic infiltrate and it was diagnosed as neurofibroma. No other neurofibromas or skin lesions were reported and in her follow-up, all symptoms had reported to be resolved.<sup>10</sup>

A 14 year-old male with dysphagia for one year was described by Singh et al. In physical examination, a pinkish irregular growth approximately 3.5 cm in transverse diameter was found in right side of base of tongue. It was immobile, non-tender, firm, and non-compressible in examination. A MRI scan was performed and reported a heterogeneous hypointens mass on T1W1 and hyperintense in T2W1. Due to a delayed post contrast enhancement, hemangiolympangioma was raised as diagnosis. Empirical treatment with oral propranolol and local sclerosant (phenol) injection was not effective in size reduction, and surgical excision with coblation was performed. Histopathological examination was suggestive for plexiform neurofibroma. They reported that

postoperative swelling was less than expected and there was a minimal pain and a quick recovery.<sup>6</sup>

In another report, Roy et al. presented a 75 year-old man with a gradually growing painless mass at the right side of the base of the tongue with slight difficulty in mobility and difficulty in swallowing for 4 months. It became painful during preceding 4 weeks. It was found as a firm in consistency, slightly tender, non-ulcerative mass with irregular surface in examination. After deep incisional biopsy, histopathology reported it as neurofibroma with typical feature. No skin pigmentation and evidence suggestive of any systemic disorders were found, and also his family history was negative. Hence, isolated neurofibroma of the tongue was raised and was advised excision, but he declined.<sup>5</sup>

Ballage et al also reported an 18 year-old male patient with a history of a slowly growing painless mass in his tongue. A pinkish red soft swelling with an irregular surface involving the right side of the mobile tongue approximately 4×5 cm in size on nonreducible and nonpulsatile on palpation were found in his oral cavity examination. Surgical removal was done after biopsy, and plexiform neurofibroma of the base of the tongue was diagnosed. After that, multiple café au lait spots were discovered on his trunk, and he was referred to a cancer center for further management and follow-up.<sup>2</sup>

Based on the afro-mentioned, awareness about the neurofibroma as one of the main differential diagnosis of tongue mass can prevent additional diagnostic procedures. Early diagnosis is essential and these patients need regular follow-up during their lifetime to detect recurrences.<sup>2</sup>

#### 4. Conclusion

The neurofibroma should be considered by otorhinolaryngologist, as a differential diagnosis of tongue mass, and more accurate physical examination are recommended for better management. Early diagnosis is essential and these patients need regular follow-up during their lifetime to detect recurrences

#### Authors' Contributions

All authors performed the surgery, reviewed the literature, wrote and revised the manuscript, and read and approved the final manuscript.

#### Conflict of Interest Disclosures

The authors declared that they have no conflict of interest.

#### Ethical Approval

The patient gave permission for the presentation of his case reports. Also The patient's written consent was obtained for the publication of this case report.

#### Funding/Support

None.

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