

Case Report

Olfactory Schwannoma-Case Report-

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Intracranial schwannomas preferentially arise from the vestibular branch of the eighth nerve, and rarely from the trigeminal nerve, facial nerve, and lower cranial nerves. Anterior cranial fossa schwannomas are extremely uncommon and few details about them have been reported. The patient was a 39-year-old woman whose chief complaints were anosmia and frontal headache for 2 years. The gadolinium (Gd)-enhanced magnetic resonance imaging (MRI) showed an extra-axial mass from ethmoid sinus to right frontal base region near the midline, with solid enhancement in lower portion and multicystic formation in upper portion. The tumor was totally resected via basal subfrontal approach. At operation, the tumor had cystic portion with marginal calcification and the anterior skull base was destructed by the tumor. The olfactory bulb was involved, and the tumor capsule did not contain neoplastic cells. The histopathological diagnosis was schwannoma. We report a rare case of anterior cranial fossa schwannoma with literature review.

KEY WORDS : Schwannoma · Olfactory nerve.

INTRODUCTION

Schwannomas are benign, slowly growing nerve sheath tumors which usually arise from any peripheral nerve containing Schwann cells including distal portions of the cranial nerve. However, the optic and olfactory nerve do not have Schwann cell layer and therefore schwannoma, theoretically, cannot develop in these nerves^{1,5-8,10,15}. Anterior cranial fossa schwannoma are very rare and only 34 such cases have been reported in the world literature so far^{1,7,11,13}. We report a rare case of anterior cranial fossa schwannoma with details of clinicoradiologic features and histopathological findings along with pertinent literatures.

CASE REPORT

A 39-year-old woman complained of anosmia with intermittent frontal headache for 2 years. Other neurological examinations were normal and her past medical and family history were unremarkable. A brain axial computerized tomography (CT) scan showed large lobulated inhomogeneous

density tumor in right anterior cranial fossa with linear or amorphous nodular calcification (Fig. 1A). Coronal CT scan showed erosion of the superomedial portion of right orbital wall and tumor localized in the cribriform plate (Fig. 1B). Three dimensional CT of the skull base demonstrated erosion of the cribriform plate (Fig. 1C). A magnetic res-

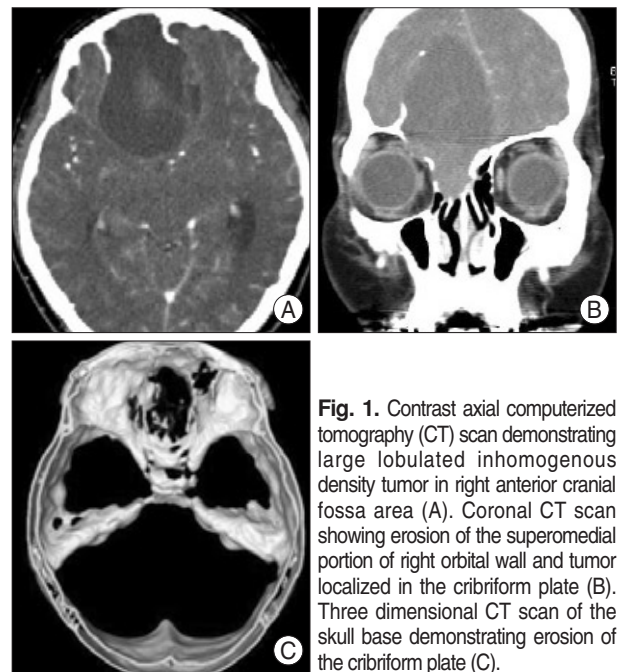


Fig. 1. Contrast axial computerized tomography (CT) scan demonstrating large lobulated inhomogeneous density tumor in right anterior cranial fossa area (A). Coronal CT scan showing erosion of the superomedial portion of right orbital wall and tumor localized in the cribriform plate (B). Three dimensional CT scan of the skull base demonstrating erosion of the cribriform plate (C).

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Fig. 2. An axial magnetic resonance imaging (MRI) with gadolinium showing an extra-axial mass in the right frontal region near midline and elevating the right frontal lobe (A). On the coronal and sagittal MRI with gadolinium showing a solid enhancement in the inferior portion of tumor and multi-septated rim enhancement in the superior portion of tumor with extension to ethmoid sinus and peri-orbital area (B and C).

onance imaging (MRI) showed an extra-axial mass in the right frontal region near midline elevating the right frontal lobe (Fig. 2A). The lesion appeared as inhomogeneous signal and the gadolinium enhanced scan showed solid enhancement in inferior portion of the tumor and multi-septated rim enhancement in superior portion of the tumor with extension to ethmoid sinus and peri-orbital area (Fig. 2B, C). The patient underwent a bifrontal craniotomy, and the tumor was totally resected via basal subfrontal approach. At operation, tumor had cystic portion with firm consistency and hypovascularity. Tumor was attached to the cribriform plate and was related to the olfactory bulb but tumor capsule did not contain neoplastic cells. The postoperative imaging scan showed a total resection of the tumor with mild hemorrhagic scar in previously tumor site (Fig. 3A, B). Histopathological examination of haematoxylin and eosin stained sections showed Antoni type A and Antoni type B cellular patterns (Fig. 4A, B). In the immunohistochemical finding, the tumor cells were strongly positive for S-100 protein and negative for epithelial membrane antigen (EMA) and Ki-67 labeling index was 0%, confirming the diagnosis of schwannoma (Fig. 4C, D). Postoperatively, the olfactory function was improved, and the patient was discharged without any neurological deficit at 2 weeks after operation.

DISCUSSION

Intracranial schwannomas represent approximately 6% to

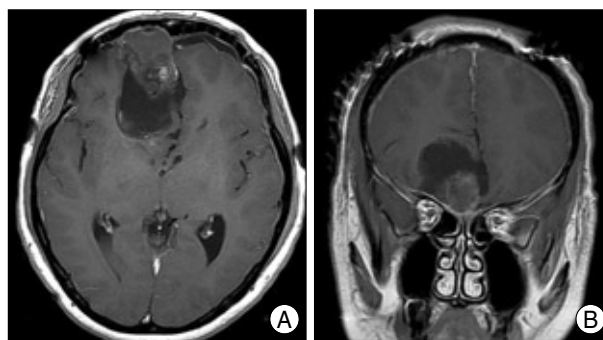


Fig. 3. Postoperative gadolinium enhanced axial (A) and coronal (B) magnetic resonance imaging scan demonstrating total tumor resection with mild hemorrhagic scar in previously tumor site.

8% of all intracranial tumors and usually arise from the Schwann cell layer of vestibular branch of eighth nerve or less commonly from the fifth nerve, seventh nerve and lower cranial nerves^{1,6-8,10,15}. However, the olfactory and optic nerve lack a Schwann cell layer and thus are not prone to tumors from these nerves¹⁴. Among several location's schwannoma, anterior cranial fossa schwannoma are quite rare lesion and only 34 cases reported in the world literature until 2007^{1,7,11,13}.

In terms of how anterior cranial fossa schwannoma arise, several theories of the origin have been put forth. Various theories regarding the possible origin of these tumors are centered around the developmental and non-developmental origins^{1,4,5,8-10}. The developmental theories suggest either transformation of mesenchymal pial cell into ectodermal Schwann cells or migration of the neural crest cells within the substance of the central nervous system. On the other hand, the non-developmental theories suggest that anterior cranial fossa schwannomas arise from Schwann cells normally present in the adjacent structures such as the perivascular nerve plexus and the meningeal branches of the trigeminal nerve and anterior ethmoidal nerve innervating the anterior cranial fossa and olfactory groove. And, reactive change after injury, that is, formation of Schwann cell from multipotential mesenchymal cells after injury in patients with pathological changes like multiple sclerosis or infarction has also been described^{2,5}. In our case, the tumor was partially attached to the dura at the cribriform plate and also involved the olfactory bulb, so we concluded that the most likely tumor origin was the fila olfactoria; it is known to acquire a Schwann cell sheath that extends approximately 0.5 mm beyond the olfactory bulb.

Anterior cranial fossa schwannoma has many different characteristics contrary to other location schwannomas as follows : beginning at an earlier age (mean 32.2 years), affects mostly men (63%), calcification is rare (8%) and in imaging finding, the tumor is composed of the solid portion in

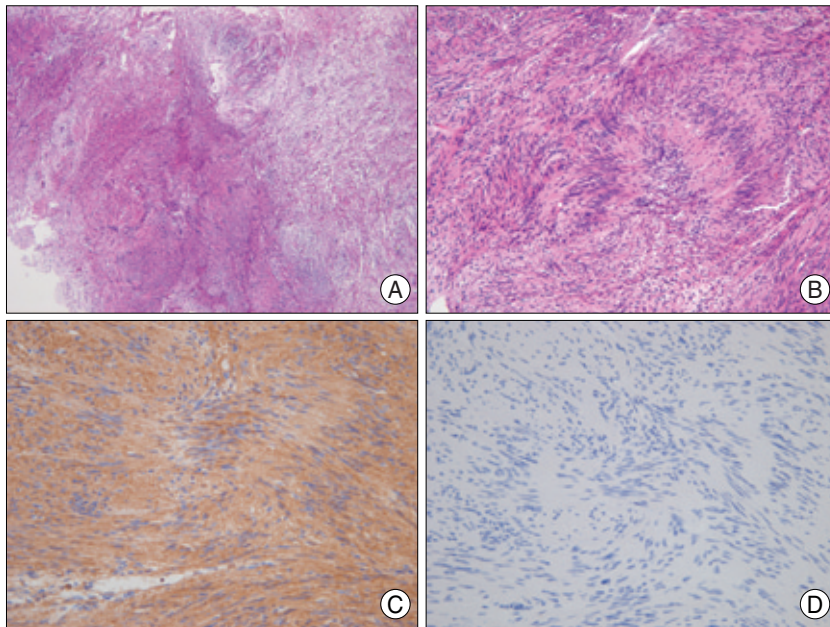


Fig. 4. Histopathological examination of the surgical specimen showing the tumor consisted of alternating areas of compact, elongated cells (Antoni type A) and less cellularized tumor areas (Antoni type B) (H & E, $\times 200$) (A and B). Photomicrographs of immunohistochemical examination showing the tumor cells were positive for S-100 protein (C) and negative for epithelial membrane antigen (IHC $\times 400$) (D).

Table 1. A comparison of 4 cases of anterior cranial fossa schwannomas in the Korean literature

Author	Sex/Age	Symptom	Olfaction	Radiologic finding	Operation finding
Yang SH ³⁾ (2002)	M/55	Headache	Normal	Solid aspect Calcification (-)	OG attachment OB not detected
Park JH ⁷⁾ (2006)	M/17	Headache Diplopia	Normal	Solid aspect Calcification (-) Erosion of skull base	CP attachment OB not involved
Woo JY ¹¹⁾ (2007)	M/38	Decreased VA	Normal	Cystic aspect Calcification (-)	OG attachment OB not involved
Our case (2008)	F/39	Headache	Anosmia	Cystic aspect Calcification (+) Erosion of skull base	CP attachment OB thinned

VA : visual acuity, OG : olfactory groove, OB : olfactory bulb, CP : cribriform plate

chiefly^{1,4-6,15}). Dissimilarly to typical characteristics of previously reported anterior cranial fossa schwannoma, our case showed cystic portion and tumor margin had thick calcification on preoperative imaging study (Table 1).

In the differential diagnosis of an extra-axial anterior cranial fossa involving cribriform plate with extension to the ethmoidal sinus, metastatic disease, meningioma, neuroblastoma and others should be included^{2,6,7,12,15}). But, in our case, intraoperative finding was sufficient for typical schwannoma because of the tumor's hypovascularity, about 30 cc cystic fluid collection with well demarcated tumor capsule margin and marked bony erosion. Also, in the immunohistochemical study, the tumor showed strong positive for S-100 and negative for EMA with low MIB-1 labeling index (0%).

Because of the tumor's benign nature, complete resection of the tumor is treatment modality and adjunctive therapy is not needed to tumor treatment. Also, the patient's prognosis is very excellent after surgical resection.

CONCLUSION

We report a rare case of anterior cranial fossa schwannoma with marginal calcification. Surgical resection is curative modality. As in this case, the schwannoma should be included in differential diagnosis of the anterior cranial fossa tumor.

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