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Extradural dumbbell schwannoma of the hypoglossal nerve: a case report with review of the literature

Lu-Ting Kuo, MD, PhD[a], Abel Po-Hao Huang, MD[a], Kuan-Ting Kuo, MD[b], Ham-Min Tseng, MD[a,*]

[a]*Division of Neurosurgery, Department of Surgery, National Taiwan University Hospital, Taipei, Taiwan 100*
[b]*Department of Pathology, National Taiwan University Hospital, Taipei, Taiwan*

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Abstract

Background: Dumbbell-shaped schwannomas of the hypoglossal nerve are very rare. This report concerns a case with an extradural, dumbbell-shaped hypoglossal schwannoma extending both intra- and extracranially.

Case Description: A 25-year-old woman presented with a right hypoglossal palsy. Imaging revealed a dumbbell-shaped tumor with considerable compression and medial displacement of the medulla oblongata, diagnosed as a hypoglossal schwannoma. The tumor mass extended extracranially to the parapharyngeal space through the enlarged hypoglossal canal. The tumor was partially excised by a right far-lateral suboccipital approach and the tumor was found to be predominantly extradural with minimal intradural extension. A histopathologic diagnosis of schwannoma was made.

Conclusions: This case emphasizes the importance of recognizing this extradural variant of schwannoma. Staged extracranial and intracranial approaches to these tumors may be necessary.

Keywords: Dumbbell-shaped tumor; Extradural; Hypoglossal canal; Hypoglossal nerve; Schwannoma

1. Introduction

Schwannomas of the hypoglossal nerve are very rare. These tumors usually originate intracranially but can also extend extracranially through the hypoglossal canal into the parapharyngeal space. Only 12 cases of dumbbell-shaped hypoglossal schwannomas have previously been reported, most of which had both extradural and intradural components [18]. The extradural, dumbbell-shaped hypoglossal schwannoma we report here has never been emphasized.

2. Case report

A 25-year-old woman was admitted to the hospital in December 2004 with a history of tongue deviation to the right for more than 2 years. Neurologic examination showed a right hypoglossal palsy with hemiatrophy of the tongue without deficits of the other cranial nerves. Neither cerebellar nor long-tract signs, such as motor or sensory disturbances, were identified. There were no manifestations of neurofibromatosis. A laboratory hemogram, blood chemistry, and urinalysis were normal. Subsequent MRI demonstrated a solitary extra-axial tumor in the right cerebellopontine angle (Fig. 1). The tumor showed inhomogeneous enhancement with Gd-DTPA. It caused considerable compression and medial displacement of the medulla oblongata without compressing the fourth ventricle. The tumor mass extended extracranially to the level of C-2 in the parapharyngeal space through the enlarged hypoglossal canal. An angiography disclosed neither a feeding artery nor a tumor stain; however, the right sigmoid sinus was not opacified and the right internal carotid artery was slightly displaced medially.

The patient underwent surgery on December 13, 2004. She was placed in a three-quarters prone position with her

Abbreviations: CT, computed tomography; Gd-DTPA, gadolinium-diethylene-triamine pentaacetic acid; MRI, magnetic resonance imaging.

* Corresponding author. Tel.: +886 2 2312 3456.
E-mail address: thm1995@ha.mc.ntu.edu.tw (H.-M. Tseng).

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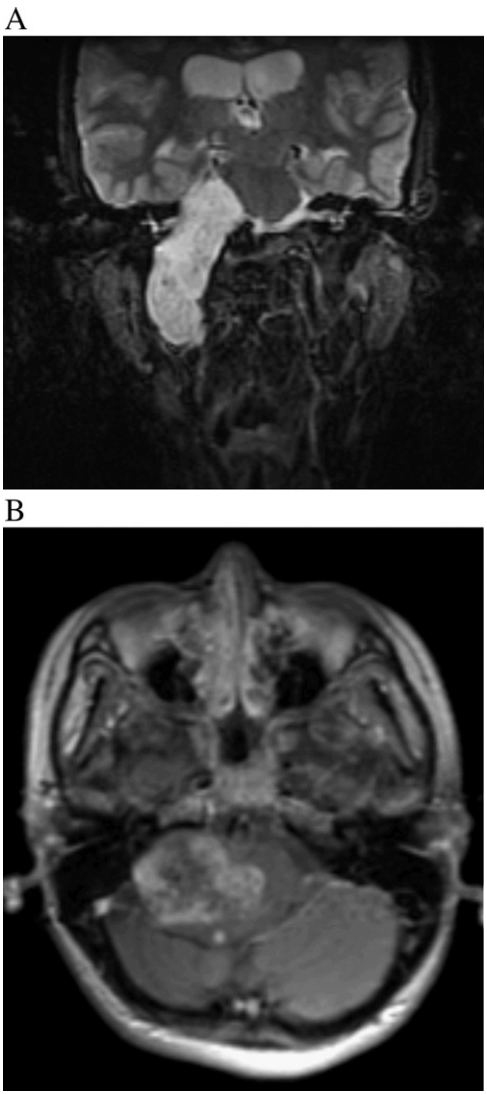

Fig. 1. Preoperative T2-weighted coronal (A) and contrast-enhanced T1-weighted axial (B) MRI scans show a tumor in the right cerebellopontine angle, causing medial displacement of the medulla, with dumbbell-shaped extracranial extension to the parapharyngeal space through the enlarged hypoglossal canal.

head rotated to the right by 45°, and a right far-lateral suboccipital approach was performed. Initially, an intradural approach was used; however, during the operation, most of the tumor was found to be extradural, with only a tiny portion extending intradurally into the premedullary cistern (Fig. 2). The other lower cranial nerves were identified and were free of any compression. The hypoglossal nerve was dissected and identified as the origin of the tumor. It was a yellowish, soft-elastic, and well-circumscribed tumor that compressed the medulla. The intracranial portion of the tumor was totally removed. The intracranial hypoglossal rootlets were normal microscopically. Pathologic inspection of the resected tumor identified it as a schwannoma, being composed of loose, palisading spindle cells with wavy nuclei against a hyaline or myxomatous background, with hyalinizing vessels, calcification, and ossification (Fig. 3). Mitotic figures were not increased and nuclear atypia was limited. These tumor cells were immunoreactive when stained with polyclonal antibodies against S-100 protein.

After the operation, the patient experienced deficits of cranial nerves IX through XII on the ipsilateral side. She was discharged 23 days after the operation. A postoperative MRI showed that the intracranial tumor mass had been removed completely (Fig. 4). A follow-up examination 6 months later showed full recovery of the cranial nerves except for a complete deficit of the right hypoglossal nerve.

3. Discussion

Schwannomas are benign, slow-growing tumors of the myelin-producing Schwann's cells, presenting as acoustic

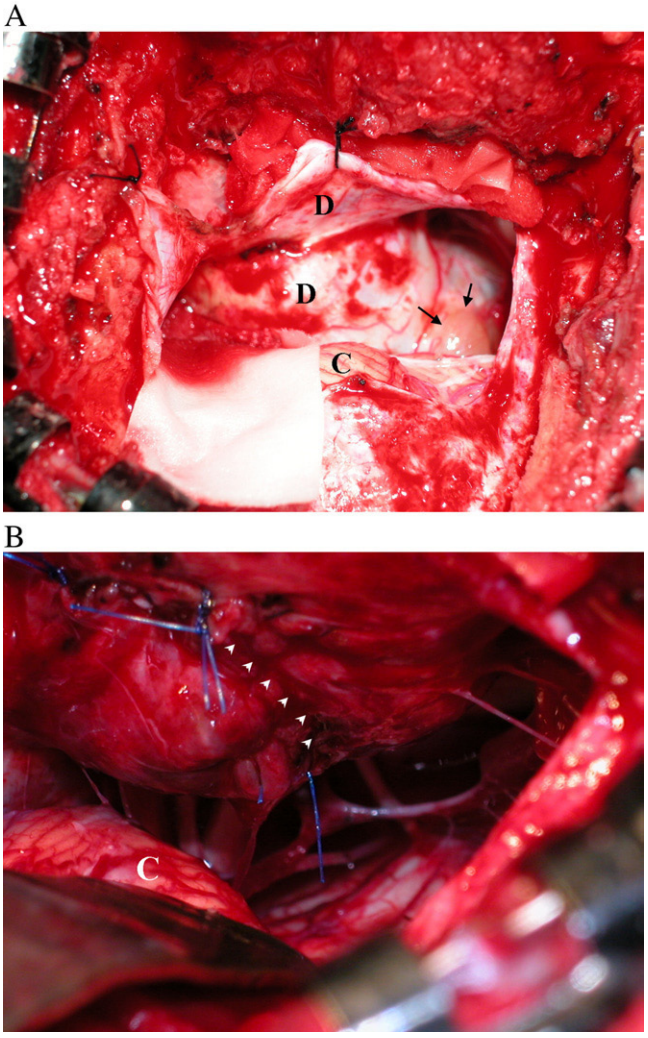

Fig. 2. A: Intraoperative photography shows exposure of the dumbbell-shaped tumor which was located extradurally with a tiny portion of intradural extension (arrow). B: After resection of the intracranial part of the tumor, dural closure was achieved (arrowheads). Lower cranial nerves were identified. C indicates cerebellum; D, dura.