

Neurofibroma in Anterolateral Aspect of Tongue

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Abstract

Neurofibromas (NFs) are rare neurogenic tumors of the oral cavity. They can present as solitary or multiple when associated with neurofibromatosis-1 (NF-1)/von Recklinghausen's disease. Solitary intraoral NFs unassociated with NF-1 are sporadic and mimic any other soft tissue tumor of the oral cavity. The authors report a case of a 49-year-old deaf-mute lady who presented with slowly growing solitary mass in the anterolateral aspect of the tongue, which was successfully managed in the outpatient clinic with complete excision and base cauterization under local anesthesia without complications.

Keywords: Tongue neurofibroma, oral lesion, surgical excision.

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Introduction

Neurofibroma (NF) is a benign tumor of neuronal origin that occurs as a solitary lesion, or as multiple lesions associated with neurofibromatosis type 1 (NF1), also known as von Recklinghausen's disease which is a systemic condition caused by a germline mutation in the NF1 tumor suppressor gene located at 17q11.2^[1]. Neurofibromas are generally asymptomatic but may become painful, irritated or pruritic. These tumors can occur anywhere but are most common in the trunk followed by the head and neck. When multiple, they can be seen in a segmental or widespread distribution. Cutaneous neurofibromas may be dermal or subcutaneous. Plexiform neurofibroma is the term used to refer to diffuse neurofibromatosis of nerve trunks, often associated with an overgrowth of the skin and subcutaneous tissues. This is a bit distinct from the neuronal type^[2].

Solitary NFs occur in any region of the body, commonly found in skin, deep nerves, and in association to other retroperitoneal tissues. Oral cavity solitary plexiform neurofibromas are rare. The first case of oral solitary NF was reported in 1954^[3].

The following is a description for the diagnosis and treatment of an anterolateral

tongue neurofibroma.

Case Report

A 49-year-old lady known to have bilateral profound hearing loss since birth presented to the outpatient clinic with a left-sided tongue mass. The history was taken with the help of her son using sign language. The patient noticed this mass five years ago, right after biting her tongue accidentally while chewing with the mass continuing to increase slowly in size since then. It was always painless and non-bloody. However, she kept accidentally biting the same lesion while chewing food causing pain and discomfort, which was the main complaint when she visited the clinic. This was her first time seeking medical help about this lesion.

She had no other similar lesions or any history of abnormal movements or seizures. She denied having chronic headache, neck pain, backache or visual problems.

Upon examination, the lesion was pinkish, soft in consistency, pedunculated and polypoid in shape. Located at the left-sided dorsum of the anterior part of her tongue, it was about 1.5 cm in its greatest dimension, non-pulsatile and non-tender, as seen in figure 1. Her entire skin was examined with no findings of café-au-lait spots, armpits freckles or other skin nodules. No

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imaging was done at that time.

The lesion was excised in the clinic under local anesthesia using lidocaine spray. A 3-0 tie was used to devascularize and separate the mass from its attachment to the tongue. Afterwards, cauterization at the site of mass attachment using silver nitrate stick prevented bleeding. The excised mass was sent for histopathology.

The final histopathology results showed a pedunculated mass, measuring 1.5 x 1.0 x 0.5cm, with an overlying orthokeratinized epithelium. The underlying dermis consisted of proliferation of spindle-shaped cells with occasional serpentine (wavy) nuclei. There were occasional scattered mast cells in a well-vascularized fibrocollagenous connective tissue stroma, interspaced with areas of myxoid matrix, as seen in figure 2. The final interpretation was a solitary dermal neurofibroma negative for malignancy.

Discussion

Peripheral nerve tumors are a heterogeneous group of mostly benign tumors that are rare in the general population. Certain types, including neurofibromas and schwannomas may occur sporadically or in association with NF⁽⁴⁾.

Symptoms and signs of peripheral nerve tumors are caused by direct nerve invasion, involvement of surrounding tissues or mass effect⁽⁵⁾. Patients present for evaluation of peripheral nerve tumors because of a soft tissue mass, pain or focal neurologic findings, approximately in that order of frequency⁽⁶⁾. The duration and progression of symptoms or signs are important, as most benign tumors have a longer duration and a slow rate of progression, while malignant tumors tend to progress rapidly in size, amount of pain and neurologic deficit^(6, 7).

The category of benign nerve sheath neoplasms includes neurofibroma, schwannoma, perineuroma, hybrid nerve sheath tumors, cellular and mixed-type neurothekeoma, dermal nerve sheath myxoma, ganglioneuroma and hemangioblastoma. A significant minority of peripheral nerve tumors are associated with either NF1 or NF2. Schwannomatosis is now considered to be a

third variety of NF⁽⁸⁾.

Neurofibromas are tumors composed of a mix of Schwann cells, perineural-like cells and fibroblasts, interspersed with nerve fibers, wire-like strands of collagen and myxoid matrix^(9,10). The majority is solitary (up to 90%) and not associated with NF1. The patient is often young, between 20 and 30 years of age⁽⁹⁾.

There is no pathological difference between neurofibromas from patients without and with NF1⁽¹¹⁾, except that systemic and hereditary factors present in the disseminated form are absent in the solitary type. However, neurofibromas are pathologically and genetically different from schwannomas. The neoplastic cell of origin is the Schwann cell but the presence of intratumor nerve fibers is a cardinal feature that helps distinguish neurofibromas from schwannomas^(10, 11).

Oral cavity involvement by a solitary and peripheral plexiform neurofibroma in patients with no other signs of neurofibromatosis is uncommon⁽¹²⁾. Sporadic cases have been reported in the submandibular gland, tongue and the periosteum at the mental foramen. This sporadic syndromic occurrence has also been seen in the cutaneous region, and several authors have suggested that these isolated neurofibromas may represent a hamartomatous growth⁽¹²⁾.

Neurofibromas are usually classified by location and appearance. The first type, cutaneous or dermal neurofibromas, are small, nodular tumors of the skin and subcutaneous tissue arising from small cutaneous nerves, which may cause local pain or bleeding but do not cause neurologic deficits. They consist of soft, fleshy, sessile or pedunculated tumors⁽¹³⁾. They move with the skin on examination and are not tender. Some are located within the dermis and can be palpated as a soft spot in the skin, often with an overlying violaceous discoloration. These cutaneous lesions tend to increase in size and number with age and vary in number from just a few to thousands, with the highest density occurring over the trunk. In NF1, they are by far the most common form of tumor. They are benign and do not carry an increased risk of malignant transformation⁽¹⁴⁾.

The second type, intraneural neurofibromas, are deeper, focal, well-circumscribed fusiform lesions involving nerve roots, nerve trunks, nerve plexuses, or peripheral nerves. The nerve enters the mass at one end, exits at the other, and is intermixed with tumor components throughout the length of the mass. These tumors usually present as a mass, with local or radicular pain, or with sensorimotor neurologic symptoms and signs [15]. Tumors within the epineurium will be encapsulated, but most extend beyond the epineurium, and though circumscribed, they are not encapsulated.

These two types (cutaneous and intraneural) could be considered discrete tumors [15], and they comprise 90% of neurofibromas.

Plexiform neurofibroma, a third type, is non-discrete. These are multinodular elongated masses affecting large nerves that may appear as a tangle of thick strands or as aropy mass resembling a “bag of worms” [10]. These are nearly pathognomonic of NF1 but rarely can be seen as solitary lesions [9].

Massive soft tissue neurofibromas are large, diffuse masses causing regional or single-limb enlargement (formerly known as elephantiasis) seen in NF1 patients [10]. Solitary intraneural neurofibromas account for approximately two-thirds of neurofibromas removed at surgery [16], with the others being associated with NF1.

Multiple neurofibromas are nearly diagnostic of NF1. They can be found affecting virtually any nerve or plexus; they affect a young patient population (20 to 30 years of age) and, for an unknown reason they occur more frequently on the right side than the left [11]. Solitary neurofibromas rarely transform to malignancy, but discrete neurofibromas in NF1 can become malignant. Moreover, plexiform neurofibromas carry a substantial risk of becoming malignant.

Imaging can be helpful in localizing the mass as well as determining its extent and any involvement of extraneural tissue. On computed tomography (CT), intraneural neurofibromas are well-demarcated masses that are hypodense to muscle [15]. Contrast enhancement is minimal, if any. On magnetic resonance imaging (MRI), neurofibromas show

low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [9]. They enhance frequently and usually heterogeneously. On T2 images, the tumors will often show a characteristic central hypointense region, called the target sign, which is presumed to be due to dense collagen and fibrous tissue in the center. This pattern is very suggestive of neurofibroma but has been reported in schwannoma and in malignant peripheral nerve sheath tumors [9].

Treatment of solitary intraneural neurofibromas is dependent on symptoms and signs. Cutaneous neurofibromas should not be removed, whether the patient has NF1 or not, unless there is a reason such as pain, bleeding, interference with function, or disfigurement [17].

Solitary neurofibromas can be managed by observation if the patient is asymptomatic. For patients with pain, with progressive neurologic deficit or in whom the diagnosis is uncertain, surgical resection becomes necessary [11, 18]. Solitary intraneural neurofibromas suspicious for malignancy should either be resected or first biopsied and then resected if malignant.

For patients with solitary or multiple intraneural neurofibromas or plexiform neurofibromas requiring treatment, surgical resection with preservation of neurologic function is the goal [19]. However, this is more likely to be accomplished in patients with solitary neurofibromas than patients with NF1.

A number of plexiform neurofibromas will degenerate into a more aggressive nodular lesion with increased 18-fluorodeoxyglucose positron emission tomography (FDG-PET) avidity [20]. These nodular lesions grow more aggressively than plexiform neurofibromas themselves and degenerate into malignancy. Special attention, and occasionally surgery, must be considered for these rapidly changing lesions.

Conclusion

A 1.5 cm neurofibroma mass uncommonly presented in the anterolateral aspect of the tongue and was easily excised in an outpatient clinic using local anesthesia without complications. The patient was able to eat and

swallow after 2 hours with minimal discomfort which resolved after few days later.

Compliance with Ethical Standards

The authors deny any potential conflict of interest with no financial support to be declared.

All procedures performed in this study were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments. Informed consent was obtained from the participant included in the study.

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ورم العصب الليفى في المنطقة الامامية الجانبية للسان

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الملخص

الاورام الليفية العصبية (NFs) في تجويف الفم أورام عصبية نادرة يمكن أن تظهر بشكل فردي أو متعدد عندما ترتبط بالورم الليفى من النوع الاول (NF 1).الاورام الليفية العصبية المنفردة داخل الفم غير المرتبطة بـ NF 1متفرقة وتشبه أي من اورام الانسجة الرخوة الاخرى في تجويف الفم. يصف المؤلفون حالة سيدة صماء بكفاء تبلغ من العمر 49 عاما اتت الى العيادة الخارجية وفي المنطقة الامامية الجانبية من لسانها كتلة انفرادية تنمو ببطء تم علاجها بنجاح في العيادة مع الاستئصال الكامل تحت التخدير الموضعي دون مضاعفات. الكلمات الدالة: الورم الليفى العصبى في اللسان، آفة الفم، الاستئصال الجراحي.