CASE REPORT

Intracranial hypoglossal nerve schwannoma with extracranial extension

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Introduction

Benign extramedullary tumours of the foramen magnum region are largely schwannomas and meningiomas.1 Tumours originating from the motor part of cranial nerves, such as the VIIth, IXth, Xth and XIIth cranial nerves, are extremely rare. Hypoglossal nerve (cranial nerve XII) schwannomas are such an example, usually arising intracranially and, in exceptional cases, extending into the extracranial compartment.2

We present the clinical and diagnostic radiological findings of a rare case of intracranial hypoglossal nerve schwannoma with extracranial extension.

Case report

A 34-year-old woman presented to the eye centre with gradual bilateral deterioration of visual acuity and mild intermittent headaches over several months. She reported that 7 years previously she had attended another institution for notable wasting of the right side of her tongue. It was unclear what, if any, investigations or diagnostic imaging were performed then. There was no other relevant medical history.

Clinical examination revealed severe wasting of the right side of the tongue with mild right-sided palatal weakness which did not appear to affect swallowing or speech. The woman maintained a visual acuity of 6/6 bilaterally and normal intraocular pressures. The remainder of the neurological examination was unremarkable. Goldman perimetry showed very slight prominence of the blind spot but Ishihara colour vision and Amsler tests were normal.

Diagnostic imaging

CT showed subtle asymmetrical distortion of the fourth ventricle without displacement or radiological evidence of hydrocephalus (Fig. 1(a)). Post-contrast images showed a 2 × 2 cm homogeneously enhancing lesion in the posterior fossa, to the right of and inferolateral to the fourth ventricle, extending to the level of the foramen magnum (Fig. 1(b)). There was no associated surrounding cerebellar oedema. On the bone algorithms, there was erosion and enlargement of the right hypoglossal canal (Fig. 1(c)).

MRI confirmed the presence of a well-defined asymmetrical bilobed extra-axial mass in the right posterior fossa, just above the foramen magnum, extending through the hypoglossal canal. The lesion was isointense relative to white-matter brain parenchyma on T1-, FLAIR and T2-weighted images and demonstrated homogeneous enhancement post-gadolinium (Fig. 2(a) and (b)). There was tonsillar herniation through to the level of C1.

High-signal changes were seen in the atrophic right tongue on both T1-weighted and FLAIR coronal sequences, consistent with neuropathic fatty degeneration (Fig. 2(c)).

Surgery and histopathology

The tumour was subtotally excised through a right retromastoid craniotomy. The firm haemorrhagic core was debulked, leaving the extracranial extent of the tumour untouched because of concern about adherence to the jugular vein and internal carotid artery. Histopathological examination of the
tumour mass confirmed that it was an Antoni type A schwannoma with areas of sclerosis.

Discussion

Intradural extramedullary tumours of the foramen magnum, according to the review by Spinnato et al., account for 2.5% to 10% of all intraspinal masses and for approximately 1% of all brain tumours. Schwannomas and meningiomas represent 20% and 70% of all such tumours, respectively; 65% of these are located in the anterolateral intradural space. Schwannomas most commonly involve the lower cranial nerves or the C1 and C2 roots; most schwannomas arise from the sensory division of the cranial nerves. Motor division involvement or schwannomas that arise at an early age often show evidence of neurofibromatosis.

The hypoglossal nerve is a pure motor nerve that innervates the intrinsic and extrinsic muscles of the tongue. Supranuclear hypoglossal nerve lesions do not result in denervation atrophy of the tongue muscle; this is seen only when the nuclear or peripheral segments of the hypoglossal nerve are involved, as in this case. Lesions involving the cisternal course of cranial nerve XII often affect cranial nerves IX, X and XI because they are in close anatomical proximity to the hypoglossal nerve roots. This is particularly the case if the lesion is of primary or metastatic malignant origin. It is therefore somewhat surprising that our patient experienced isolated XIIth nerve involvement despite the sizeable cisternal extension of the schwannoma.

De Martel et al. first described hypoglossal schwannomas in 1933. They are very rare tumours; only 18 cases have been reported in the literature in the last 10 years. Patients are commonly middle-aged and predominantly female. Hypoglossal schwannomas arise usually intracranially, causing enlargement and erosion of the hypoglossal canal, and rarely extend over the occipital condyle into the extracranial compartment towards the jugular foramen, carotid canal or infratemporal fossa. The extracranial extension of the tumour results in a dumb-bell-shaped appearance of the tumour. Only 7 cases of these dumb-bell-shaped tumours,

![Figure 1](a) Precontrast CT of a 34-year-old woman with a right hypoglossal schwannoma. Note the subtle distortion of the fourth ventricle on the right. (b) Post-contrast axial acquisition CT showing the homogeneous enhancement of the right hypoglossal schwannoma. (c) CT axial acquisition on bony algorithms, showing the enlargement of the right hypoglossal canal as a result of the extension of the hypoglossal schwannoma.
including our case, have been described in the literature over the last 10 years.

The presenting symptoms are often misleading, resulting in a long clinical course between onset of symptoms and definitive diagnosis, by which stage the tumour may have reached a significant size.\textsuperscript{1,3} Neurological deficits include lower cranial nerve palsy with long tract signs due to compression at the cervicomedullary junction. Hypoglossal palsy with ipsilateral hemiatrophy and weakness of the tongue is the most common sign. Patients may develop cerebellar deficits as a result of increased intracranial pressure, further confounding the clinical diagnosis.

High-resolution CT and MRI of the cervicomedullary junction are the imaging investigations of choice and provide information for surgical planning based on the origin of the tumour and its pattern of extension. High-resolution CT on bone algorithms is important in order to demonstrate bone involvement.

Most of the reported cases in the literature have been classified histopathologically as neurinomas, neurofibromas, neurilemmomas or schwannomas.\textsuperscript{3} Schwannoma tumour tissue consists of Antoni A and B type cells. Type A tissue is identified by a compact texture composed of interwoven parallel or palisaded bundles of long spindle cells. The type B areas are distinguished by the loose texture and polymorphism of tumour cells embedded in a fine honeycomb eosinophilic myxomatous matrix. Haemorrhage from adjacent tissue, necrosis, hyalinization and cystic degeneration can occur. These may give rise to the clearly circumscribed and partly cystic heterogeneous signal appearance on MRI. Immunohistochemistry involves the S-100 protein as a diagnostic marker for schwannomas.\textsuperscript{7}

Only 2 cases of malignant degeneration have been reported in the literature to date. As they are largely benign tumours, the mainstay of treatment is complete surgical excision. Complete removal of tumours located in a complex region of the skull base may be difficult.\textsuperscript{4} The surgical approach is determined by the type of tumour extension. Dumbbell-shaped schwannomas have represented a surgical challenge, due to both intra- and extracranial extension.\textsuperscript{3}

\textbf{Figure 2} (a) Post-gadolinium contrast T1-weighted (TE=15, TR=525) axial MRI of the right hypoglossal schwannoma with dumb-bell appearance. (b) Post-gadolinium contrast-enhanced T1-weighted (TE=15, TR=525) coronal acquisition showing the extension of the intracranial hypoglossal schwannoma on the right. (c) T1-weighted (TE=15, TR=525) coronal MRI showing significant neuropathic fatty atrophy of the right side of the tongue.
Conclusion

Our case illustrates several pertinent points. The long, protracted history of presentation and vague symptoms before reaching the definitive diagnosis are typical. The case is also a good illustration of the complementary roles of MRI and CT in evaluation of skull-base lesions for diagnostic and surgical planning purposes. Although there are no pathognomonic radiological signs for the diagnosis of a hypoglossal schwannoma, in our case there was strong radiological evidence that pointed to the final diagnosis.

References