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Case Report

Dumbbell-shaped solid-cystic hypoglossal schwannoma: An unusual case report☆☆

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ABSTRACT

Hypoglossal Schwannomas are extremely rare benign slow-growing neoplasms, which originate from the 12th cranial nerve. To date, and to the best of our knowledge, only 40 cases of dumbbell-shaped Hypoglossal Swchannomas have been published in the world literature. We report our experience with a 66 years old male patient, who was diagnosed with a solido-cystic lesion at the right cerebello-pontine angle arising from XIIth cranial nerve. He was treated with surgery via midline suboccipital approach which led to sub-total removal of the tumor and improvement of the symptoms within 3 months. This case highlights the importance of an accurate suspicion diagnosis of hypoglossal schwannoma as well as the treatment options including surgery and radiosurgery.

Introduction

Hypoglossal Schwannomas (HS) are extremely rare benign slow-growing neoplasms, which originate from the myelin-producing cells of the 12th cranial nerve. They account for only less than 5% of all non-vestibular neurinomas. These tumors can be completely intracranial, intracranial/extracranial, or completely extracranial. Intracranial HS are typically challenging to distinguish from other cerebello-pontine lesions because of their low frequency. To date, and to the best of our knowledge, only 40 cases of dumbbell-shaped HS have been published in the world literature. Due to the rarity of this tumor, mainly case reports and small case series have been reported. Here, we report our experience with a patient presenting intracranial/extracranial HS operated in our department. Based on recent reports, we describe the clinical and radiological features of HS, to allow an early diagnostic suspicion and to devise the optimal therapeutic strategy.

Case presentation

We report a case of a 66 years old man with history of hypertension and hearing loss, presented 1 month prior to

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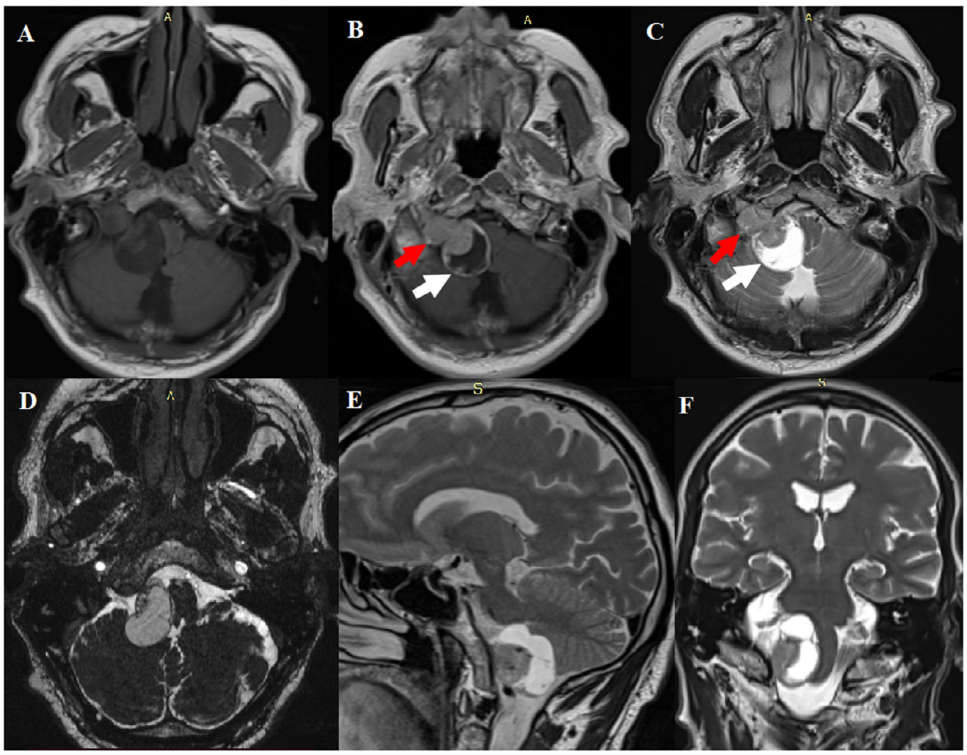

Fig. 1 – Preoperative Brain Magnetic Resonance Imaging (MRI) axial T1-WI (A), axial T1-WI with gadolinium (B), axial T2-WI (C), axial Constructive Interference in Steady State (CISS) sequence (D), sagittal T2-WI (E) and coronal T2-WI (F) showing a large expanding mass in the right cerebellopontine angle with a cystic component (white arrow). The tumour extends extracranially over the occipital condyle area with marked compression of the cervico-medullary junction (F) and through the enlarged hypoglossal canal with dumbbell-shaped extension to the parapharyngeal space (red arrow).

admission to our department of neurosurgery for occipital headache, vertigo and dysphagia to solids. On physical examination, we noted tongue deviation to the right side when pushed out with fasciculation and fissuring in the right side of tongue due to atrophy. The neurological exam showed also a palsy of the right glosso-pharyngeal nerve with static and kinetic cerebellar syndrome. Rest of the neurological examination was normal. A computed tomography scan showed an extra axial solido-cystic mass of the posterior fossa compressing the cerebellar hemisphere and the brainstem. A brain magnetic resonance imaging (MRI) with contrast is performed revealing an extra axial lesion at the right cerebello-pontine angle region. This lesion appeared as a double component mass (nodular and cystic) with hypointense portion and hyperintense portion on T1-weighted images and a hyperintense portion and an isointense portion on T2-weighted images. T1-weighted gadolinium-enhanced MR imaging revealed an enhancement in the nodular portion (Fig. 1). This mass was arising from XIIth cranial nerve with extension to the right internal carotid artery space. The lesion was abutting right IX and Xth cranial nerve and compressing the brainstem. The patient underwent an operation for tumor removal via midline suboccipital approach. Subtotal removal of the lesion was performed and the cystic cavity was drained. Histopathological examination revealed the characteristic features of a schwannoma. Correlating the signs, symptoms and imaging, the diagnosis of a hypoglossal schwannoma was established. The symptoms showed marked improvement following the surgery. The patient was discharged without any further treatment. Six month Follow-up imaging showed complete resection of the intracranial part of the mass. The patient was referred to oncology department for stereotactic radiosurgery treatment (Table 1).

Discussion

Schwannomas are rare benign neoplasms, originating from myelin-producing Schwann's cells, with predilection for sensory nerves. They represent less than 1% of the neoplasms seen in the head and neck region. The most commun is vestibular schwannoma accounting more than 90% of cranial cases. Among all non vestibular schwannomas, hypoglossal schwannomas (HS) is found in only 5% of cases [1]. These pure motor nerve-associated schwannomas are not commun. Currently, and to the best of our knowledge, there are only 98 cases of HS have been published in the world literature.

Schwannomas are generally detected in middle-aged patients with mean age of 44.6 years and predominantly in women (64%) [1]. Our review of the 40 cases showed that our patient is the oldest male case in the literature with 66 years-old. The most common symptoms are classically, tongue deviation or speech disturbance (38%), headaches (33%) followed

by dysphagia (30.7%) and dizziness (23.1%) [2]. Signs of cerebellar or brainstem compression are also reported. Hypoglossal nerve dysfunction was detected in more than 80% of HS cases with unilateral tongue atrophy on the side of the tumor [1].

On neuroimaging, MRI confirmed the exact location of the HS: Intradural (Type A), transdural/dumbbell shaped (Type B) or extra-dural (Type C) [3]. The MRI appearance of these lesions are usually homogeneous and low to intermediate in signal intensity on T1WI, intermediate to high in signal intensity on T2WI. Zones of cystic degeneration, hemorrhage or necrosis are also reported [4]. In our case the mass was solid-cystic. Only 6 cases of solid-cystic HS have been described in the world literature.

Surgical excision is the preferred option for HS. It was performed in 93% of reported cases [5]. Larger symptomatic tumors at the cerebello-pontine angle must be resected. Retromastoid retrosigmoid suboccipital craniotomy is the preferred route. A Midline suboccipital craniotomy can also be used in case of caudal extension through the foramen magnum. A gross total resection was possible in only 41% of intradural cases [6]. Stereotactic radiosurgery tend to be reserved for small tumors or patients with hight risk for surgery. Some authors report its efficacity in tumor growth control with low morbidity [7]. Therefore, for larger tumors, sub-total resection followed by radiosurgery can be the best management strategy.

Late complications was noted in 23% in world literature [8]. Usually, there is no improvement in hypoglossal function. Recurrences was reported in some studies with a median time of 16.5 months requiring repeat surgeries [1].

Conclusion

Hypoglossal neurinomas are a rare entity, in which an accurate suspicion diagnosis must be established regarding their clinicoradiological profile. Surgery remains the main treatment for this benign tumor, allowing complete cure in most cases. However, many factors can hinder gross total resection. Radiosurgery offers now more alternatives for management of these tumors.

Authors' contributions

All authors contributed equally to the manuscript and read and approved the final version of the manuscript.

Patient consent

The patient and his family consented to participate and publish their clinical data.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2021.12.044.

REFERENCES

[1] Bindal S, El Ahmadieh TY, Plitt A, Aoun SG, Neeley OJ, El Tecle NE, et al. Hypoglossal schwannomas: a systematic review of the literature. J. Clin. Neurosci. 2019;62:162–73. doi:10.1016/j.jocn.2018.11.037.
[2] Nonaka Y, Grossi PM, Bulsara KR, Taniguchi RM, Friedman AH, Fukushima T. Microsurgical management of hypoglossal schwannomas over 3 decades: a modified grading scale to guide surgical approach. Oper. Neurosurg. 2011;69:ons121–40. doi:10.1227/NEU.0b013e31822a547b.
[3] De Foer B, Hermans R, Fossion E, Sciot R, Baert AL. Hypoglossal schwannoma. Ann. Otol. Rhinol. Laryngol. 1995;104(6):490–2 no. doi:10.1177/000348949510400614.
[4] Ohba S, Inaba M, Kinoshita Y, Nakagawa T, Shimizu K, Murakami H. Hypoglossal nerve schwannoma with intratumoral hemorrhage. Brain Tumor Pathol 2012;29(1):43–6 no. doi:10.1007/s10014-011-0060-2.
[5] Shimansky VN, Shevchenko KV, Poshataev VK, Tanyashin SV, Abdurakhimov FD. Hypoglossal schwannoma: case report and literature review. Vopr. Neirokhirurgii Im. NN Burdenko 2019;83(5):51 no. doi:10.17116/neiro20198305151.
[6] Suri A, Bansal S, Sharma S, Mahapatra AK, Kale SS, Chandra S, et al. Management of hypoglossal schwannomas: single institutional experience of 14 cases. J. Neurol. Surg. Part B Skull Base 2014;75(03):159–64 no. doi:10.1055/s-0033-1356924.
[7] Tucker A, Miyake H, Tsuji M, Ukita T, Nishihara K, Ohmura T. Intradural microsurgery and extradural gamma knife surgery for hypoglossal schwannoma: case report and review of the literature. Min - Minim. Invasive Neurosurg 2007;50(6):374–8 no. doi:10.1055/s-2007-993206.
[8] Di Pascuale I, Brito N, Zerpa J, Roa C, Faria G. «Hypoglossal nerve schwannoma: case report and literature review». World Neurosurg 2020;135:205–8. doi:10.1016/j.wneu.2019.12.006.