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Case Report

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Hypopharyngeal schwannoma: A case report of a rare entity

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Abstract

We report the case of a 55-year old Caucasian female who presented with dysphagia after being operated for a brain glioblastoma. Fiberoptic endoscopy and magnetic resonance imaging showed a submucosal tumefaction of the posterior hypopharyngeal wall. Direct laryngoscopy and biopsy did not reveal a definitive diagnosis. The lesion was completely removed using a transoral C02 laser, and histopathological examination of the lesion showed a diagnosis of hypopharyngeal schwannoma. The patient recovered uneventfully and has remained clinically and radiologically disease-free for 6 months. Surgical excision and S100 protein immunohistochemistry remain the gold standards for treatment and diagnosis of hypopharyngeal schwannomas.

Keywords: Hypopharynx, schwannoma, S100 proteins, surgery.

Introduction

Schwannomas are benign nerve sheath tumors mostly affecting cranial nerves with a known genetic cause (mutation of a gene coding for a Schwann cell protein) and distinct histological features.[1] Only a dozen hypopharyngeal cases, originating from the pharyngeal plexus, have been reported in the literature.[2] Published epidemiological data report an incidence of all schwannomas to be 1/50,000, of which the majority are intracranial and asymptomatic.[1] Clinical presentation is due to the local effects of the tumor mass and are thus non-specific. Magnetic resonance imaging (MRI) was shown to be the superior radiological modality, while ultrasonography, computerized tomography (CT), scintigraphic evaluation and fine-needle aspiration cytology provide additional data to guide the diagnostic process.[1,3,4] Here, we present a case of hypopharyngeal schwannoma due to its rarity and aim to provide a reminder to clinicians of the

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need to consider schwannomas in the differential diagnosis of hypopharyngeal tumors.

Case report

A 55-year-old Caucasian woman presented to our otolaryngology-based outpatient clinic with a one-year history of difficulty swallowing. Her symptoms began shortly after being operated for a brain glioblastoma, after which she underwent chemoradiotherapy. At the time of presentation, she had no other symptoms apart from dysphagia, and clinical examination using fiberoptic endoscopy showed a posterior pharyngeal wall tumefaction at the oro- and hypopharynx border.

A neck MRI showed an oval-shaped expansive tumor 3x2 cm in size at the oro-hypopharyngeal border, with inhomogeneous signals on T2-weighted MRI sequence, and intensive opacification on T1-weighted post-contrast sequence. The tumor extended beyond the midline and caused contralateral displacement of the epiglottis (Figure 1).

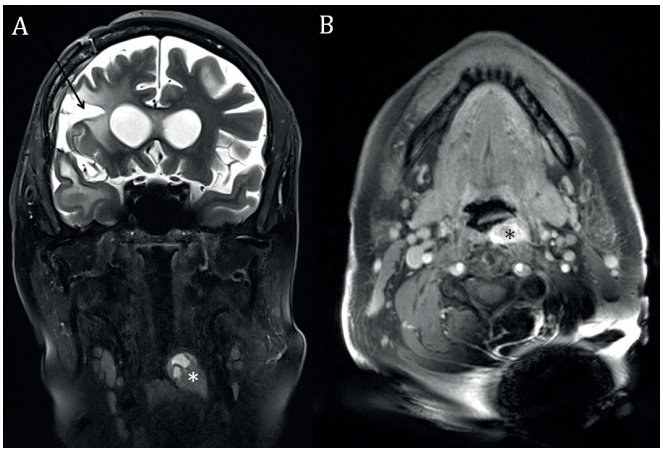

Figure 1. Head and neck magnetic resonance image (MRI) showing an ovoid tumor with mixed-signal intensity on the T2 weighted image of the oro-hypopharyngeal border on the left side (A); Intense contrast enhancement of the tumor is seen on the axial T1 image and the epiglottis is dislocated contralaterally by the mass (B); asterisk indicates the schwannoma and arrow indicates postoperative changes in the brain parenchyma.

First, microdirect laryngoscopy and biopsy were performed, however, histopathological examination failed to yield a definitive diagnosis. Next, a surgery was scheduled and the lesion was completely excised using a transoral C02 laser (Figure 2). Histopathological analysis of the lesion revealed features of a tumor consistent with a diagnosis of schwannoma, namely, Verocay bodies, specific for schwannomas, were observed on hematoxylin and eosin staining and the sample showed S100 protein immunoreactivity (Figure 3). The postoperative period was uneventful and the patient remains clinically and radiologically disease-free at the 6-month follow-up. Written informed consent was obtained from the patient to report his condition as a case report.

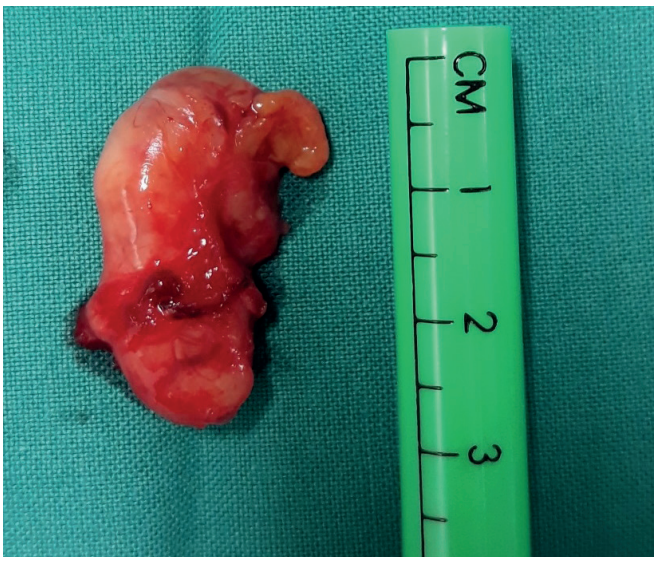

Figure 2. Macroscopic image of the excised specimen.

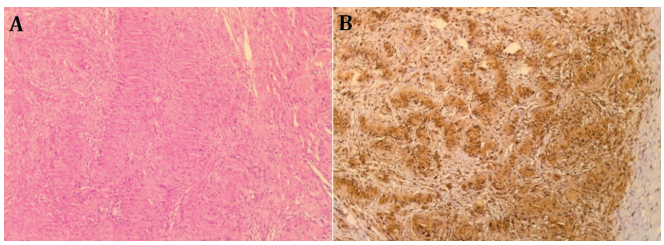

Figure 3. Histopathological image of the tumor tissue sample. Histopathological image of the tissue sample: (A) Spindle tumor cells are arranged in interlacing fascicles, cells are whirling and palisading, and Verocay bodies are seen (HE x 100); (B) Diffuse and intense S100 protein immunoreactivity (x100).

Discussion

Hypopharyngeal schwannomas occur as either sporadic

cases or as neurofibromatosis-associated tumors.[1] Malignant transformation and recurrence of these tumors are very rare.[5] Our patient had a unique co-occurrence of a brain glioblastoma and hypopharyngeal schwannoma, yet her additional clinical data and genetic analysis did not show her to harbor the NF stigmata or mutations. This fact is important, since it is known that glioblastoma occurs more often in NF patients.[6] Even though the patient was diagnosed with a hypopharyngeal schwannoma with a one-year delay following glioblastoma surgery, it is considered a co-occurrence, since the schwannoma was visible, yet overlooked, on the MRI performed during glioblastoma workup.

Epidemiological data on pharyngeal schwannomas is non-existent, due to their rarity; however, epidemiological data for vestibular schwannomas show that these tumors mostly occur in Caucasians in the fifth decade and with either sex.[7] On the other hand, glioblastoma occurs more often at a later age and in the male sex.[6] Our patient was a "typical" schwannoma patient, yet she suffered from glioblastoma at a younger age than the average glioblastoma patient.

Most schwannomas are benign, asymptomatic, sporadic tumors; however, they also occur in several tumor suppressor syndromes, most notably neurofibromatosis.[8] The typical radiological appearance of schwannomas is not as specific as their histopathology and includes homogeneous hypo- or isodensity and contrast enhancement on CT and T1 hypointensity and contrast enhancement and T2 hyperintensity on MRI.[8] Pathognomonic histopathological features of these tumors include alternations of two types of areas – cellular Antoni A areas in which Schwann cells are arranged into dense spindles and hypocellular Antoni B areas composed of scarce cells enclosed in the myxoid stroma. Also, schwannomas show diffuse and strong S100 immunopositivity in contrast to neurofibromatosis.[8] Previously described hypopharyngeal schwannomas, as well as the tumor reported herein, share these radiologic and histologic findings.[2,9]

Our patient presented with dysphagia, a clinical feature typical for hypopharyngeal tumors.[2] By itself, dysphagia has a wide differential diagnosis ranging from central nervous system lesions and neuromuscular disease to obstructive lesions of the alimentary tract. In the present case, the differential diagnosis was significantly narrowed, as fiberoptic endoscopy showed a submucosal impaction of the posterior hypopharyngeal wall. As in other regions, pharyngeal tumors can be either malignant or, less commonly, benign. The chief differences in clinical presentation between the two groups are that malignant lesions tend to progress rapidly and, apart from local signs, present with general symptoms. Differential diagnosis of benign lesions, which are less frequent in this region than malignant ones, includes neurofibromas, lymphangiomas, hemangiomas, lipomas, etc.[2] Both clinical presentation and imaging findings are informative, yet non-specific in delineating between individual entities, therefore a histopathological examination of the lesion is required for definitive diagnosis.

Surgery is considered the curative treatment modality for hypopharyngeal schwannomas and different open and endoscopic approaches have been described so far.[2] Our patient underwent a successful total tumor removal using a C02 laser. The postoperative course was uneventful and the patient remains clinically and radiologically disease-free at the 6-month follow-up. The rarity of hypopharyngeal schwannomas makes it difficult to develop evidence-based recommendations on follow-up regimes. Such consensus among experts is lacking for more frequent regions, such as vestibular schwannomas. On average, neurosurgeons and otolaryngologists follow their patients five years after surgery, during which period the patients undergo five MRI scans.[10] Since both hypopharyngeal and vestibular schwannomas share a common biology and it was shown for the latter that the risk of recurrence after surgery is minute, we believe that the prevailing follow-up practice for vestibular schwannomas can also be applied to hypopharyngeal cases.

Conclusion

Preoperative diagnosis of head and neck schwannoma is difficult due to their rarity and non-specific presentation. For these lesions, surgical excision is the curative gold standard and histopathological S100 protein immunoreactivity is its diagnostic counterpart. Fiberoptic endoscopy and MRI are useful tools in the postoperative follow-up.

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