Case Report: Intraneural Intracanalicular Ganglion Cyst of the Hypoglossal Nerve Treated by Extradural Transcondylar Approach

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Abstract

We report a case of an intraneural ganglion cyst of the hypoglossal canal. The patient presented with unilateral hypoglossal nerve palsy, and magnetic resonance imaging showed a small lesion in the hypoglossal canal with no contrast enhancement and high signal on T2-weighted imaging. The lesion was assumed to be a cystic schwannoma of the hypoglossal nerve. Stereotactic irradiation was considered, but in accordance with the patient's wishes, surgical exploration was performed. This revealed that, rather than a schwannoma, the patient had an intraneural ganglion cyst, retrospectively contraindicating irradiation as an option. This case illustrates a very rare location of an intraneural ganglion cyst in the hypoglossal nerve. To our knowledge there are no previous reports of an intraneural ganglion cyst confined to the hypoglossal canal.

Keywords
- intraneural ganglion cyst
- hypoglossal nerve palsy
- hypoglossal canal
- transcondylar approach

Introduction

Most intraneural ganglion cysts present in the peripheral nerves near joints or tendon sheaths.[1] Cranial nerves are rarely affected. A common location of an intraneural ganglion cyst is the peroneal nerve behind the fibular head.[2–4] The pathogenesis of ganglion cysts is still unclear but is likely to be developmental. The cyst can compress and invade the adjacent nerves and become symptomatic.

In the present case a small cyst was confined to the hypoglossal canal, making interpretation of magnetic resonance imaging (MRI) difficult. This report demonstrates the importance of an intraneural ganglion cyst as a differential diagnosis in a patient with hypoglossal nerve palsy.

Case Report

Examination revealed left-sided tongue atrophy and slurred speech. MRI obtained 13 months prior to surgery was initially interpreted as negative, but retrospectively it showed a nearly imperceptible T2 hyperintense 3-mm lesion related to the hypoglossal canal (►**Fig. 1**). Eight months later, MRI was repeated due to the progression of symptoms. This MRI revealed a 7-mm T2 lesion with gadolinium enhancement into the left hypoglossal canal (►**Fig. 2**).

A standard transcondylar extradural approach without transposition of the vertebral artery allowed evacuation of the cyst, and 360-degree neurolysis of the hypoglossal nerve was performed to disconnect the cyst from the C0–C1 joint.

The patient had no complications and was discharged on day 2. No recurrence was shown on MR images obtained 7 months postoperatively. The slurred speech resolved after a few weeks.

Discussion

We know isolated hypoglossal nerve palsy is rare and most commonly associated with other cranial nerve involvement.[5–8] Isolated nerve palsy is reported to be caused by hypoglossal nerve schwannomas,[6,7,9–11] dural arteriovenous fistulas, enlarged emissary veins of the hypoglossal canal, aneurysms of the stump of a persistent hypoglossal artery, internal carotid and vertebral artery dissections, metastatic

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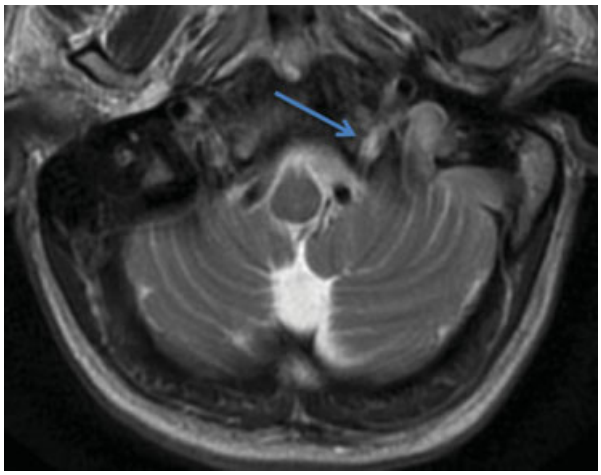

Fig. 1 Initial axial T2-weighted magnetic resonance image showing hyperintense legion related to the hypoglossal canal (indicated by arrow).

lesions to the skull base, arachnoid cysts, occipital condyle fractures, after a neck surgery, or with no apparent cause.[6,7]

Many theories have been proposed to explain the pathogenesis of juxtafacet intraneural ganglion cysts. Although the theories suggest association with a joint,[3] myxoid degeneration and cyst formation in the synovial tissue,[12,13] and prior traumatic injury[14,15] all may play a role, the origin and pathogenesis of these cystic tumors remain unknown.

To our knowledge, five cases present a patient with an isolated hypoglossal palsy due to a cranial nerve ganglion cyst.[5,7,16,17] Mujic et al[5] and Elhammady et al[6] classified their cases as atlantooccipital joint synovial cysts. Mujic et al called it synovial cyst because of the presence of fibrous connective tissue with myxoid change, and no associated neural tissue was present. Elhammady et al reported a case of an atlantooccipital juxtafacet cyst that can be classified as a synovial cyst, lined with synovial cells and containing clear or xanthochromic fluid, or as a ganglion cyst, which does not have a synovial lining and contains gelatinous content. The other three cases reported by Baldauf et al,[16] Nonaka et al,[17] and Gambhir et al[7] describe an intraneural ganglion cyst due to

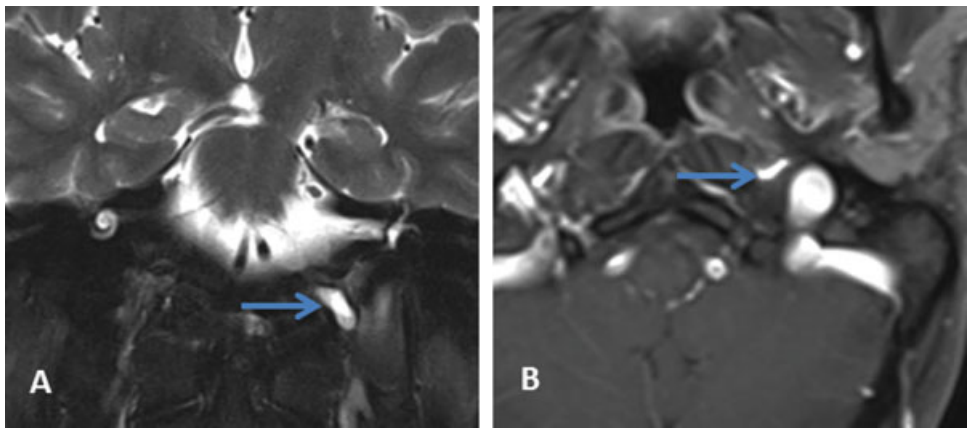

Fig. 2 (A) Preoperative coronal T2-weighted magnetic resonance image showing the hyperintense legion (indicated by the arrow). (B) Axial gadolinium-enhanced T1-weighted image showing the cystic lesion (indicated by the arrow).

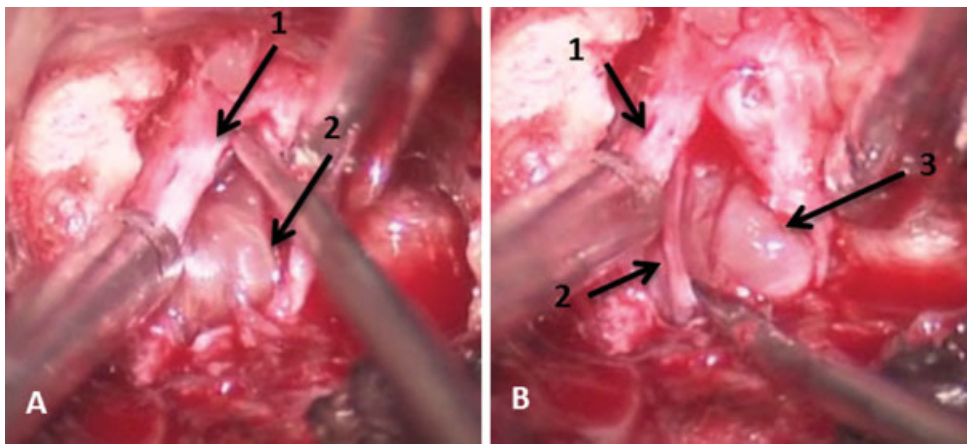

Fig. 3 Intraoperative images. Surgical exploration revealed an intraneural ganglion cyst, and the transcondylar approach allowed complete evacuation of the cyst content. (A) Dura (arrow 1) and cyst (arrow 2), below which is concealed the hypoglossal nerve. (B) Dura (arrow 1), hypoglossal nerve (arrow 2), and gel-like cyst content (arrow 3).

the presence of neural tissue affecting the hypoglossal nerve. The presented case differs by being confined to the hypoglossal canal.

The present lesion was interpreted as a possible schwannoma, and stereotactic radiosurgery was offered based on this and the small size of the lesion. Fortunately, the patient preferred surgery. The lesion was extradural and in the left hypoglossal canal. Given the location of the lesion, the extradural transcondylar approach was performed. Tumors at the craniovertebral junction are difficult to remove because of their location and complex anatomical relations. The transcondylar approach is a versatile approach to this area and allows access to a variety of intra- and extradural tumors. In this case, the transcondylar approach allowed complete evacuation of the cyst content and microneurolysis (►**Fig. 3**).

Based on intraoperative findings, the lesion was classified as an intraneural ganglion cyst of the hypoglossal nerve. The treatment of a cyst is surgical excision.

Conclusion

The extreme rarity of an intraneural ganglion cyst in the hypoglossal canal makes it challenging to recognize. Due to the favorable prognosis for recovery of muscle function after surgical decompression, it is important to consider this diagnosis in cases of intracranial hypoglossal palsy.

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