NEUROFIBROMA OF THE LINGUAL NERVE: A CASE REPORT

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A neurofibroma of the lingual nerve is a rare clinical finding, the most common lesion site of a lingual neurofibroma being the tongue. In most situations, it is difficult to determine the precise nerve origins. Herein, we report a case of lingual nerve neurofibroma that presented as a submandibular mass, mimicking a submandibular gland tumor or solitary lymphadenopathy. Complete surgical excision of such a lesion for histopathologic examination provides a better treatment and final diagnosis. For a patient presenting with neurofibromatosis and a submandibular mass, a neurofibroma of nerve origin should be considered in the differential diagnosis.

Key Words: lingual nerve, neurofibroma


CASE PRESENTATION

A 24-year-old woman who visited our otolaryngology department complained of a bulging mass over the left submandibular area, which had persisted for the last 2 years. According to the patient’s statement, the mass had progressively enlarged over the last 1 month; she denied any inflammatory or infective conditions during this period, and claimed no abnormalities associated with food and taste. There was no weight loss, night sweats, rigors or low-grade fever associated with the patient’s condition. An investigation of patient and family past medical history showed that the patient, her mother, and her younger brother had suffered from NF. Scattered café au lait spots and multiple cutaneous neurofibromas were distributed around the patient’s neck and trunk.

Physical examination of the patient confirmed a palpable, firm, well-defined, and nontender mass in the left submandibular triangle, with a diameter of approximately 4 cm. Computed tomography (CT) examination of the region showed, on heterogenous enhancement, a spherical, nodular tumor located in the left submandibular space (Figure 1). Under the diagnostic impression of a submandibular gland tumor or lymphadenopathy, surgical exploration was undertaken via a standard submandibular approach. The surgical findings highlighted a well-defined mass adherent to the left submandibular gland. On removal
and dissection of the submandibular gland, it was unexpectedly found that this mass had originated from the main trunk of the lingual nerve without any apparent feeding vessels (Figure 2). Complete excision of the well-defined mass was then performed. Upon gross inspection, the lesion appeared to be a 5×3×2.5 cm well-encapsulated, yellowish mass of a rather soft consistency. Histopathologic examination showed well-demarcated tumor growth with proliferative spindle cells intermingled with myxoid and fibrotic stroma (Figure 3). The final diagnosis of a neurofibroma arising from the lingual nerve was confirmed by pathology. There was no tumor recurrence during the subsequent 6-month follow-up period.

**DISCUSSION**

NF is a heterogeneous spectrum of disorders, and, to the best of our knowledge, a positive family history is reported for perhaps half of all confirmed cases [2,3]. NF type 1 (NF-1) is the most common form, a range of diverse neurocutaneous and soft-tissue lesions constituting the major manifestations. NF-2 is characterized by the development of bilateral acoustic neuromas [2,4,5]. To the best of our knowledge, at the time of writing, no definite clinical manifestations have been used to specifically identify or differentiate between these two forms of NF. The present case was categorized as NF-1, showing specific features of café au lait spots, cutaneous neurofibromas, and a neurofibroma of the lingual nerve.

Among patients suffering from NF, head and neck neurofibromas typically feature a predilection to develop in the deep planes of the neck, for which the incidence of occurrence is typically low [6]. Neurofibromas have also been reported to occur in the parotid and parapharyngeal spaces [1,7], being characterized by lesions that feature progressive and rapid enlargement causing obvious clinical symptoms [7]. As regards CT manifestations of neurofibroma, they can best be determined with heterogeneous
enhancement. This may be related to the mixed-fat content of involved Schwann cells, myxoid tissue, and cystic degeneration [3]. The characteristic CT features of NF are virtually the same as those of salivary gland tumors and lymphadenopathies. A neurofibroma of the lingual nerve is, reportedly, extremely rare; such a lesion typically originates from one of several terminal branches of the lingual and hypoglossal nerves [4], and it is often difficult to determine the specific origin of the involved nerves. As a consequence, a hypertrophied tongue featuring enlarged fungiform papillae is the most common clinical expression of NF leading to a firm diagnosis without detection of the specific nerve origins [2–5].

In our case, the submandibular mass was the first sign of this condition upon initial patient presentation. On heterogeneous enhancement of the left submandibular gland during preoperative CT, a well-defined mass featuring associated compression of the left submandibular gland constituted a characteristic feature. It was, thus, reasonable to suspect that the tumor had developed from the submandibular gland or lymph nodes. As a consequence, it would appear to be difficult to differentiate a lingual nerve neurofibroma from a submandibular gland tumor or lymphadenopathy. It may be possible that a thin and flat submandibular gland may arise as a result of the mass effect of a neurofibroma, thus providing another means of preparing a comprehensive differential diagnosis. In addition to radioimages, fine needle aspiration may be another procedure that can assist in preoperative diagnosis, especially in patients with known NF-1.

With regard to this very rare and, for us, unexpected finding of a neurofibroma originating from the main trunk of the lingual nerve in the submandibular space, we suggest that complete surgical intervention is the best method to provide the most accurate diagnosis. Malignant transformation of NF-I is reported to occur in 2–30% of cases [1,2,4]. When solitary neurofibroma compresses adjacent tissues or produces malignant manifestations, complete surgical excision of the lesion is usually possible for solitary and well-encapsulated neurofibromas [1,2]. With regard to this patient’s postoperative status, the sensation of taste was slightly decreased, but the motor function of mastication and swallowing was the same as her preoperative condition. We should weigh and compare the consequences of malignant transformation and permanent lingual nerve dysfunction when deciding whether or not to proceed with surgical intervention.

In conclusion, a neurofibroma of the lingual nerve is very rare. It is, generally, quite difficult to differentiate a neurofibroma from a submandibular gland tumor from preoperative imaging. Consequently, a neurofibroma arising from either the lingual or hypoglossal nerve should be considered in the differential diagnosis of a patient suffering from NF and featuring a submandibular mass.

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REFERENCES

源自舌神經之神經纖維瘤 — 病例報告

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源自於舌神經之神經纖維瘤在臨床上極為少見，病灶處以舌頭最常出現。大部分的神經纖維瘤均無法明確判斷神經的來源。我們報告一位神經纖維瘤病的患者，以下頜部腫瘤為表現的臨床病例，術前診斷為來自下頜腺或是淋巴腺之腫瘤。經由完整的手術切除及病理組織學檢查，提供正確的診斷。因此罹患神經纖維瘤病的患者，出現下頜腺腫瘤時，分佈於此處的神經所長出的神經纖維瘤亦須納入鑑別診斷。

關鍵詞：舌神經，神經纖維瘤

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