

“Pure” Suprasellar Schwannoma Presented with Communicating Hydrocephalus: A Case Report

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Schwannoma is a benign peripheral nerve sheath tumor originating from Schwann cells. Most intracranial schwannomas arise from vestibular nerve and schwannoma in the suprasellar region is extremely rare. A 64-year-old man presented with walking disturbance and blurred vision for three months. Lateral hemianopsia in the left eye and brachybasia were observed. Magnetic resonance imaging revealed a suprasellar tumor with strong contrast enhancement associated with communicating hydrocephalus. The cerebrospinal fluid tap test improved gait disturbance. Hypothalamic stimulation test revealed hypo-reaction of GH, FSH and LH. After ventriculo-peritoneal shunting, the tumor was totally removed via a bilateral frontobasal approach with a clinical diagnosis of craniopharyngioma. No adhesion was observed between the tumor and surrounding structures such as meninges and brain. The histopathological diagnosis was schwannoma. Here we report a case of suprasellar schwannoma associated with communicating hydrocephalus that has not ever been previously reported, with special reference to its pathogenesis.

Keywords: suprasellar schwannoma, communicating hydrocephalus, CSF protein

Introduction

The area around the sella turcica is anatomically complex, which makes differential diagnosis of specific tumors in this region difficult. Intracranial schwannomas, comprising 8–10% of all primary intracranial neoplasms,^{1–4} commonly arise from the vestibular nerve in the cerebellopontine angle, so schwannoma arose in the intrasellar or suprasellar is very rare. Here we present clinical course and immunohistological findings of a case of a suprasellar schwannoma with communicating hydrocephalus.

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Case Report

A 64-year-old man presented with gradually worsening gait disturbance for three months. He noted blurred vision while watching the television or reading the newspaper during the same period. He had a past history of hypertension, urolithiasis and retinal detachment in the right eye. Neurological examination revealed wide-based gait with short strides. It took 20 seconds for the 3-meter up and go test. Neither motor paralysis nor sensory disturbance was observed. On ophthalmological examination, lateral hemianopsia was revealed in the left eye. Visual acuity of the right eye was limited to perception of hand motion, which was attributed to his previous retinal detachment. The other cranial nerves were normal. The patient had cognitive failure and urinary incontinence on admission. Magnetic resonance imaging (MRI) showed 24 × 20 × 24-mm heterogeneous enhanced mass, which compressed the optic chiasm upward, and severe communicating hydrocephalus (Fig. 1). The blood test was normal, but hypothalamic stimulation test revealed hypo-reaction of GH, FSH and LH. Lumbar puncture was performed, and the cerebrospinal fluid (CSF) protein levels were slightly elevated (143 mg/dl). Following this procedure, the patient's performance of gait in the 3-meter up and go test improved to 14 seconds.

The preoperative diagnosis was craniopharyngioma associated with normal pressure hydrocephalus. After ventriculo-peritoneal shunting (Medos programmable system), the gait disturbance significantly improved and the patient became able to walk stably by himself. Subsequently, the tumor was excised via a bilateral frontobasal approach. The tumor was solid hard with light yellowish color and had high vascularity. The stalk of the pituitary gland was compressed to the left side. It seemed that feeding arteries of the tumor came from back and rightward of the tumor. No adhesions were observed between tumor and surrounding structures such as diaphragm sellae, pituitary stalk, chiasma and internal carotid artery and brain (Fig. 2).

Postoperatively, the patient temporally developed diabetes insipidus for a few days. The left lateral hemianopsia improved after tumor resection. With the improvement of his gait, vision, cognitive and urinary function, he was able to perform normal activities of daily living.

The histopathological diagnosis of the tumor was schwannoma. Microscopically, the tumor consisted of spindle cells with nuclear palisading and Antoni A/B

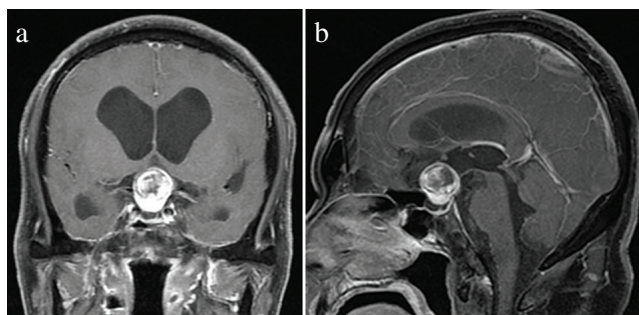


Fig. 1 Coronal (a) and sagittal (b) contrast-enhanced T1-weighted magnetic resonance images. Target-like and indistinct central enhancement was observed in the suprasellar tumor.

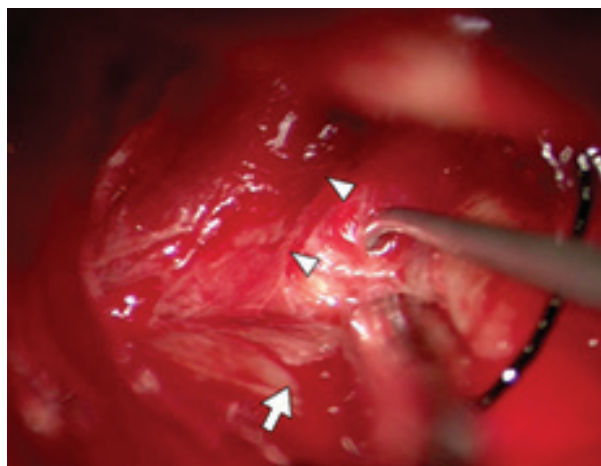


Fig. 2 An intraoperative view of the tumor (arrow). Tumor adhesion with the surrounding structures including the hypophyseal stalk (arrow head) was not seen.

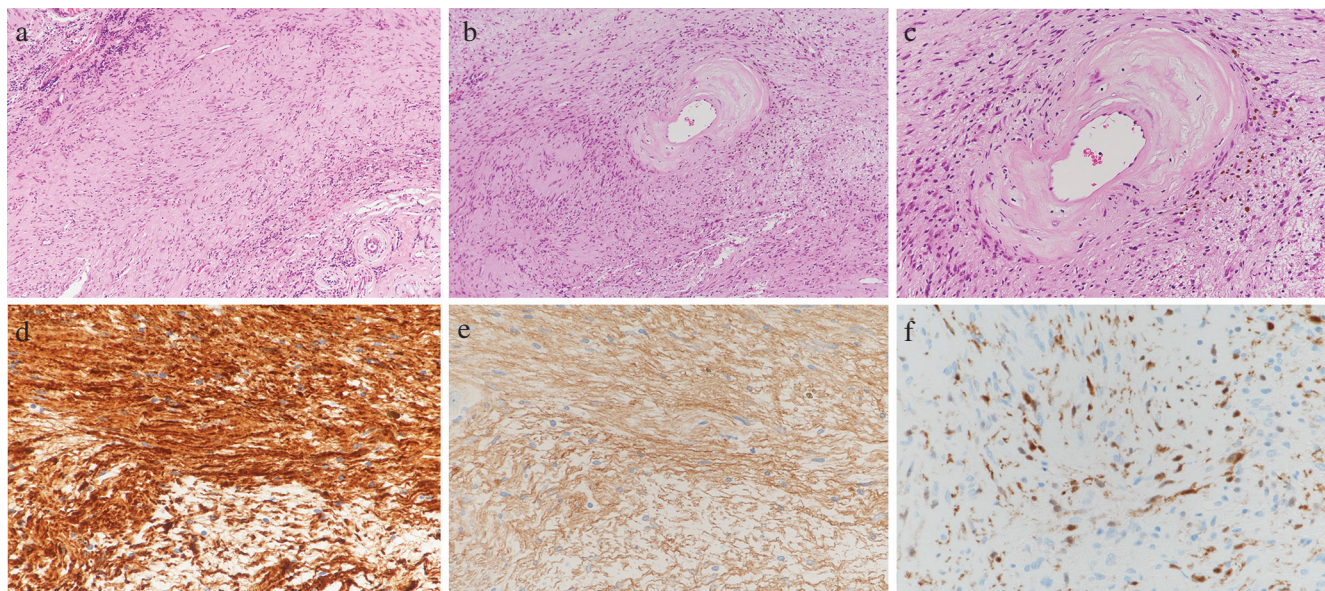


Fig. 3 (a–c) The tumor is consisted of spindle cells arranged in a fascicular pattern. Nuclear palisading (a, $\times 100$), Antoni A/B pattern (b, $\times 100$), and perivascular hyalinization and hemosiderin laden cells (c, $\times 200$) are observed (H&E). (d) Strong staining of S-100 protein ($\times 400$). (e) Strong intercellular staining of basement membrane for collagen type IV ($\times 400$). (f) Scattered immunostain for calretinin of the tumor cytoplasm and nuclei ($\times 400$).

(Figs. 3a and 3b). Strong hyalinization was observed in the center of the tumor and blood vessels. Perivascular accumulation of hemosiderin-laden macrophages was found (Fig. 3c). There was mild atypia in tumor cells but no mitosis was observed and the MIB-1 labeling index was approximately 1.0%. The tumor cells were not immunoreactive for any epithelial markers (AE1/AE3, EMA), glial fibrillary acidic protein (GFAP), CD34 and signal transducer and activator of transcription 6 (STAT6) but immunoreactive for S-100 protein (Fig. 3d) and B-cell lymphoma (Bcl-2). Intercellular space was stained by collagen type IV (Fig. 3e). Scattered nuclear and cytoplasmic expression of calretinin was identified in the tumor cells (Fig. 3f).

Discussion

Schwannomas, relatively common intracranial tumors, mostly originate from vestibular and trigeminal nerves but rarely from other cranial nerves.²⁾ Schwannomas arose in the suprasellar region are very rare. Here we briefly reviewed a total of 18 cases, including the present case, of schwannomas located in the suprasellar region^{2–16)} (Table 1).

The patient's age was ranged from 19 to 79 (average age 49.0) and half of the patients were male (55.6%). In 16 cases, the suprasellar mass was extended from the intrasellar region (88.9%) and mass was located purely in the suprasellar region in only two cases including the present case. No involvement of cranial nerves was noted in our patient, as was true in most of the reported cases, with the exception of the 3rd and 5th nerve involvement in one case each. Temporal hemianopsia is one of the major symptoms of schwannoma located in the suprasellar region. Gait disturbance was seen in two patients with hydrocephalus and endocrine dysfunction was reported in 11 cases.

Table 1 Summary of 18 cases of schwannoma located in suprasellar region

Case	Author, year	Age	Sex	Location	Hydrocephalus	Symptoms	Endocrine disfunction	Nerve involvement	Surgical approach
1	Perone, ⁸⁾ 1984	39	M	Intrasellar and suprasellar	ND	Headache	–	5th?	Transsphenoidal
2	Wilberger, ⁴⁾ 1989	62	F	Intrasellar and suprasellar	ND	Visual loss	+	–	Transsphenoidal and transcranial
3	Civit, ⁹⁾ 1997	41	M	Intrasellar and suprasellar	ND	Bitemporal hemianopsia	–	–	Transsphenoidal
4	Bhagat, ¹⁰⁾ 2002	68	M	Intrasellar and suprasellar	ND	Bitemporal hemianopsia	+	–	Transsphenoidal
5	Whee, ¹¹⁾ 2002	39	M	Intrasellar and suprasellar	ND	Bitemporal hemianopsia	+	–	Transsphenoidal
6	Maatens, ⁵⁾ 2003	33	F	Intrasellar and suprasellar	ND	Left frontal headache, diminished visual acuity, amenorrhea	+	–	Transsphenoidal
7		56	F	Intrasellar and suprasellar	Obstructive	Bitemporal hemianopsia, cognitive decline, personality change, gait disturbance	+	–	Transsphenoidal
8	Esposito, ¹²⁾ 2004	73	M	Intrasellar and suprasellar	ND	Hyponatremia, bitemporal hemianopsia	+	–	Transsphenoidal
9	Honegger, ³⁾ 2005	79	F	Intrasellar and suprasellar	ND	Syncope, headache, fatigue, left temporal visual field defect	+	–	Transsphenoidal
10	Yoon, ¹³⁾ 2005	34	M	Intrasellar and suprasellar	ND	Headache	+	–	Transsphenoidal
11	Park, ¹⁴⁾ 2009	49	F	Intrasellar and suprasellar	ND	Head ache, vomiting, bitemporal hemianopsia	–	–	Transsphenoidal and transcranial
12	Mohammed, ²⁾ 2010	19	F	Primary suprasellar	Obstructive	Inferior left visual defect, right temporal hemianopsia	ND	–	Bifrontal
13		45	F	Intrasellar and suprasellar	ND	Irregular menses, headache, right-sided facial pain	+	–	Transsphenoidal
14	Cugati, ⁶⁾ 2012	42	M	Intrasellar and suprasellar	ND	Bifrontal headache, progressive visual loss	–	–	Transsphenoidal
15	Senapati, ⁷⁾ 2014	24	F	Sella, suprasellar and parasellar area	ND	Diplopia and ptosis of left eye, visual loss, headache,	ND	3rd	Left pterional
16	Liu, ¹⁵⁾ 2016	50	M	Intrasellar and suprasellar	ND	Decreased visual acuity	–	ND	Transsphenoidal
17	Kong, ¹⁶⁾ 2016	65	M	Sella, suprasellar and parasellar area	ND	Temporal hemianopsia	+	ND	Transsphenoidal
18	presented case, 2016	64	M	Primary suprasellar	Communicating	Gait disturbance, left lateral hemianopia	+	–	Bifrontal

ND: not described.

Radiological features of the parasellar schwannoma are less distinctive. Parasellar schwannoma shows isointensity on T1, high intensity of T2 with homogeneous or heterogeneous enhancement.^{1,2,6,11,13,15)} Major differential diagnoses of the pure suprasellar mass are meningioma and craniopharyngioma. Typical meningioma shows iso-hypointensity on T1, iso-hyperintensity on T2 and often has flow void in the mass, with homogeneous enhancement and dural tail sign.¹⁷⁾ Majority of craniopharyngioma have cysts whose fluid is usually hyperintensity on T1.¹⁷⁾ Calcification often observed in 20–50% of meningioma and up to 80% of craniopharyngioma,¹⁷⁾ however, calcification is not identified in parasellar schwannoma. In 15 cases, the tumors were removed via a transsphenoidal route under a tentative preoperative diagnosis of pituitary adenoma.

The histological origin of suprasellar schwannoma remains obscure. Some authors have hypothesized that these tumors arise from Schwann cells that are located in the nerve plexus adjacent to the medial wall of the pituitary fossa or from perivascular nerve cells and autonomic vasomotor nerve plexi.^{1–3)} Some have suggested that ectopic Schwann cells in the walls of the sella turcica give rise to this tumor,^{2,3)} while others attribute it to the transformed pial or pluripotent mesenchymal cells; the putative origin of the cell gives rise to intracranial schwannoma.^{2,3)} Considering the position of the pituitary stalk, we speculate that the origin of the tumor is located in the inferior to the chiasm and right and forward from pituitary stalk.

Histopathologically, typical schwannoma consists of biphasic Antoni A tissue and Antoni B tissue. Long club-shaped nuclei, nuclear palisading is seen in Antoni A tissue. Perivascular hyalinization and accumulation of hemosiderin-laden macrophages are characteristic diagnostic findings for a slowly-growing tumor like schwannoma.¹⁸⁾ Immunohistochemically, schwannomas, contrary to meningiomas, are usually negative for the epithelial marker such as EMA. Schwannomas are also not immunoreactive for GFAP, while most pituitary adenomas are immunoreactive for GFAP. One characteristic features observed in schwannoma, but rarely in pituitary adenoma, is pericellular basal lamina, as demonstrated by immunoreactivity for collagen type IV¹⁸⁾ (Fig. 3e). Immunonegativity for CD34, STAT6 and immunoreactivity for Bcl-2 suggest the tumor has no reliability for solitary fibrous tumor. Additionally, multifocal nuclear and cytoplasm expression of calretinin may suggest schwannoma (Fig. 3f). Fine et al. have shown immunoreactivity for calretinin in 96% of schwannoma but in 7% of neurofibroma.¹⁹⁾ Takei et al. reported calretinin may help distinguishing intracerebral schwannoma from its mimicker.²⁰⁾ In the present case, microscopic appearance and immunoreactivity for collagen IV and S-100 led to pathological diagnosis of schwannoma.

This is the second case of schwannoma purely within the suprasellar region and the first reported case of suprasellar schwannoma with communicating hydrocephalus. Communicative hydrocephalus is a common complication of vestibular schwannoma, with a prevalence of approximately 11%.²¹⁾

A proposed pathogenesis in such cases is the sloughing of tumor proteins into CSF. A high CSF protein level results in plugging at the arachnoid granulations and reducing CSF resorption.²¹⁾ Our patient had a slightly elevated CSF protein level (143 mg/dl), suggesting that his communicating hydrocephalus might have developed as a consequence of the tumor. Among 18 cases of suprasellar schwannoma, obstructive hydrocephalus was noted in two cases due to compression by the tumor.^{2,4)} Communicating hydrocephalus has never been reported, though hydrocephalus was not resolved despite adequate debulking and required ventriculoperitoneal shunting in one case.⁴⁾ To our knowledge, there are only two reported cases for communicating hydrocephalus due to other pathology of seller region tumors for craniopharyngioma and iatrogenic one.^{22,23)} Therefore, schwannoma would be one of the tumor types presenting with communication hydrocephalus with suprasellar region tumors.

Conflicts of Interest Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or, devices in the article. All authors have registered on-line self-reported COI Disclosure Statement Forms through the website for the Japan Neurosurgical Society members.

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