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Case Report

Case of submandibular schwannoma and review of literature ☆

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ABSTRACT

Schwannomas are slow growing, benign tumours arising from Schwann cells. They are usually solitary and are sometimes associated with Neurofibromatosis type 1 and 2. As reported by Okada et al., while approximately 25%-40% of extra-cranial schwannomas occur in the head and neck region, Schwannomas of the oral cavity are very uncommon, accounting for only 1% of all Schwannomas. We report a case of a sublingual schwannoma in a 47-year-old female, discovered incidentally during the workup for tinnitus. The radiological and histopathological findings, along with a literature review, are presented.

Introduction

Schwannomas are a benign neoplasm arising from epineural Schwann cells (which produce the myelin sheaths surrounding cranial and peripheral nerves) [1]. Schwannomas typically present as a slow-growing, smooth-surfaced mass, and although usually solitary, can also be multifocal (usually in the setting of an underlying genetic inherited disorder such as NF1 or NF2). They are typically asymptomatic and most commonly occur between 30 and 50 years of age, with no strong gender predilection [1]. Schwannomas can occur along any central or peripheral nerve.

Symptoms and presentation relate to local mass effect dependent on the location and size of the lesion [1]. Detection of these lesions is often incidental, owing to their slow-growing nature and frequently deep-seated location [3,4]. As such, imaging is often critical and superior to clinical evaluation [1,2].

Schwannomas can sometimes provide a diagnostic dilemma, particularly when occurring in atypical locations, such as in the oral cavity. However, even when presenting in

Abbreviations: SMA, Skeletal muscle actin; CD43, Cluster of differentiation 43; STAT 6, Signal transducer and activator of transcription 6; NF1, neurofibromatosis 1; NF2, Neurofibromatosis 2.

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an atypical location, Schwannomas typically retain the same key imaging features, and correct imaging diagnosis can be important in avoiding more invasive surgical intervention.

We present a case of an incidentally detected floor of mouth schwannoma, which was suspected radiologically based on overall imaging characteristics, and was able to be proven histologically on core biopsy.

Case report

A 47-year-old female presenting with a history of chronic headaches and new onset left sided pulsatile tinnitus was referred by an ENT specialist for an MRI brain (to exclude an underlying intra-cranial pathology).

MRI findings

An MRI brain demonstrated no intra-cranial abnormality and no clear cause for the patient's pulsatile tinnitus. However, an incidental finding was made of a well circumscribed ovoid lesion within the left floor of mouth (only visible on the post-contrast T1 sequences). The lesion was located at the posterior aspect of the left sublingual gland, posterior to the mylohyoid sling and measured 21 × 13 × 20 mm (AP x TR x CC). It demonstrated relatively homogeneous contrast enhancement, with some low signal intensity foci centrally within the lesion (Fig. 1) thought to most likely representing cystic change. Based on the salivary gland location, well circumscribed margins and vivid contrast enhancement, the initial imaging differential diagnosis given was of a pleomorphic adenoma (or other salivary gland neoplasm), or a nerve sheath tumor (such as a schwannoma).

The patient proceeded to have a dedicated MRI of the region of interest to further characterize the lesion. A multiphase, multisequence contrast MRI of the floor of mouth demonstrated that the lesion was isointense to muscle on T1 and hyperintense on T2, with intense post contrast enhancement (Fig. 2). There were no locally aggressive features or restricted diffusion evident, with no other lesions identified within the head/neck and no locoregional lymphadenopathy. Based on overall imaging characteristics, the favored differential remained of a nerve sheath tumor or a salivary grand neoplasm such as a pleomorphic adenoma.

After discussion with the referring ENT surgeon, decision was made to proceed to an attempted ultrasound guide core biopsy. Under ultrasound guidance, four passes with a 20 G core needle were made into the left sublingual gland lesion.

Histopathology

Four core biopsy specimens were received, measuring 12, 11, 8, and 7 mm respectively. Microscopic analysis revealed tissue with a variably cellular populations of bland spindle cells and focal Verocay body formation (Fig. 3). The lesional cells were diffusely positive for S100 (Fig. 4) and negative for SMA, CD34 and STAT6. The features were in keeping with a Schwannoma, with no evidence for malignancy.

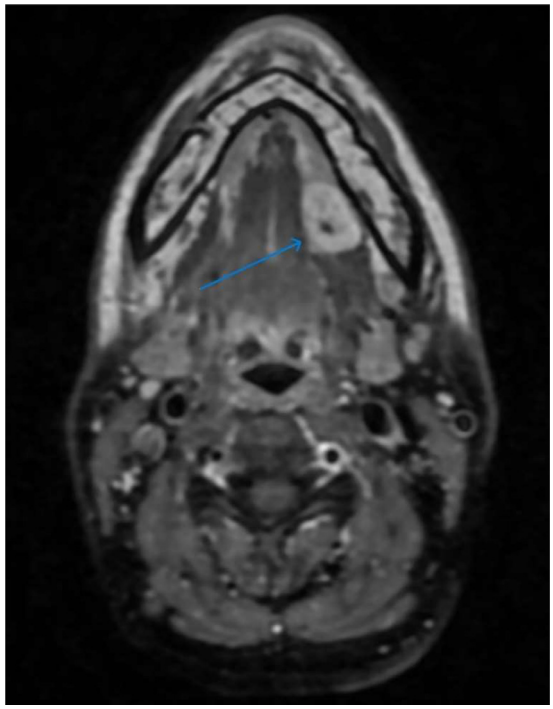

Fig. 1 – Axial post contrast T1. Incidentally detected enhancing lesion left floor of mouth (blue arrow) on MRI brain performed for work-up of tinnitus. Lesion is located at the posterior aspect of the left sublingual gland, and measures 21 x 13 x 20 mm (AP x TR x CC). Central region of non-enhancement within the lesion reflecting an internal cystic component.

Discussion

Oral cavity, and specifically floor of mouth Schwannomas are exceedingly rare in the literature. Oral cavity schwannomas are estimated to account for approximately 1% of the total incidence of schwannomas, with the most common oral cavity site being the tongue [1,5]. A 10-year review of Schwannoma cases across two tertiary centres in Brazil documented only 1 of 12 cases of intra-oral schwannomas occurring in the floor of mouth [6]. A literature search of the terms "sublingual schwannoma, floor of mouth schwannoma, Sublingual neurilemmoma" from 2000 to 2022 in English yielded 15 cases, 1 of the mylohyoid nerve, 10 of the hypoglossal nerve, 3 within the submandibular gland, and 1 unspecified [4,5,7–9].

Imaging plays an important role in schwannoma detection and diagnosis. Imaging appearances may be strongly suggestive or raise the possibility of the diagnosis, based on a combination of lesion location and the presence of typical imaging findings [8]. On computed tomography, schwannomas usually appear as well encapsulated, hypodense lesions with vivid contrast enhancement (although the degree of enhancement can be variable and depends on the volume of Antoni B fibres within the lesion). On MRI schwannomas are usually T1 iso to hypointense and T2 hyperintense with intense post-contrast enhancement [1].

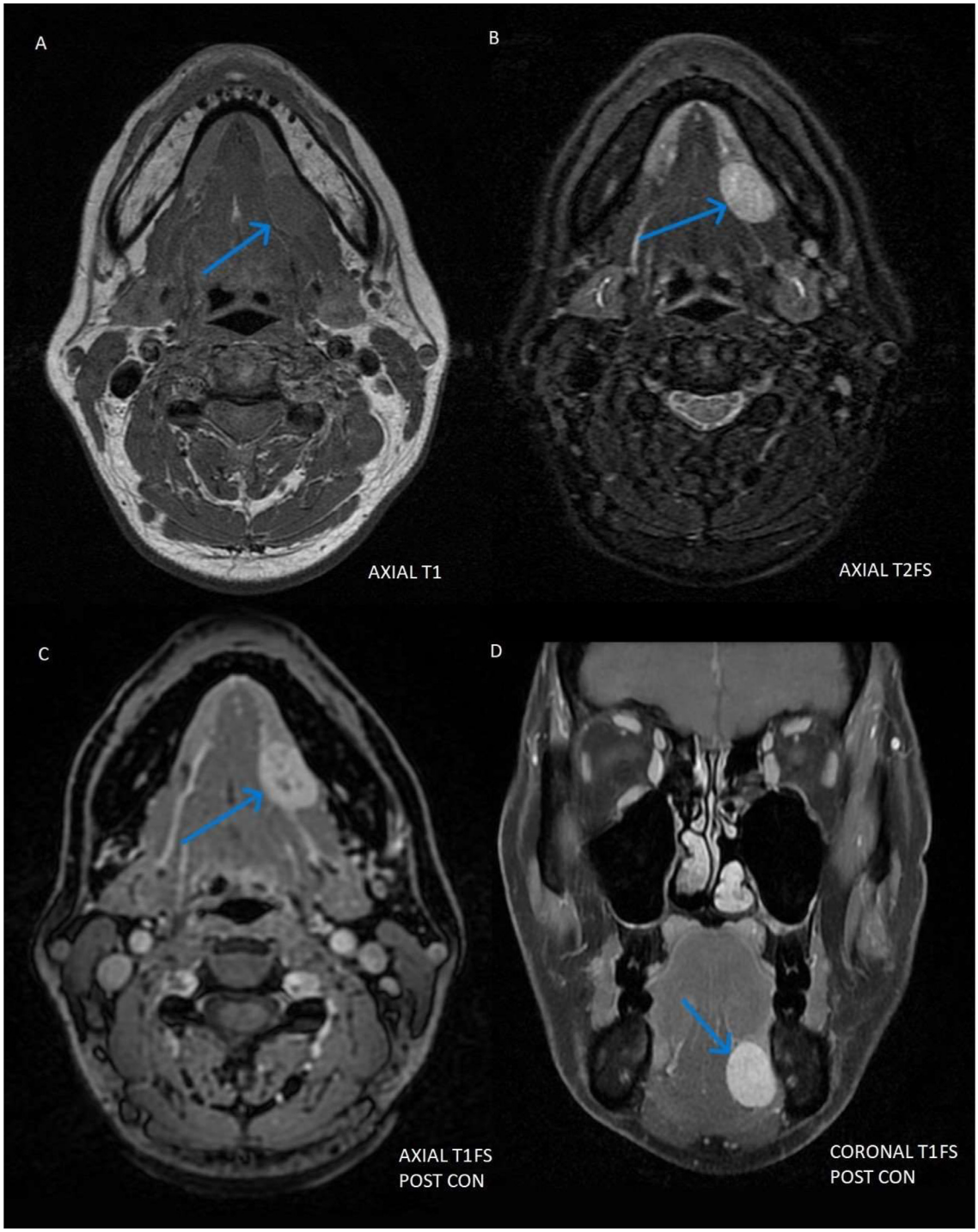

Fig. 2 – Multisequence MRI floor of mouth. A dedicated contrast MRI of the floor of mouth demonstrates the lesion (blue arrows) to be isointense to muscle on T1 (A) and hyperintense on T2 (B), with intense post contrast enhancement (C, D). The lesion has relatively well circumscribed margins and was not associated with any restricted diffusion or features of local invasion. No further oral cavity lesions or locoregional lymphadenopathy evident.

In addition to the perineural location, there may be several other suggestive morphological features, such as an elongated appearance (due to growth along the course of a nerve) and internal cystic or fatty degeneration (particularly in larger lesions), occasionally with hemorrhage [7]. Due to their slow growing nature, they may be associated with adjacent smooth bony remodelling, and typically displace adjacent structures rather than invade them.

Microscopically, conventional schwannomas are composed of spindle cells that demonstrate 2 growth patterns:

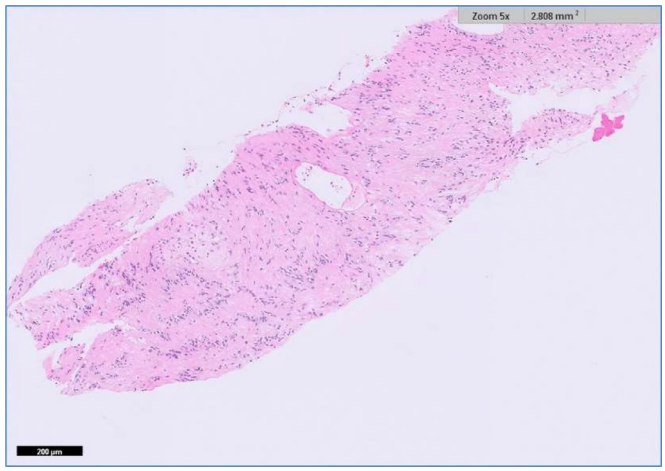

Fig. 3 – H and E stain of the specimen demonstrating spindle cell tumor with Verocay bodies.

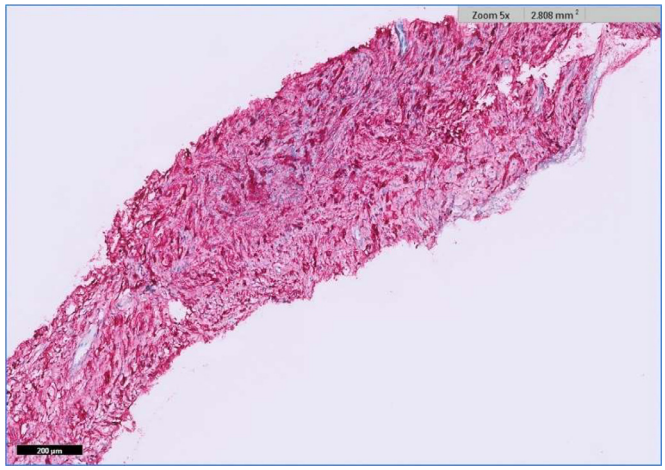

Fig. 4 – Immunohistochemical staining of lesional cells were diffusely positive for S100 (red chromogen).

Antoni type A and Antoni type B. The Antoni type A pattern is characterized by elongated or fusiform cells form dense bundles in organized fascicles. A pallisaded distribution surrounding eosinophilic regions, Verocay bodies, may be evident [1,5].

In contrast, Antoni type B feature less compact cell arrangements that are prone to cystic degeneration. The cells are scattered haphazardly within a loose myxoid matrix, with vacuolated cytoplasm and fibres without specific orientation associated with interstitial oedema and microcysts. The respective proportions of Antoni A and B fibres affect the imaging appearance of the lesion, with high Antoni B fibre populations being associated with greater T2 hyperintensity and post contrast enhancement [6]. Further immunohistochemistry (IHC) stains include diffuse positivity for s100, and positivity with Vimentin and Neuron specific enolase is seen [1,6]. Negative IHC stains for SMA excludes muscle origin, while negative CD34 and STAT 6 distinguishes solitary fibrosis tumors as a schwannoma mimic [10,11].

The gold standard for management of schwannomas remains surgical excision with definitive diagnosis by histological examination [12]. Fine needle aspiration has been reported as providing overall low accuracy, and may not be adequate for diagnosis particularly if imaging features are uncertain [2,11]. Malignant transformation of schwannoma is rare [1,6].

Although associated with low rate of recurrence, excision is not without its risks in otherwise asymptomatic patients and is highly dependent on the specific location of the individual lesion and proximity to adjacent structures [6]. In a surgical review of 9 cases of hypoglossal nerve schwannomas, 3 cases of asymptomatic presentations underwent complete excision, of which 1 case required transection of the main hypoglossal nerve trunk, resulting in hypoglossal nerve palsy [4,8]. A recently published consensus guideline on management of Schwannomatosis recommends surgical excision only in symptomatic individuals, where surgery is unlikely to result in neurological deficits [13]. In this case, the patient was asymptomatic and opted for radiological surveillance. There

is no consensus on timing of repeat imaging. The ERN recommends frequency be guided by clinical judgement based on presence of changing symptoms [13]. Follow up MRI performed at 6 and 12 months demonstrated slow increase in size: 25 × 15 × 23 mm and 26 × 15 × 25 mm (previously 21 × 13 × 20 mm). The patient remained asymptomatic.

Conclusion

We present an unusual location of a biopsy proven schwannoma, detected incidentally on imaging. Such cases highlight the importance of recognising the typical imaging features of Schwannomas, with an appreciation for the wide variety of locations in which they can occur. The floor of mouth is a very rare location, with our literature search yielding only one other reported case of a sublingual schwannoma.

Imaging diagnosis remains key in the work-up of Schwannomas, as they are often detected incidentally and may be difficult to access surgically depending on the location. Given their indolent nature it is important to differentiate them from more aggressive mimickers which may occur in similar locations, however, may have differentiating imaging features. Given their peri-neural location, surgical excision has the potential to result in neurological deficits and associated morbidity in otherwise asymptomatic and incidental presentations. In some cases, early imaging diagnosis or differential consideration of a schwannoma may help reduce the need for more aggressive intervention (such as surgical biopsy or excision).

Patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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