CASE REPORT

Schwannoma of the tongue

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SUMMARY

Schwannoma, neurilemmoma or neurinoma is a rare, benign nerve sheath neoplasm composed of Schwann cells. It is usually solitary, slow growing and asymptomatic. Approximately 1–12% of the tumours occur intraorally with the tongue being the most common location. We report a rare case of lingual schwannoma in a 20-year-old female patient involving the tip of the tongue, which was slow growing and asymptomatic. For diagnosis histopathological examination and also immunohistochemistry testing were carried out to confirm the nature of tissue fragments. The treatment was complete surgical excision.

BACKGROUND

Schwannoma is a benign, most commonly encapsulated nerve sheath tumour.^{1 2} The neoplasm can occur alone³ or is a part of genetically inherited diseases such as neurofibromatosis type 1 (NF1), NF type 2 (NF2) and schwannomatosis.^{4 5} Aetiology is connected to the mutation of the *NF2* gene which is located on chromosome 22q12.2 and encodes the merlin protein. *NF2* gene functions as a tumour suppressor and a regulator of Schwann cells.^{4 6 7} Precise prevalence figures are not known.^{2 8} The tumour can affect the peripheral, cranial and autonomic nerves.⁹ Occurrence of all schwannomas in the head and neck is 25–45% and of these 1–12% can be found intraorally.⁹ In the oral cavity the tongue is most frequently affected, followed by the roof of the mouth, the floor of the mouth, the buccal mucosa, the gingiva, the lips and the vestibular mucosa.¹⁰ In the tongue, identification of the originating nerve (hypoglossal, glossopharyngeal and lingual) is difficult given their proximity. These lesions most commonly appear between the second and fourth decade of life. There is no predilection of gender or race.¹¹ The tumour presents as a solitary, slow growing and smooth surfaced mass, which is generally asymptomatic but can also cause pain and discomfort. The intensity of symptoms is determined by the location and size of the tumour. The usual treatment is simple surgical excision. Recurrence rate after surgical excision is low.¹² Malignant alteration is rare.⁵ The case of a female patient with a schwannoma of the anterior part of the tongue, which was excised in local anaesthesia, will be presented.

CASE PRESENTATION

A 20-year-old female patient was sent to the Department of Maxillofacial and Oral Surgery at

the University Medical Center Ljubljana. The patient reported a slowly enlarging painless mass on the tip of the tongue. The mass was first noticed a year ago. The patient was otherwise healthy with no significant personal or family medical history. Intraoral clinical examination revealed a nodule 13×13 mm in size (figure 1). The mass was firm and non-tender. There was no fluctuation. Cervical lymph nodes were not palpable. The remaining clinical examination was unremarkable. Fine-needle aspiration biopsy (FNAB) was performed, which showed tissue fragments consisting of cells aligned to produce nuclear palisading as well as dissociated nuclei and elements of peripheral blood. Immunohistochemistry showed positive staining for S-100 protein. The diagnosis of schwannoma was suggested. No radiological investigations were needed because the mass was easily visible and palpable. The mass was excised under local anaesthesia (figure 2). About 2 mL of articaine solution (40 mg of articaine chloride per 1 mL, 0.005 mg epinephrine per 1 mL) was applied at the apex of the tongue to allow a painless removal of the tumour. A small (10 mm) incision was needed and the tumour was easily extirpated (figure 3). No haemostasis was needed. Absorbable sutures were

Figure 1 Clinical manifestation of the mass on the tip of the tongue.

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Figure 2 Intraoperative view of the schwannoma.

Figure 4 The wound was closed using absorbable sutures.

Figure 3 Intraoperative view after tumour extirpation.

used to close the wound (figure 4). The procedure and the post-operative period were uneventful. After 1 week the sutures were removed (they were causing only a minor discomfort to the patient). The wound had healed without any signs of secondary intention (figure 5). Gross examination revealed an encapsulated nodule 15×9×9 mm in size with a smooth surface which had a moderately firm white grey cut surface (figure 6) and with histopathological examination the diagnosis of schwannoma was confirmed (figure 7). The final examination was carried out 1 month after the operation (figure 8). The patient reported a very slightly changed sensation on the tip of the tongue that

Figure 5 Clinical presentation after 1 week.

posed no functional disturbances. It was explained to the patient that further improvement in this symptom can be expected. Additional control visit only in case of problems was agreed with the patient.

Figure 6 Gross view of the excised tumour.

Figure 7 Histological picture with a typical biphasic appearance and Verocay bodies (H&E, original magnification $\times 100$). Image courtesy of assistant Dr Katarina Odar, DMD, PhD, Institute of Pathology, Faculty of Medicine, University of Ljubljana.

Figure 8 Clinical presentation after 1 month.

DISCUSSION

Schwannoma is a benign tumour, which clinically appears as a slow-growing mass. It originates from Schwann cells of the nerve sheath.⁹ The most commonly affected nerve in the head and neck region is the vestibulocochlear nerve. In the oral cavity, however, schwannoma is rare (1%).¹³ The tongue is the most common location of occurrence intraorally. Wright and

Jackson reviewed 146 cases of oral schwannomas, where they found that the tongue was represented in 52%, buccal or vestibular mucosa in 19.86%, soft palate in 8.9% and the gingivae and lip in 19.24% of cases.¹⁴ Schwannoma has no affinity for gender or race and it can occur throughout a wide age range.^{12 15} Most commonly it occurs between the ages of 10 and 40 years. In the tongue, schwannomas appear on two different parts. The majority of cases (around two-thirds) involve the oral portion of the tongue. Approximately one-third of cases involve the base of the tongue. In these cases symptoms such as pain, dysphagia, dysphonia, sleep apnoea, bleeding and infection can be present. The majority of schwannomas of the tongue, however, clinically appear as a painless submucosal nodule.¹¹ Butler, Patel and McHugh performed a retrospective study on the clinicopathological features of schwannomas. The majority of cases were asymptomatic. Local symptoms were, however, caused by tumours in the oral cavity and larynx and neurological symptoms were most often associated with tumours arising adjacent to or within the bone. Although schwannomas are generally encapsulated, this was not always the case in their study. Tumours arising from the nasal cavity, paranasal sinuses and larynx lacked a capsule.² On histopathological examination, schwannoma is characterised by two patterns of tissue growth. The first is composed of Schwann cells arranged in a cellular palisaded pattern (Antoni type A) with Verocay bodies. The second (Antoni type B) has a looser and disorganised arrangement.¹⁶ Both patterns are also seen in the presented case (figure 7). The diagnosis is confirmed by immunohistochemical markers S-100 and SOX10, which are used in most cases.¹¹ In the presented case, S-100 protein was positive. Malignant transformation of schwannoma is unusual and when it does occur it is usually in the form of malignant peripheral nerve sheath tumour (MPNST) or even more rarely angiosarcoma. The lifetime risk of MPNST in the setting of NF 1 is $\sim 2\%$.⁵ The diagnosis is suggested by FNAB and confirmed by histopathological examination.¹ Fine needle aspiration was performed because in the oral cavity a round tumour can be malignant and consequently the treatment would be different.¹⁷ Imaging investigations include ultrasound scanning, CT and MRI.¹ Most schwannomas are shown by non-enhanced CT to be well-circumscribed, dense and homogeneous soft-tissue masses.¹⁸ MRI is the imaging modality of choice for detailing the extent of the tumour¹ and is the first choice among other imaging modalities for examination of the base of tongue. MRI shows the tumour as a smooth and well-demarcated lesion which is isointense to muscle on T1-weighted images and homogeneously hyperintense on T2-weighted images.³ In the presented case no imaging was necessary since the tip of the tongue presented an easily accessible location with no neighbouring structures causing any dilemmas regarding the treatment plan. Multiple schwannomas can be a manifestation of NF 2 or schwannomatosis, which have to be considered as separate entities. The hallmark signs of NF 2 are vestibular schwannomas, whereas in case of schwannomatosis, which is a form of NF 2, these tumours are absent.⁴ Differential diagnosis should, in addition to schwannomas and neurofibromas, include granular cell tumours, leiomyomas, irritation fibromas, hemangiomas, rhabdomyomas, lymphangiomas, lipomas, pyogenic granulomas and benign salivary gland tumours.⁹ Complete surgical excision is the treatment of choice. Schwannoma rarely recurs, but when it does, it is usually due to incomplete excision.¹⁰ In the presented case the outcome was good with complete functional and aesthetic restoration.

Learning points

- ▶ Schwannoma of the tongue is a rare benign neoplasm.
- ▶ It is usually solitary, slow growing and asymptomatic.
- ▶ The diagnosis is suggested by fine-needle aspiration biopsy and confirmed by a histopathological examination.
- ▶ The treatment is complete surgical excision.
- ▶ Malignant transformation and recurrence of schwannoma are rare.

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