CASE REPORT

Lingual neurofibroma causing dysaesthesia of the tongue

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Neurofibroma is a benign tumour derived from the neural sheaths of peripheral nerves and composed of Schwann cells, fibroblast-like cells and intermediate cells. The usual clinical presentation of an oral neurofibroma is swelling. A 62-year-old woman was referred to our department of otorhinolaryngology with irritation and dysaesthesia of the lateral aspect of the tongue. The only finding was a slightly red area from which a biopsy was taken. The macroscopic findings observed by the surgeon were consistent with normal tongue tissue. The histopathological examination showed a small, rounded tumour closely approximated to an invagination of the surface epithelium and with a small lymphatic infiltrate. The tumour was a neurofibroma. A Schwannoma type B was considered but the presence of small nerves and positive neurofilament reaction favoured a neurofibroma. The patient had no other neurofibromas or skin lesions. At a 3-week follow-up visit all symptoms had resolved.

BACKGROUND

A neurofibroma is a rare slow-growing benign tumor derived from the neural sheaths of peripheral nerves and composed of Schwann cells, fibroblast-like cells and intermediate cells.^{1 2} The histopathological findings include spindle-shaped cells with thin, wavy nuclei immersed in a collagenous matrix. Often small axons pass through the tumour and mast cells are abundant.^{2–6} About 25% of all neurofibromas arise in the head and neck region, but may be found anywhere on the body. Isolated neurofibromas in the oral cavity are very rare. The usual clinical presentation of oral neurofibroma is swelling which in some cases is accompanied by discomfort. Neurofibromas can be part of the neurofibromatosis, but these patients often display multiple tumours affecting different anatomical sites as well as other stigmata of the disease, such as café au lait macules (neurofibromatosis Recklinghausen).^{3–7} The risk of malignant transformation of a solitary neurofibroma is extremely low but can occur, most often when associated with neurofibromatosis Recklinghausen.^{2 3 8} The correct treatment of a neurofibroma is complete surgical resection after which the frequency of recurrence is low.³ We report a case of an isolated microscopic neurofibroma of the tongue presenting only as dysaesthesia. With this report we want to raise the awareness that patients with unusual symptoms of irritation or dysaesthesia of the oral cavity might have microscopic disease.

CASE PRESENTATION

A 62-year-old woman born and raised in Turkey was referred to our department of otorhinolaryngology with symptoms of excessive mucus production of the nose and irritation of the rhinopharynx and oropharynx. The patient described an unusual sensation of constant irritation on the right lateral aspect of her tongue, which made her spit in a handkerchief several times during the first visit. The sputum was unremarkable. The otorhinolaryngeal objective assessment showed some extent of non-purulent mucous in her nasal cavity and dysaesthesia of the right posterolateral part of the tongue. The mucosa was slightly erythematous, but no tumour or ulcer was visible. A biopsy, which at the subsequent histopathological examination was shown to be an excisional biopsy of a microscopic lingual neurofibroma, was taken from the area. No other lesions, especially no café au lait macules, were found on the patient and her family's medical history was negative for neurofibromatosis Recklinghausen. When the patient returned for her follow-up after 3 weeks she reported that the symptoms from the tongue had disappeared after the biopsy was taken. As she persisted to suffer from excessive mucus production from the nose and sinuses, she eventually had functional endoscopic sinus surgery performed with success.

INVESTIGATIONS

The only visible change of the mucosa was a slightly erythematous area from which a biopsy was taken using local anaesthesia (lidocaine–epinephrine 0.5%) and a sharp Weil Blakesley through cutting forceps (Miltex, Germany). The macroscopic findings of the biopsy observed by the surgeon were consistent with normal tongue tissue. No unusual bleeding was observed. The histopathological examination using H&E staining showed a small, rounded, unencapsulated tumour closely correlated to an invagination of the surface epithelium and a small lymphatic infiltrate. The tumour was excised in toto, when taking the biopsy. The tumour cells were elongated spindle cells with pale, eosinophilic cytoplasm and elongated, buckled nuclei. In between small nerves could be identified. The stroma was fibromyxoid and small capillaries were observed. There were scattered mast cells (figure 1). The tumour was positive for S-100 and was also stained by immunohistochemistry using neurofilament antibodies (figures 2 and 3). The tumour was diagnosed as a neurofibroma. A Schwannoma type B was considered but the

To cite: Lykke E, Noergaard T, Rasmussen ER. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-010440

Figure 1 Histopathological slide showing H&E staining. The tumour cells are elongated spindle cells with pale, eosinophilic cytoplasm and elongated, buckled nuclei (arrow).

presence of small nerves and positive neurofilament reaction favoured a neurofibroma.

A CT revealed no mucosal swellings of the tongue, but it seemed that the drainage from the frontal sinuses were not sufficient. No other imaging was performed in this case since a tumour was not visible.

An MRI would be the suitable imaging modality to assess tumours of the oral cavity, both as a diagnostic tool and to plan a possible surgical intervention.

DIFFERENTIAL DIAGNOSIS

- ▶ Isolated schwannoma
- ▶ Neurofibromatosis, that is, Recklinghausen or Schwannomatosis
- ▶ Localised infections of the mucosa
- ▶ Stomatitis
- ▶ Rheumatological or other systemic disease with oral manifestations
- ▶ Malignant tumours of the oral cavity
- ▶ Dermoid cyst

TREATMENT

Further treatment was not indicated since the tumour was excised in toto when taking the biopsy. The frequency of recurrence is extremely low after complete resection³

Figure 2 Immunohistochemical staining using S-100 antibodies indicating the presence of neural crest derived cells; in this case Schwann cells (arrow).

Figure 3 Immunohistochemistry using neurofilament antibodies for staining. The brown colour indicates the presence of neurofilaments (arrow).

OUTCOME AND FOLLOW-UP

When the patient returned for her follow-up visit after 3 weeks, she reported complete remission and has not since returned to our clinic with symptoms arising from the oral cavity.

DISCUSSION

The described patient was atypical compared with other patients suffering from neurofibromas of the tongue, since the clinical findings usually include a visible tumour.^{3 4 7 8} The most frequent intraoral sites involved are the gingiva and the alveolar ridge,³ further emphasising the uniqueness of this case. This neurofibroma clearly had arisen in a small sensory nerve originating from the glossopharyngeal nerve which innervates the posterior third of the tongue. An MRI is a useful tool when diagnosing and planning surgical excision of tumours of the oral cavity, but might not be readily accessible on all locations. In such cases CT should be considered, especially to determine the size and vascularity of the tumour before surgery, since the surgeon needs to obtain clear surgical margins to avoid recurrence. To the best of our knowledge we are the first to report on a microscopic neurofibroma only presenting as dysaesthesia. We would like to raise the awareness of this possibly underdiagnosed disease.

Learning points

- ▶ Neurofibromas of the tongue do not always present as a visible mass.
- ▶ CT is not a sensitive imaging modality for small tumours of the soft tissue.
- ▶ Even small neurofibromas can cause significant discomfort.
- ▶ Biopsy of symptomatic areas should most often be performed.

Acknowledgements We would like to thank Hilleroed Hospital ENT department for our use of an examination room.

Contributors EL was responsible for the idea, writing of manuscript, finding valuable reference articles, patient communication and approval of the final manuscript. TN was involved in histopathological examination, writing and revising of the manuscript and approval of the final manuscript. ERR was responsible for the

idea, writing of manuscript, critical revision, finding of reference articles, patient communication and approval of the final manuscript.

Competing interests ERR has received payment for teaching at one scientific meeting sponsored by MSD Norway.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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