CASE REPORT

Submandibular schwannoma: is excision a 'gold standard treatment'?

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SUMMARY

A 58-year-old man was referred to ear, nose and throat (ENT) with an 8-week history of painless right submandibular swelling with no history of weight loss, dysphagia or difficulty with mastication. On examination, there was enlargement of the right submandibular gland, which was diffuse, nodular and firm; but no calcification of the duct or cervical lymphadenopathy. ENT examination including fiberoptic laryngoscopy was normal. Fine-needle aspiration of the lump was inconclusive. Neck ultrasound scan showed a well-defined 3 cm×3.8 cm×4.3 cm heterogeneous mass with significant internal vascularity, anterior to the right submandibular gland. MRI (T2-weighted MRI) revealed a neurogenic tumour or a non-sarcomatous soft tissue lesion. Complete excision of the lump was performed by intracapsular dissection and the specimen was submitted for histology, which confirmed the diagnosis of 'Ancient schwannoma' of the submandibular gland. The management resulted in complete resolution of symptoms with no cranial nerve deficits.

BACKGROUND

Schwannomas of the submandibular gland are rare causes of extracranial neurogenic tumours.¹ Twenty-five per cent of extracranial tumours occur in the head and neck region, with 1% occurring in the intraoral region.² The current literature has shown reports and studies on head and neck schwannomas, nonetheless, no study has been conducted specifically on the optimal management of submandibular schwannoma.

CASE PRESENTATION

A 58-year-old man presented to the general practitioner with an 8-week history of painless right submandibular swelling with no history of weight loss, dysphagia or difficulty with mastication. He was subsequently referred to the ear, nose and throat (ENT) department using the 'urgent suspected cancer' pathway. On examination, there was enlargement of the right submandibular gland, which was diffuse, nodular and firm, and measured approximately 5 cm×4 cm in diameter. There was no calcification of the duct or cervical lymphadenopathy. ENT examination including fiberoptic laryngoscopy was normal. Medical history included ulcerative colitis (no medical management) and no surgery. Family history was significant for brain cancer (mother). The patient was an artist/painter. He was a non-smoker with minimal alcohol consumption of 1–2 units/month. He had no known drug allergies and was not on any regular medication.

INVESTIGATIONS

Fine-needle aspiration of the lump was inconclusive. Ultrasound scan of the neck showed a well-defined heterogeneous mass with significant internal vascularity, which was anterior to the right submandibular gland and measured 3 cm×3.8 cm×4.3 cm. The rest of the neck examination was normal. These findings were inconclusive and thus necessitated MRI for further evaluation.

MRI (T2 weighted), revealed a well-defined lesion with displacement of surrounding structures including the tongue, mylohyoid muscle, and submandibular and sublingual glands with no aggressive features or adenopathy (figures 1 and 2). Findings were suggestive of a neurogenic tumour or a non-sarcomatous soft tissue lesion.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of submandibular swelling includes:

- ▶ Sialadenitis
- ▶ Benign and malignant neoplasms
- ▶ Cystic swellings
- ▶ Mumps
- ▶ Calcification
- ▶ HIV infection
- ▶ Salivary gland enlargement
- ▶ Lymphadenopathy.

Figure 1 MRI—no contrast T2-weighted MRI: mass in the right submandibular space.

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Figure 2 Postcontrast MRI: mass in the right submandibular space.

Figure 3 Intraoperative procedure picture: intraoperative procedure showing submandibular lump delivered from the incision.

TREATMENT

Complete excision of the lump was performed by intracapsular dissection and the specimen was submitted for histopathological examination to establish the final diagnosis (figures 3 and 4).

OUTCOME AND FOLLOW-UP

The patient was reassessed 6 months postoperatively, and showed no tongue paraesthesia or paralysis suggestive of nerve damage.

Figure 4 Histology picture: schwannoma composed of uniform spindle shaped cells with focal palisading of nuclei (Antoni A pattern)—black arrows, lying in a pale oedematous stroma with focal cyst formation (Antoni B pattern)—blue arrow. Prominent thick walled blood vessels are also a feature of this neoplasm—green arrows.

DISCUSSION

Neurilemmomas often present as painless solitary nodules of variable size; however, they may present as multiple lesions commonly associated with neurofibromatosis type II.³ The aetiology of neurilemmomas is associated with deletion and/or monosomy of chromosome 22.³ Histologically, the morphology usually appears circular with encapsulation, characterised by biphasic pattern of Antoni A and Antoni B cells microscopically.³ Additionally, schwannomas exist as five variants, namely, 'plexiform schwannoma', 'ancient schwannoma', 'cellular schwannoma', 'melanotic schwannoma' and 'pacinian schwannoma' schwannomas.³ The above case report showed a lesion consistent with that of an ancient schwannoma, and this usually occurs as a degenerative deep-seated lesion with manifestations of nuclear pleomorphism and rare mitotic activity.² Ancient schwannomas are characterised by four features, namely: (1) Verocay body; (2) palisading cells; (3) biphasic pattern of Antoni A and Antoni B cells and (IV) encapsulated mass with degenerative changes.^{3 4} With this, pain and paraesthesia are uncommon symptoms and these were concordant with the symptoms displayed by the patient and, thus, suggestive of this specific diagnosis. Clinical manifestations depend on the tumour location, size and nerve of origin.⁵ Evidently, the patient discussed had no symptoms other than the slow-growing submandibular mass.

Although most are benign in nature, malignant schwannomas can also occur.⁶ The incidence of malignant peripheral nerve sheath tumour is rare: 0.0001%.⁷ Malignant peripheral nerve sheath tumours strongly express protein S-100 and collagen IV—laminin. Imaging findings correspond to heterogeneous enhancement, irregularity of contour and necrosis, in some cases. Compression and local invasion of adjacent structures are also noted, but some malignant tumours are indistinguishable from benign lesions. There are no specific radiological signs oriented towards malignant changes. The risk of malignant transformation of head and neck schwannomas varies from 8% to 10%.⁸ Therefore, all clinical and radiologically diagnosed schwannomas need to have a histological diagnosis. In our case, the fine-needle aspiration or core biopsy was inconclusive, although radiologically, it was diagnosed to be a schwannoma.

The histological distinction between neurofibroma, neurofibroma with atypical features and malignant schwannomas is difficult to make, because there is a continuum between these lesions.⁷ Preoperative diagnoses of this tumour is challenging because of non-specific clinical symptoms and signs. Radiological investigations in the form of CT, MRI or ultrasound yield very low specific diagnosis. Fine-needle aspiration

cytology is accurate in only 20% of cases.⁵ Hence, the treatment is complete surgical excision of the tumour and postoperative histopathological examination to establish the final diagnosis.⁹

Usually, the tumour is located eccentrically in the nerve and is well enveloped by a capsule consisting of perineum from the nerve bundle of origin. The remaining nerve fascicles are contained within the epineurium and, in most instances, are functional.¹⁰ Bondi *et al*¹⁰ described methods of dissecting the healthy fascicles with longitudinal incisions on the nerve and then exposing the tumour. Their experience with 18 patients confirmed minimal neurological deficit after such procedures.¹⁰

The nerve of origin of the tumour was not identified in this case, but a careful intracapsular dissection was performed to remove the lesion in its entirety.¹¹

A study by Calcaterra *et al*¹¹ noted that the nerve of origin is usually not identifiable and nerve preserving intracapsular nucleation does not guarantee intact nerve function after surgery.

Patient's perspective

Overall, the surgical team members were fantastic and I couldn't speak too highly of them. With such a rare tumour and all the things that could go wrong with a major surgery, they did a marvellous job. I have the highest praise for all the medical staff involved in my care.

Learning points

- ▶ Extracranial schwannomas are rare and the majority of them are asymptomatic.
- ▶ Preoperative clinical and radiological diagnosis is difficult.
- ▶ Surgical excision, particularly intracapsular dissection, and histopathological examination are the appropriate diagnostic and management approach.
- ▶ Preoperative counselling of these patients is important, especially with regard to the complications of nerve paralysis or paresis following excision.

Postoperative neurological deficit can sometimes prove the origin of the tumour. However, the patient was reassessed 6 months postoperatively, and showed no tongue paraesthesia or paralysis suggestive of nerve damage. In conclusion, schwannomas are rare and difficult to diagnose without intracapsular surgical excision and histopathological examination.

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Contributors SSL had the primary responsibility of obtaining patient records and investigative findings as well as deriving the concept and draft of the case report and was also involved in ensuring accurate presentation of patient information in the case report. VP conducted the surgical procedure and ensured appropriate management and follow-up of this patient. He also made a significant contribution to the written discussion component of the case report as well as conducting a critical review on the overall written piece in ensuring accurate content. CO critically reviewed the histology slides and commented on the histopathological diagnosis of schwannoma. He also made a contribution to the overall written case report in ensuring that it had the necessary pre-requisites for publication.

Competing interests None declared.

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