CASE REPORT

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Olfactory Groove Schwannoma

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We present a case of olfactory schwannoma in a 16-year-old boy with headache and diplopia. Brain computed tomography (CT) scan and magnetic resonance(MR) imaging showed a huge mass in the subfrontal area resembling an olfactory groove meningioma. We performed a bifrontal craniotomy and found out the mass was attached to cribriform plate but was not related to the olfactory tract or bulb. The histopathological diagnosis of schwannoma was confirmed by immunohistochemical staining for S-100, vimentin and others. We describe the clinical manifestations, radiological characteristics, histological aspects, and differential diagnosis of this tumor with literature review.

KEY WORDS : Schwannoma · Olfactory groove · Immunohistochemistry · Olfactory tract or bulb.

Introduction

Intracranial shwannomas usually arise from the vestibular nerve, and less commonly from the trigeminal nerve, facial nerve and lower cranial nerves. Optic nerves and olfactory nerves are devoid of Schwann's cell layer, therefore, schwannomas can not develop from these nerves theoretically[5,12].

Solitary schwannomas arising from the olfactory groove are very rare and about twenty six cases of solitary schwannomas of the olfactory groove have been reported[1-4,6-11,13,15].

We report a case of isolated schwannoma in the region of the olfactory groove and review the pertinent literature, the neuroradiological and pathological features of the case as well as the origin of the schwannomas in this unusual location.

Case Report

A 16-year-old boy presented with headache of 5 days' duration and diplopia in 1 day prior to admission. There was no family history of neurofibromatosis. Physical examination was normal. He complained double vision when looked at left side, and mild hypesthesia on the left face.

A brain axial computed tomography(CT) scan showed a huge (7×6cm) frontal mass with irregular enhancement and a poorly defined margin (Fig. 1A). Coronal CT scan showed a large mass localized on the left side of the cribriform plate (Fig. 1B), and erosion of the left frontal skull base was noticed on three-dimensional CT scan (Fig. 1C). A brain MR imaging showed a left extra-axial mass with intense and heterogeneous enhancement in the subfrontal region (Fig. 2).

With a bicoronal incision and bifrontal craniotomy, a total excision of the tumor was performed. At operation, the tumor

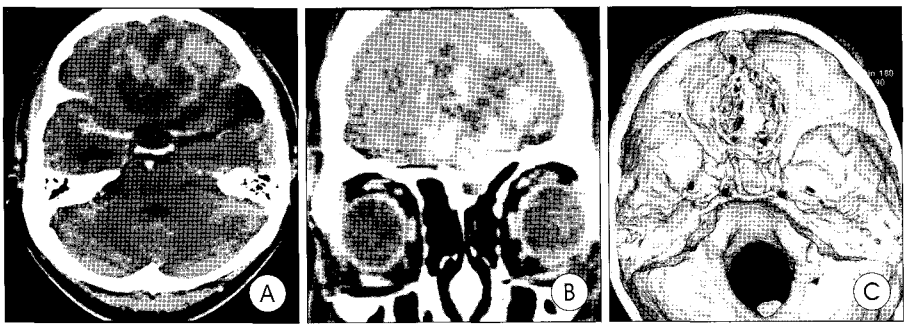

Fig. 1. A : Brain axial computed tomography(CT) scan shows a large frontal mass with a relatively irregular contour and with heterogeneous enhancement. B : Contrast enhanced coronal CT scan shows a mass localized on the left cribriform plate which is displaced downward and eroded. C : Three-dimensional CT scan of skull base shows the erosion and depression of the left cribriform plate.

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proved to be attached to the dura in the region of the cribriform plate and to the dural surface over crista galli. There was severe adhesion to the falx, but the olfactory bulbs and tract did not appear to be involved.

Table 1. Results of immunohistochemical staining

Immunohistochemical staining	Result
S-100 protein	Positive
Vimentin	Positive
GFAP	Weak positive
Pan CK	Negative
EMA	Negative
Chromo A	Negative
NSE	Negative
Synaptophysin	Negative
Ki-67	Less than 1%

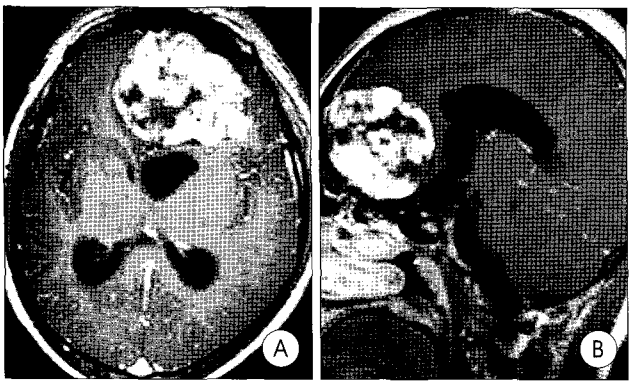

Fig. 2. Brain magnetic resonance imagings(axial and sagittal contrast-enhanced T1-weighted images) show an extra-axial mass with intense and heterogeneous enhancement in the left subfrontal area.

Histopathological examination revealed a partially encapsulated, paucicellular neoplasm which was composed of bipolar spindle shaped cells. They also showed Anthoni A type cellular areas and characteristic Verocay bodies (Fig. 3A) and Anthoni B type areas (Fig. 3B). In immunohistochemical staining, the cells were intensely positive for S-100 protein and negative for epithelial membrane antigen(EMA) (Fig. 3C, D). The immunohistochemical results are presented in Table 1. The pathologic diagnosis was confirmed to be schwannoma.

The patient recovered well and discharged without any neurological deficit two weeks after operation.

Discussion

Schwannomas commonly arise from the nerve sheaths of the peripheral and the cranial nerves and represent approximately 8% of all intracranial tumors[12]. Even though schwannomas have been reported to originate from all the cranial nerves except the optic nerve, the majority of intracranial schwannomas arise in particular from the vestibulocochlear nerve, and most of them originate in its vestibular division[12]. The next most frequent type of schwannoma arises from the trigeminal nerve, but these account for 2~3% of all intracranial schwannomas[12]. Very few cases of intracranial schwannomas without any cranial nerve or dural attachment have been reported in the literature[5,12]. Furthermore, solitary intracranial extraparenchymatous schwannomas related to the olfactory groove are very rare[1-4,6-11,13,15]. Since Sturm et al.[8], reported the first case of an olfactory schwannoma as a separate entity in 1968, 26 cases have been reported. Of these only three cases were found to be attached to the olfactory nerve[2,3]. In the case of an olfactory schwannoma, the pathogenesis of these intracranial schwannomas is puzzling as the olfactory nerve does not contain any Schwann cells[5,12]. Various theories regarding the possible origin of these tumors are centered around developmental and nondevelopmental origins[2,5,10,12,15]. The developmental theories suggest either transformation of mesenchymal pial cells into ectodermal Schwann cells or migration of the

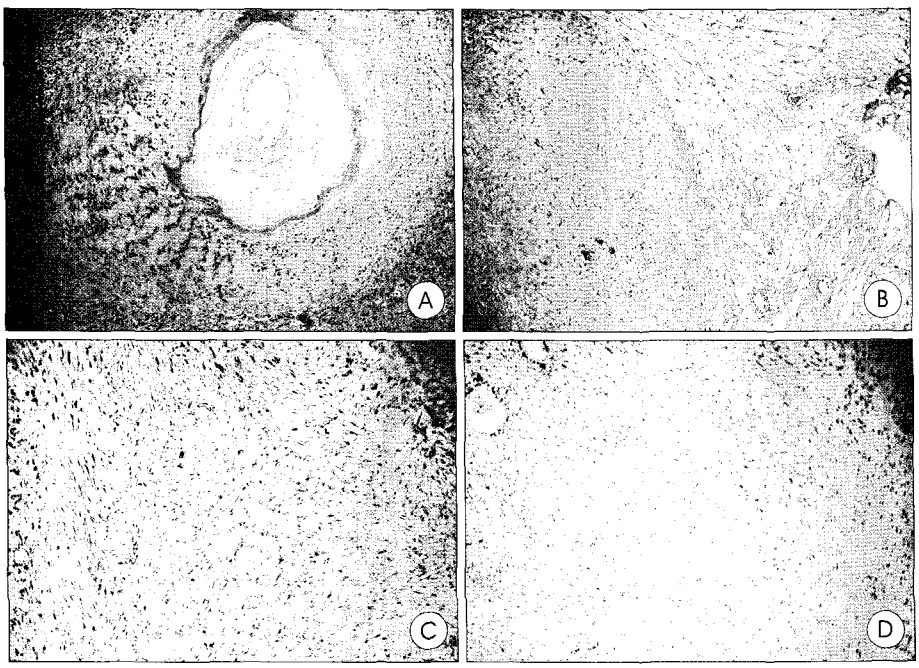

Fig. 3. Photomicrographs of the tumor specimen. A, B : The tumor consists of alternating areas of compact, elongated cells(A : Anthoni A type) and less cellular, loosely textured tumor areas(B : Anthoni B type)(H & E, ×40). Verocay bodies are characteristics of the schwannoma. C, D : Immunohistochemical staining shows the tumor cells strongly and diffusely expressed S-100 protein(×400)(C) and negatively expressed epithelial membrane antigen(×400)(D).

neural crest cells within the substance of the central nervous system[2,5,12]. On the other hand, the proponents of the non-developmental theories postulate that intracranial schwannomas arise from Schwann cells normally present in the adjacent structures such as the perivascular nerve plexus and the meningeal branches of the trigeminal and anterior ethmoidal nerves innervating the anterior cranial fossa[10,12,15]. Timothy et al.[10], advocated that the term olfactory groove schwannoma represents this pathological entity more appropriately than olfactory schwannoma, because the olfactory groove schwannoma almost certainly arose from the anterior ethmoidal nerve. In our case, the tumor was attached to the dura in the region of the cribriform plate and to the falx, but the olfactory bulbs did not appear to be involved. It suggested that the schwannoma might have originated from the small nerves in the basal dura matter.

Recently Yako et al.[13], performed the molecular genetic examination for a case of subfrontal schwannoma and revealed that neither NF2 gene mutation nor loss of heterozygosity of chromosome 22q, unlike common schwannomas. These genetic alterations are found in as much as 60~70% of sporadic ordinary schwannomas and regard as the essential for the tumourigenesis of common schwannomas. However, they did not detect any of them and suggested that this rare tumour may not share the major mechanism of schwannoma tumourigenesis.

The age and sex distribution of these schwannomas show some characteristics[4,14,15]. Many cases of olfactory groove schwannomas have occurred in males. The average age of diagnosis is 30 years old. In most cases the tumor had reached a large size by the time the patients were presented. Common presentations include headache, seizures, anosmia, and increased intracranial pressure. Neurological deficits are uncommon.

In the differential diagnosis of an extra-axial anterior fossa mass involving cribriform plate, one must initially consider meningioma[1,3,7]. Bony changes in meningioma are usually sclerotic but can rarely be destructive or scalloped[1,15]. The age at presentation is older in meningioma than in schwannoma patients. About 50% of patients with subfrontal schwannomas are younger than 30 years. Signal intensity in MRI does not help in the differential diagnosis between meningioma and schwannoma[1,3]. So most of olfactory groove schwannomas, most cases were misdiagnosed to be olfactory groove meningioma preoperatively.

Neuroblastoma, hemangiopericytoma, esthesioneuroblastoma, and metastatic disease are other unusual entities to include in the differential diagnosis[1,12,13]. Hemanigopericytoma has a polylobulated shape and very prominent flow voids. Esthesioneuroblastoma and metastatic disease usually show invasion of paranasal sinuses and extensive bony destruction.

Complete excision is the main therapeutic modality and cure due to the benign nature of the tumor[3,7,10,15].

Conclusion

We report a case of olfactory groove schwannoma without involvement of the olfactory bulb or tract. It is difficult to diagnosis this case before surgery and it deserves a certain interest due to its rarity. It is necessary to recognize the possibility of occurrence of a schwannoma in unusual location.

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