Large Cystic Hypoglossal Schwannoma with Fluid-Fluid Level: A Case Report

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ABSTRACT

Hypoglossal schwannomas are rare skull base tumors. Furthermore, cystic hypoglossal schwannomas are extremely uncommon. We report the first case of a large cystic hypoglossal schwannoma with a fluid-fluid level. A 36-year-old woman presented with increased intracranial pressure and cerebellar signs without hypoglossal nerve palsy. Magnetic resonance imaging showed a predominantly cystic mass with a fluid-fluid level in the foramen magnum region extending into the hypoglossal canal. The intracranial tumor was largely removed via a midline suboccipital subtonsillar approach, leaving only a tiny residue in the hypoglossal canal. Histology confirmed a schwannoma with relative hypervascularity. Twenty months later, the tumor recurred and presented as a multicystic dumbbell-shaped lesion, extending intra- and extracranially through the enlarged hypoglossal canal. A complete resection of the intracranial and intracanalicular parts of the tumor was achieved with a small extracranial remnant treated by radiosurgery. Histology revealed a focal increased Ki67 proliferative index. In this report, we discuss the possible reasons for the absence of hypoglossal nerve palsy and the potential mechanism of the formation of the fluid-fluid level, and we consider the treatment of this lesion.

KEYWORDS: Hypoglossal schwannoma, cystic, fluid-fluid level, schwannoma

Schwannomas are benign, slow-growing neoplasms of the myelin-producing Schwann cells in the peripheral sensorimotor nervous system. In more than 90% of cases, these present as vestibular schwannomas.¹ Schwannomas that arise from the hypoglossal nerve, a pure motor nerve, are very rare, accounting for only 5% of all nonacoustic intracranial schwannomas.² To date, around 100 cases of hypoglossal schwannoma have been reported in the English literature.³

Hypoglossal schwannoma usually originates intracranially but can also extend extracranially through the hypoglossal canal in a so-called “dumbbell” fashion or can arise purely from the extracranial portion of the

XII nerve. According to Kaye et al’s⁴ classification of the jugular foramen neurinomas, the hypoglossal schwannomas documented in the literature are divided into three types: type A, intracranial in 31.5%; type B, dumbbell-shaped or extra- and intracranial in 50%; type C, extracranial in 18.5%.⁵

Schwannomas are frequently solid masses, and cystic patterns are uncommon. A fluid-fluid level has been observed in a few intracranial cystic schwannomas on magnetic resonance image (MRI), such as schwannomas that originate from the V, VIII, and IX nerves.^{6–8} Here we describe a unique case of a monocystic hypoglossal schwannoma with fluid-fluid level. To our

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Skull Base 2010;20:193–197. Copyright © 2010 by Thieme

Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

Received: April 15, 2009. Accepted: September 13, 2009. Published online: January 12, 2010.

DOI: <http://dx.doi.org/10.1055/s-0029-1246219>. ISSN 1531-5010.

knowledge, this type of presentation of a hypoglossal schwannoma has not been reported previously.

CASE REPORT

A 36-year-old woman presented with intermittent right occipital headache that had lasted for about a year and a half. For the 2 weeks prior to admission, she had persistent generalized headache accompanied by vomiting, vertigo, and an unbalanced gait. A computed tomography (CT) scan of the head obtained 6 months prior to admission showed no apparent abnormality. Neurological examination demonstrated horizontal nystagmus, ataxia, and bilateral papilledema without deficits of the lower cranial nerves. Long tract signs, such as sensory and motor disturbances, were not observed. There were no manifestations of neurofibromatosis.

An MRI showed a predominantly cystic mass with a fluid-fluid level in the foramen magnum region extending into the right hypoglossal canal, which measured $4.7 \times 3.8 \times 2.9$ cm. The right cerebellum, the brain stem, and the fourth ventricle were markedly compressed. The subnatant fluid was hyperintense on T1-weighted image when compared with the supernatant fluid. However, the fluid-fluid level was not visible on T2-weighted image. A contrast-enhanced MRI showed diffuse enhancement on the solid portion of the tumor and a rim enhancement on the cystic capsule (Fig. 1A).

Surgery was performed via a midline suboccipital subtonsillar approach. The tumor had a smooth and dark green surface. The dissection from the surrounding cerebellum, brain stem, and accessory nerve was performed easily as a good dissection plane was present. Xanthomatic fluid and brownish old unclotted blood

were aspirated from the cyst. The intracranial portion of the neoplasm was totally removed. The hypoglossal nerve was dissected and identified as the origin of the tumor. The nerve trunk in the hypoglossal canal was completely engulfed by the tumor. A tiny tumor residue was left attached to the trunk to preserve its integrity. The solid portion of the tumor was well vascularized.

Histopathology revealed a relatively hypervascular schwannoma with dilated thin-walled vessels (Fig. 2). Immunohistochemical examination showed positive for S-100 protein. Only 4% of cells were positive for Ki67.

The patient did not receive postoperative radiotherapy and had a slight deviation of the tongue, which improved noticeably 6 months after surgery. Twenty months later, the patient was referred with a 2-month history of mild hoarseness and hypoglossal nerve palsy. Neurological examination revealed a right deviation, atrophy, and fasciculation of the tongue. A three-dimensional CT of the skull base disclosed destruction of the hypoglossal canal (Fig. 3). The contrast-enhanced MRI showed a rim-enhanced multicystic dumbbell-shaped mass extending intra- and extracranially through the enlarged hypoglossal canal (Fig. 1B).

The patient underwent a second surgery via a right suboccipital retrosigmoid approach. The intracranial portion of the tumor was totally removed; its content consisted of blood breakdown products and necrotic materials. The intracranial part was then resected apart from a small extracranial remnant (Fig. 4). Histopathology confirmed that the mass was a recurrent cystic schwannoma with hypervascularity. The focal Ki67 proliferative index was ~15%. The postoperative course was uneventful. The patient underwent gamma knife surgery to treat the extracranial residual tumor.

Figure 1 (A) The first preoperative contrast-enhanced magnetic resonance imaging (MRI) revealed a predominantly cystic mass with a fluid-fluid level (black arrowhead) in foramen magnum region extending into the right hypoglossal canal (white arrow). (B) The second preoperative contrast-enhanced MRI showed a multicystic dumbbell-shaped mass extending intra- and extracranially through enlarged hypoglossal canal.

Figure 2 Photomicrograph showing spindle-shaped tumor cells and relative hypervascularity with thin-walled dilated vessels (hematoxylin and eosin, $\times 200$).

Figure 3 Three-dimensional computed tomography of the skull base revealed enlargement of the hypoglossal canal and the jugular foramen and the erosion of jugular tubercle.

DISCUSSION

Hypoglossal schwannomas are rare skull base tumors that usually cause ipsilateral hypoglossal nerve palsy. Hypoglossal nerve palsy has particular diagnostic value, as $\sim 85.7\%$ of patients with hypoglossal schwannomas present with this symptom.⁹ Hypoglossal nerve palsy antedates other symptoms from 5 months to 10 years, on an average of 3.1 years.¹⁰ However, several reports have described hypoglossal schwannomas without hypoglossal nerve palsy. In these instances, the neoplasm arose from the intracranial rootlets of the XII nerve.¹¹ Our patient presented without XII nerve deficit, and the tumor originated from the trunk. The tumor extended intracranially away from the hypoglossal nerve itself with only

a small portion located in the canal. This discrepancy may depend not only on the anatomic location of the tumor's origin and the direction of tumor's extension but other factors may also be at play. The extremely slow growth of benign schwannoma may allow for a higher degree of tolerance of the nerve.¹² The abundant tumor blood flow supports not only the schwannoma itself but also helps preserve hypoglossal nerve integrity.¹³ Finally, the formation and expansion of a cyst lead to rapid neurological deterioration, which makes the patient seek medical assistance prior to the onset of hypoglossal nerve palsy.

Fluid-fluid levels in intracranial schwannomas are exceptionally rare.⁶ The mechanism of the fluid-fluid

Figure 4 (A) Intraoperative photograph showed exposure of the cystic tumor. (B) After resection of the tumor, the right vertebral artery (white arrowhead) was visible and Gelfoam (the 3rd Pharmaceutical Factory of Nanjing, Nanjing, China) was used to fill in the enlarged hypoglossal canal (white arrow).

level formation is not completely clear yet, but formation is probably due to the differing densities of the cyst constituents. Fluid levels in cystic schwannomas can be caused by collection of liquefactive necrotic materials or by hemorrhage from hypervascular schwannomas.⁸ Most often, this blood does not clot, so that some of the fluid is unclotted blood, which may create a level against serous (interstitial) fluid.¹⁴ Hypervascular schwannomas may have a higher tendency to result in a spontaneous intratumoral hemorrhage. Takahashi et al⁵ reported that a hypervascular hypoglossal schwannoma with hyaline degeneration of vessels had extensive intratumoral hemorrhage. In our case, we think that the hemorrhage resulted in the formation of cyst and the fluid-fluid level. This is confirmed by the direct intraoperative evidence that the brownish old unclotted blood settled at the bottom due to its higher density. The histopathology revealed hypervascularity with thin-walled dilated vessels. The cystic enlargement may also have been augmented by fluid accumulation due to the gradual lysis of red blood cells and the direct osmotic effect of hypertonic fluid.

As the tumor described here was type A, we resected the tumor via a midline suboccipital subtonsillar approach. The approach was first described by Tatagiba et al¹⁵ to treat a hypoglossal schwannoma. It permits a straight primary intradural view to the hypoglossal canal without the necessity of a condylar resection. If surgery is to be undertaken, the objectives of modern skull base surgery should be considered, including anatomic and functional cranial nerve preservation, protection of the great vessels, and microscopic gross total tumor removal.¹⁶ In our case, we were in a dilemma of whether to perform a radical resection of the neoplasm with XII nerve sacrifice or whether to leave a small residue within the hypoglossal canal. We choose to preserve the nerve after the following considerations: (1) preoperative absence of hypoglossal nerve palsy, (2) tight adherence to the trunk of the origin nerve, (3) abundant blood supply in the intracanalicular tumor, (4) and slow-growing characteristics of the benign schwannoma. Unfortunately, rapid regrowth of the residual tumor led to complete palsy of the hypoglossal nerve and reoperation was necessary. It may be a better choice in cystic hypoglossal schwannomas to totally excise the tumor including the sacrifice of the XII nerve, as total excision may reduce the risk of tumor regrowth, thus avoiding reoperation. Furthermore, the dysfunction of a unilateral hypoglossal nerve does not cause disruption of speech or dysphasia.¹⁷

Little is known about the long-term natural history of residual hypoglossal schwannoma, but the growth rate of schwannoma is generally believed to be slow. In our case, however, the tumor recurred markedly 20 months after surgery. Interestingly, the recurrence was also cystic. Furthermore, the K₆₇

proliferative index that reflects the tumor growth potential increased from 4 to 15%. This strongly suggests that the residual cystic hypoglossal schwannoma had a tendency of accelerated regrowth. Kameyama et al¹⁸ noted that the residual cystic acoustic neuroma was associated with rapid regrowth and required reoperation, as compared with the residual acoustic neuroma without cyst formation. We choose radiosurgery to manage the extracranial residual tumor because of the extracranial complex neurovascular structures that would make a surgical approach more hazardous. However, further long-term follow-up will be needed to establish the role of radiosurgery in the treatment of residual cystic hypoglossal schwannomas.

ACKNOWLEDGMENTS

I thank Doctor Ivar Mendez for grammatical revision of the manuscript.

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