Schwannoma of the nasal septum presenting as a multicentric neuronal tumour

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Abstract. Schwannoma of the nasal septum presenting as a multicentric neuronal tumour. Problem: Schwannomas (neurilemmomas) are benign primary tumours that arise from Schwann cells. Schwannomas arising from the nasal septum are exceptionally rare. Here, we report a unique case of schwannoma of the nasal septum presenting as a multicentric neuronal tumour.

Results: A 40-year old male sustained intermittent left tinnitus. Magnetic resonance imaging revealed masses near the nasal septum and upper cervical cord in addition to a tumour in the left cerebellopontine (CP) angle. The tumour in the nasal septum was completely resected by endoscopic endonasal surgery and diagnosed as a typical schwannoma. The CP angle tumour was treated with stereotactic radiosurgery, while the asymptomatic cord lesion showed no significant growth and remains under observation.

Conclusion: Endoscopic endonasal surgery is useful for the resection of schwannomas of the nasal septum. Schwannomas of the nasal septum may present as multiple neuronal tumours.

Introduction

Schwannomas (neurilemmomas) are benign primary tumours that arise from Schwann cells. Between 25% and 45% of schwannomas are found in the head and neck region.1 The vestibular nerve is thought to be the most predisposing site of origin.1 Schwannomas affecting the nasal and paranasal cavities are infrequent and estimated to comprise 4% of all cases.2 Schwannomas arising at the nasal septum are exceptionally rare and reportedly comprise only 6.9% of cases originating from the nasal and paranasal regions.3 Of note, a fraction of schwannomas are thought to occur in association with neurofibromatosis.4 Here, we report a unique case of schwannoma of the nasal septum presenting as a multicentric neuronal tumour.

Case report

A 40-year-old man patient was referred to our department for intermittent left tinnitus, which he experienced for one month. His medical history was unremarkable, and he had no family history of genetic disorders. At presentation, the patient showed no focal neurological deficit. A physical examination found no skin abnormalities such as stigmata, café-au-lait spots, or cutaneous neurofibromas. The patient did not show any skeletal deformity, and the neuro-ophthalmological examination did not identify any pathology.

Pure-tone audiometry did not show hearing loss. Cerebral magnetic resonance imaging revealed a heterogeneously enhancing mass at the left cerebellopontine (CP) angle, which was 2.5 cm in diameter, and compressing the pons and cerebellar hemisphere (Figure 1A). Unexpectedly, two additional tumourous lesions were found at the same time at the base of the nasal septum and upper cervical cord. The former was round, heterogeneously enhancing, 3 cm in dimension, and extended upward into the frontal sinus with fluid accumulation. Invasion into the intracranial, orbit, or sphenoethmoidal sinuses was not noted (Figure 1B-D). The latter was located intramedullar and was fusiform, inhomogeneously enhancing, and 5 mm in diameter (Figure 1C, D). The tumour in the nasal septum was asymptomatic; however, it...
Figure 1

Post-contrast axial (A), coronal (B), and sagittal (C) magnetic resonance images and a sagittal T2-weighted sequence (D) showing a heterogeneously enhancing mass, 2.5 cm in diameter, at the left cerebellopontine (CP) angle (A, arrowheads). Two additional tumours appear at the base of the nasal septum (A-D, asterisk) and upper cervical cord (C, D, arrow). The nasal lesion was round, heterogeneously enhancing, 3 cm in dimension, and extended upward into the frontal sinus with fluid accumulation, whereas invasion into the intracranial cavity, orbit, or sphenethmoidal sinuses was not found. The cord lesion is located intramedullary, and was fusiform, inhomogeneously enhancing, and 5 mm in diameter.
Schwannoma of the nasal septum

was of considerable size, whitish in color, capsulated, elastic and soft, and its vascular was fed by the anterior ethmoidal arteries.

Recently, progress has been made in the field of otolaryngology with regard to the use of endoscopic endonasal surgery. In recent years, this type of surgery has also been successfully performed on patients with skull-base tumours. Therefore, we decided to resect the tumour by endoscopic endonasal surgery guided by a neuronavigation system.

The surgery was performed as follows. A Killian incision was made from the left nasal cavity to the nasal septum mucosa, and the mucosa was separated subperiosteally as in septoplasty. Around the border with the perpendicular plate, the tumour could be clearly observed in the submucosa. An endoscope was inserted from this site into the other nasal cavity, and the tumour was resected even though the nasal mucosa on both sides of the nasal cavity was attached. However, the tumour in the frontal sinus was not resected. Using a neuro-navigation system, the portion of the tumour attached to the anterior cranial base was accurately identified and managed. The obstruction of the nasofrontal duct was released, and the part of the tumour extending into the frontal sinus was withdrawn through the duct. Finally, the tumour was removed without postoperative hyposmia and other functional impairments. The tumour was between the 2 mucoperichondrium.

The histologic diagnosis was consistent with Antoni type-A schwannoma (Figure 2). With the peripheral venous blood as a sample, neurofibromatosis-related genomic derangements were analyzed by direct polymerase chain reaction sequence and Multiplex Ligation-dependent Probe Amplification (MLPA) methods. These examinations found no abnormalities.

The CP angle tumour was treated using gamma knife radiosurgery, which resulted in a gradual regression without hearing loss or facial nerve paresis. The asymptomatic intramedullary tumour showed no neurological deficits or significant growth, and the patient has been under observation for 5 years.

Discussion

The present case is unique because the neuroimaging study for tinnitus simultaneously and unexpectedly delineated a CP angle tumour, schwannoma of the nasal septum, and an intramedullary cervical cord tumour. The schwannoma of the nasal septum was asymptomatic, although it had reached a considerable size, with obstruction of the nasofrontal duct. Therefore, we decided to resect it.

Sympathetic nerves distributing to the sepal blood vessels, parasympathetic fibers distributing to the sepal mucous glands, and sensory nerves distributing to the nasal septum have been suggested as sites of origin for schwannomas of the nasal septum. The schwannoma of the nasal septum was asymptomatic, although it had reached a considerable size, with obstruction of the nasofrontal duct. Therefore, we decided to resect it.

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Conclusion
Endoscopic endonasal surgery is useful for the resection of schwannomas of the nasal septum. Schwannomas of the nasal septum may present as multiple neuronal tumours.
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