Intracranial Hypoglossal Neurinoma without Preoperative Hypoglossal Nerve Paresis
—Case Report—

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Abstract

A 46-year-old female presented with an intracranial hypoglossal neurinoma manifesting only as spasticity in the lower extremities without hypoglossal nerve paresis. Magnetic resonance imaging greatly aided in the early detection of the tumor with this atypical presentation. Unilateral suboccipital craniotomy with resection of the occipital condyle allowed us to approach the tumor in front of the medulla from an inferolateral direction and to remove it successfully. We emphasize the need to pack dead space with fatty tissue to prevent cerebrospinal fluid leakage.

Key words: hypoglossal nerve, hypoglossal nerve palsy, magnetic resonance imaging, neurinoma

Introduction

Neurinomas of the hypoglossal nerve are rare. The significance of preoperative evidence of ipsilateral hemiatrophy and weakness of the tongue has been emphasized in the diagnosis. We describe a patient with an intracranial hypoglossal neurinoma with only spasticity in the lower extremities and no hypoglossal nerve paresis.

Case Report

A 46-year-old female was admitted to our hospital with spasticity of both lower extremities which had begun 2 years prior to admission, but no complaints of headache or nuchal pain. She was alert and well oriented. Neurological examination found no cranial nerve abnormalities, no weakness or atrophy of the tongue, and no motor or sensory abnormalities. Deep tendon reflexes were increased in both lower extremities. Other examinations including cerebellar testing were within normal limits. There were no stigmata or family history of von Recklinghausen’s disease.

Radiography of the skull and whole spine were unremarkable. Precontrast axial computed tomography (CT) disclosed an isodense mass in the lateral medullary cistern (Fig. 1). Anteroposterior skull tomography and CT showed no change of the hypoglossal canal, jugular foramen, or occipital condyle. T1-weighted sagittal magnetic resonance (MR) imaging with gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA) demonstrated a well-demarcated multi-nodular mass extending from the cerebellopontine cistern to the lateral medullary cistern (Fig. 2 left). T1-weighted axial and coronal MR imaging showed the mass compressing the medulla and pons, but not encroaching on the jugular foramen or the hypoglossal canal, or extending extracranially (Fig. 2 center, right). Vertebral angiography showed no displacement of the major vessels or pathological vascularization, and the venous phase showed no occlusion of the jugular sinus. Right carotid angiography found no abnormality.
She was placed in the semi-prone position and her head turned to the contralateral side. A hockey-stick skin incision was made from the tip of the mastoid process up to the superior nuchal line, curved toward the midline, and straight down to the spinous process of C-3. In addition to the standard right suboccipital craniotomy, the posteromedial one-third of the occipital condyle and the posterior portion of the mastoid process were removed. The bone covering the sigmoid sinus was resected with a high-speed drill, and the entire length of the sigmoid sinus down as far as the jugular bulb was exposed. These procedures allowed us to approach the tumor in front of the medulla from an inferolateral direction. Dural incision exposed the tumor extending inferiorly beneath the right cerebellar hemisphere. The tumor was easily separated from the ninth, tenth, and eleventh cranial nerves. After intracapsular removal of the tumor, we found that the capsule had adhered to the sectioned and free distal stump of the nerve at the entrance of the hypoglossal canal. Therefore, we concluded that the origin of the tumor was the hypoglossal nerve. The wound was closed after applying lyophilized dura and fibrin glue to the dural defect. Histological examination of the specimen showed evidence of typical neurinoma (Fig. 3).

After the operation, persistent tongue deviation to the right was observed. Difficulty in swallowing, due to dysfunction of the ninth and tenth cranial nerves, was slight and transient. Postoperative MR imaging showed no residual tumor. A postoperative leak of cerebrospinal fluid into the subcutaneous space was successfully treated with packing of abdominal fatty tissue, and she was discharged.

Discussion

Forty-four patients with intracranial hypoglossal neurinoma have been described previously. Including the present patient, the median age at diagnosis was 43.0 years (range 16–64 yrs), and 33 of the 45
patients were females. The left hypoglossal nerve was involved slightly more frequently than the right: 24 on the left, 20 on the right, and one unspecified. Hemiatrophy and weakness of the tongue were noted by the first hospitalization in every patient except four. This is the most common and characteristic finding in hypoglossal tumors. However, many patients did not notice or complain of hypoglossal deficits, because dysfunction of one hypoglossal nerve does not cause disruption of speech or dysphagia. The tumor irritates the nerve root and meninges, and often causes suboccipital and nuchal pain. Signs and symptoms of increased intracranial pressure such as headache, vomiting, and papilledema are often present, but are less common in recent reports. Cerebellar signs, motor disturbance, and sensory disturbance are also frequently present. Our patient presented with spasticity in both lower extremities and no evidence of twelfth cranial nerve dysfunction. Such atypical symptoms made the diagnosis difficult.

The importance of demonstrating an enlarged hypoglossal canal in the diagnosis of intracranial hypoglossal neurinoma has been emphasized. Kuramitsu et al. suggested that a difference of more than 2 mm in diameter between the two hypoglossal canals was significant. In our patient, neither skull tomography nor CT revealed an enlarged hypoglossal canal, and the origin of the tumor could not be determined preoperatively.

MR imaging demonstrated the presence of the tumor, and allowed us to make an early diagnosis in spite of the atypical signs and symptoms. MR imaging revealed the location and extension of the tumor accurately and was more effective than CT. The tumor did not encroach on the hypoglossal canal in the MR image. We consider that the tumor originated from the rootlets of the hypoglossal nerve and grew mainly between the medulla and the hypoglossal canal, which may explain the atypical symptoms and absence of an enlarged hypoglossal canal.

Unilateral suboccipital craniectomy has been used for removal of the tumor. However, the lateral portion of the foramen magnum, the posterosmedial portion of occipital condyle, and the jugular tubercle obstruct the operative field in this approach. Seeger recommended removal of these structures in addition to the standard unilateral suboccipital craniectomy, using the term “dorsolateral approach with resection of the condylus occipitalis and tuberculum jugulare.” This useful approach was improved by Hakuba et al. as the “transcondylar approach.” Removal of occipital condyle should be limited to within the posterior one-third to prevent postoperative instability. We chose this approach for the present case. Based on our experience with this patient, we emphasize the importance of packing of the dead space with fatty tissue after bone resection to prevent postoperative cerebrospinal fluid leakage.

Early diagnosis, microsurgical techniques, and improved supportive care may have contributed to the improvement in the operative results. No deaths have occurred among recently reported patients. Careful use of microsurgical techniques obviates the need for tracheotomy. Intracapsular resection is recommended if the tumor adheres to the brainstem firmly, since MR imaging can detect regrowth of the tumor accurately.

References

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