

# Suprasellar Hemangioblastoma Mimicking a Craniopharyngioma: Result of Extended Endoscopic Transsphenoidal Approach—Case Report

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## Abstract

Hemangioblastoma in the suprasellar region is rare. We present a case of a suprasellar hemangioblastoma that underwent surgical resection using an extended endoscopic transsphenoidal approach. A 64-year-old female patient presented with headache and decreased visual acuity for the last four years, computed tomography (CT) and magnetic resonance imaging (MRI) revealed a 2.5 cm irregular lesion in the suprasellar region. Our preoperative presumptive diagnosis was craniopharyngioma. The patient underwent an extended endoscopic transsphenoidal approach, the mass was subtotally removed. An endoscopic endonasal repair was needed due to the cerebrospinal fluid (CSF) leak. However, 1 month later, the patient got disturbance of consciousness because of the hydrocephalus. Ventriculoperitoneal shunt was used to solve the problem. Pathological findings were compatible with hemangioblastoma. Suprasellar hemangioblastoma is very rare. Any highly vascular lesions located in the suprasellar region should alert the surgeon to the possibility of hemangioblastoma. Extended endoscopic transsphenoidal approach adopted by us should not be the first choice of the treatment procedure for this kind of large and vascular tumor.

Key words: hemangioblastoma, suprasellar, endoscopic endonasal approach

## Introduction

Hemangioblastoma is currently classified as a benign tumor of uncertain histogenesis. It occurs predominantly in the cerebellar hemisphere, spinal cord, and medulla oblongata of adults between the age of 40 and 60.<sup>1)</sup> However, hemangioblastoma in the suprasellar region is very rare, information on them is merely limited to a small number of case reports. As endoscopic techniques evolve, a few neurosurgeons have tried to use endoscopic endonasal approaches to handle lesions in the suprasellar region.<sup>8)</sup> We now present a case of a suprasellar hemangioblastoma that underwent surgical resection through an extended endoscopic transsphenoidal approach.

## Case Report

### Clinical and imaging evaluation

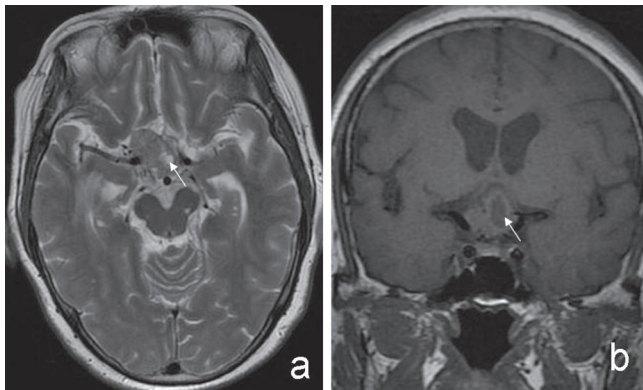
A 64-year-old female presented with headache and decreased visual acuity for the past 4 years. She described

these headaches as dull. Ophthalmologic examination revealed 0.6 oculus dexter (OD), finger counting oculus sinister (OS) and bi-temporal field defect. The patient's endocrinological examinations were within normal limits (Table 1). The computed tomography (CT) and magnetic resonance imaging (MRI) examinations revealed a solid suprasellar mass of 2.5 cm diameter (Fig. 1a, b). The mass was isointense with gray matter on T<sub>1</sub>-weighted images and hyperintense on T<sub>2</sub>-weighted images. Curvilinear areas of flow void were evident on both sequences. And strong enhancement was observed after intravenous (i.v.) administration of Gd-DTPA (gadolinium diethylene triamine pentacetate) (Fig. 2a). The tumor was shown to contain a "cystic" component (Fig. 1a, b). It was hypointense on T<sub>1</sub>-weighted images and hyperintense on T<sub>2</sub>-weighted images. The presumptive diagnosis was craniopharyngioma. After enhancement, the "cystic" component was more prominent than solid tumor on 3D-MPRAGE MR (three dimensional magnetization prepared rapid acquisition with gradient echo sequence magnetic resonance) (Fig. 3a). In our department, 3D-FIESTA (three dimensional fast-imaging employing steady acquisition sequence)

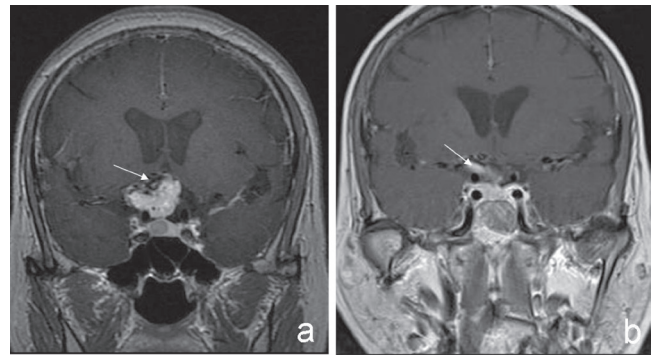
**Table 1** Pre- and post-operative endocrinological data of the patient

Hormone (normal value)	On admission	Follow-up (1 year)
FT3 (2.8–7.1 pmol/l)	3.17	4.6
FT4 (12.0–22.0 pmol/l)	23.2	21.8
TSH (0.27–4.2 uIU/ml)	2.91	3.22
PRL (131–649 mIU/l)	156.9	78.3
GH (<10 ng/ml)	< 0.05	< 0.05
ACTH (7.2–63.3 pg/ml)	47.0	60.0
Cortisol (7–10 am, 171–536 nmol/l)	472.0	116.6
FSH (25.8–134.8 mIU/ml)	16.7	29.1
LH (7.7–58.5 mIU/ml)	5.2	7.8

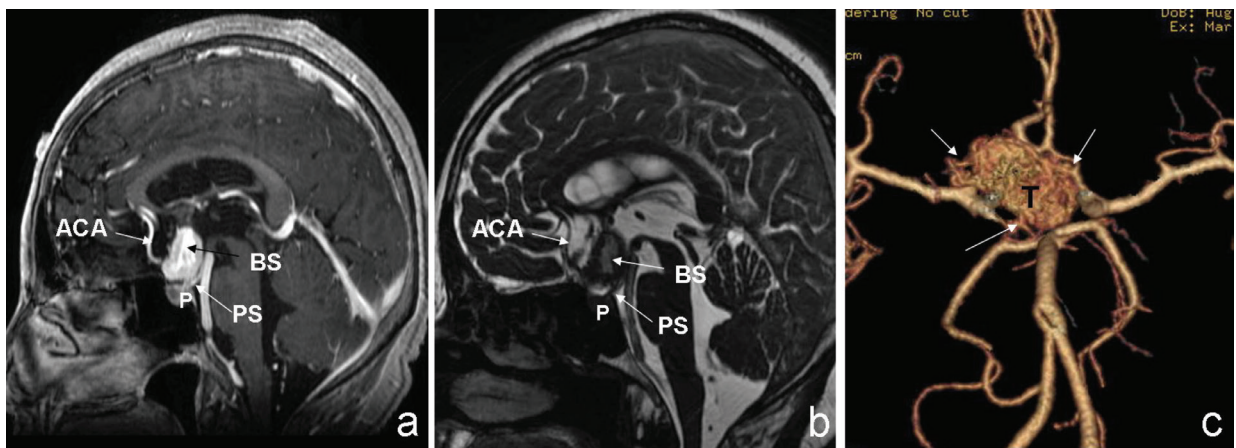
ACTH: adrenocorticotropic hormone, FSH: follicle-stimulating hormone, FT3: free T3, FT4: free T4, GH: growth hormone, LH: luteinizing hormone, PRL: prolactin, TSH: thyroid-stimulating hormone.



**Fig. 1** a, b: Preoperative nonenhanced images reveal a suprasellar mass. The “cyst” (arrows) is hypointense on T<sub>1</sub>-weighted image and hyperintense on T<sub>2</sub>-weighted image.



**Fig. 2** a, b: The preoperative and postoperative images with contrast. a: Showing a markedly enhancing suprasellar mass. The arrow notes the hypointense area of flow voids. b: Showing the small piece of tumor remained (arrow).



**Fig. 3** a: The 3D-MPRAGE MR image showed the blood sinus in the tumor with stronger enhancement. Both 3D-MPRAGE MR and 3D-FIESTA MR, b: revealed the small vital structures around the tumor, c: CTA image demonstrates blood supply by multiple small vessels (arrows). ACA: anterior cerebral artery, BS: blood sinus, CTA: computed tomography angiography, P: pituitary, PS: pituitary stalk, T: tumor, 3D-FIESTA MR: three dimensional fast-imaging employing steady acquisition sequence magnetic resonance, 3D-MPRAGE MR: three dimensional magnetization prepared rapid acquisition with gradient echo sequence magnetic resonance.

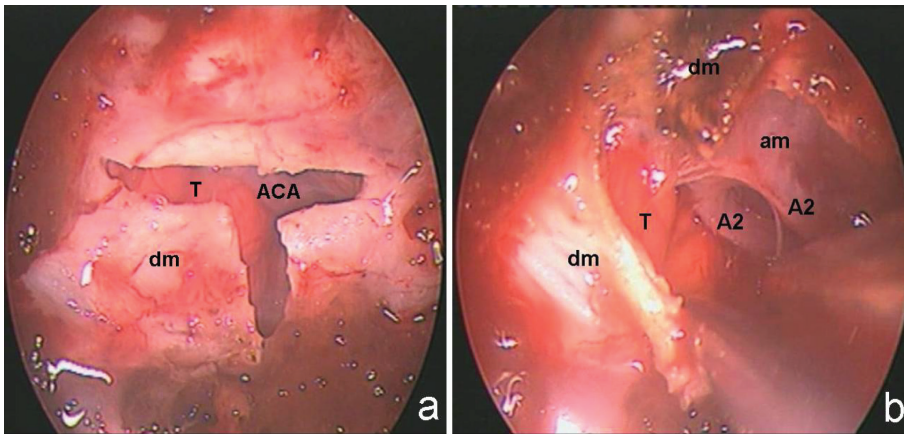


Fig. 4 Pictures during the operation. a: After we open the dural matter, the tumor was found through the semitransparent arachnoid membrane. b: Dissecting the tumor from the complex of anterior cerebral artery. ACA: anterior cerebral artery, A2: segment of the anterior cerebral artery, dm: dural matter, am: arachnoid membrane, T: tumor.

and 3D-MPRAGE were used to evaluate the suprasellar mass and detect vital small structures in and around the tumor.<sup>19)</sup> The anterior cerebral artery (ACA) could be found in front of the tumor, while the pituitary stalk was behind the tumor (Fig. 3b). Computed tomography angiography (CTA) demonstrated blood supply to the mass from multiple small perforating arteries arising from the ACA and posterior communicating artery (Fig. 3c).

#### Operation and postoperative course

Under a preoperative diagnosis of craniopharyngioma, a standard, expanded endoscopic endonasal opening<sup>8)</sup> was performed. The upper half of sellar floor and tuberculum sellae were opened. The dural matter was cut in “T” shape and coagulated at the bilateral edges (Fig. 4a). After opening the arachnoid membrane, we observed the ACA complex on the tip of the tumor (Fig. 4b). The pituitary stalk and chiasma were pushed behind the tumor. After meticulous extracapsular dissection, an easily bleeding reddish mass with a smooth surface was removed piece by piece. During the resection, the bleeding was aspirated by the angled suction to make a clear vision. Blood loss totaled 600 ml. A small piece of the tumor was retained because of the adherence to the surrounding artery.

Histopathologically, the tumor had numerous capillary vessels, each composed of a single layer of endothelial cells without nuclear heteromorphism and characteristically lipid-filled stromal cells (Fig. 5). Immunostaining revealed that the tumor was positively immunoreactive for the endothelium-specific markers CD31 and CD34. The histological diagnosis was hemangioblastoma.

Postoperatively, endoscopic endonasal repair was done because of the cerebrospinal fluid (CSF) leak. After operation her visual improved to 0.8 OD and 0.2 OS. However, the patient got disturbance of consciousness 1 month after the operation. CT showed communicating hydrocephalus. Ventriculoperitoneal shunt was

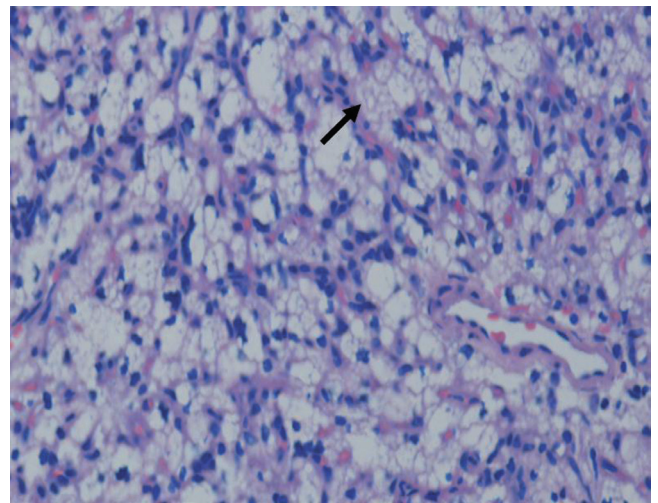


Fig. 5 Histopathology of the resected specimen shows thin-walled vessels and lipid-filled stromal cells (arrow) (H and E  $\times 200$ ).

used to solve the problem. MRI during the follow-up period demonstrated subtotal removal of the tumor (Fig. 2b). She was advised to undergo radiotherapy.

#### Discussion

Hemangioblastoma is benign, highly vascular neoplasm that occurs most commonly in the posterior fossa.<sup>1)</sup> Suprasellar hemangioblastomas are very rare; only 17 cases have been reported to date (Table 2). Our patient in this case did not present with Von Hippel-Lindau disease.

In the previously documented cases in the literature, the most common presenting complaint was visual disturbance.<sup>14)</sup> The other complaints were endocrinological disturbance, including amenorrhoea, galactorrhoea, and diabetes insipidus.

**Table 2** Previous published cases of suprasellar haemangioblastomas

Case	Series	Clinical presentations	Angio	Tumor size (mm)	Treatment
1	O'Reilly et al. (1981) <sup>13)</sup>	Vertigo, vomiting, ataxia, nystagmus	Supraclinoid nodule	NS	Craniotomy, total excision
2	Grisoli et al. (1984) <sup>5)</sup>	Galactorrhoea	Highly vascular lesion	NS	Craniotomy, total excision
3	Neumann et al. (1989) <sup>11)</sup>	Amenorrhoea,	NS	NS	Craniotomy
4	Niemelä et al. (1996) <sup>12)</sup>	Hemianopsia, SIADH?	NS	24	GK radiosurgery
5	Sawin et al. (1996) <sup>16)</sup>	Visual loss, headache	NS	NS	Transphenoidal biopsy, craniotomy and subtotal excision, radiotherapy
6	Kachhara et al. (1998) <sup>7)</sup>	Diplopia, left abducens palsy	NS	NS	Craniotomy, subtotal resection
7	Kouri et al. (2000) <sup>9)</sup>	Amenorrhoea, polydipsia, panhypopituitarism	NS	12	Transphenoidal approach, total resection
8	Kouri et al. (2000) <sup>9)</sup>	Asymptomatic	NS	14	Transphenoidal approach, total resection
9	Goto et al. (2001) <sup>4)</sup>	Irregular menstruation	Tumor blush	10	Craniotomy, total excision
10	Ikeda et al. (2001) <sup>14)</sup>	Visual disturbance	Tumor blush	NS	Craniotomy, total excision
11	Rumboldt et al. (2003) <sup>15)</sup>	Visual loss, bitemporal hemianopsia panhypopituitarism	Intratumoral vessels	40	Transphenoidal biopsy
12	Wasenko and Rodziewicz (2003) <sup>18)</sup>	Memory disturbances, oligomenorrhoea	Supply from distal, internal carotid arteries and thalamoperforating arteries	32	Subtotal excision with craniotomy, GK radiosurgery
13	Peker et al. (2005) <sup>14)</sup>	Visual loss	NS	NS	Craniotomy, total excision
14	Peker et al. (2005) <sup>14)</sup>	Visual loss	NS	NS	Craniotomy, subtotal excision
15	Fomekong et al. (2007) <sup>2)</sup>	Blurred vision	NS	20	Craniotomy, total excision
16	Miyata et al. (2008) <sup>17)</sup>	General fatigue, loss of volition, and decreased vision	Supply from the left superior hypophyseal artery	35	Craniotomy
17	Cao et al. (2010) <sup>20)</sup>	Galactorrhea and alopecia	NS	4	Oral bromocriptine

Angio: angiogram, GK: gamma-knife, NS: not stated, SIADH: syndrome of inappropriate antidiuretic hormone secretion.

The CT and MRI showed the suprasellar mass, which demonstrated intense enhancement with contrast material. And it was thought to be containing a small cyst. All clues favored the diagnosis of craniopharyngioma. However, 3D-FIESTA, 3D-MPRAGE MR, and CTA supplied much more information. The cyst should be the blood sinus formed by hemangioblastoma. And flow voids around the tumor were more clearly on 3D-FIESTA MR. CTA revealed that the mass was hypervascular supplied by multiple small blood vessels.

Surgical resection is the first line of treatment for the hemangioblastoma. To our knowledge, in the 17 cases (Table 2) previously described in the literatures, two of them were diagnosed only at autopsy, one underwent radiosurgery, one chose medical treatment, and 13 cases underwent surgical resection. Among the 13 cases, only two were selected for modified transsphenoidal approach and craniotomy was performed in the other 11 cases. In the cases with modified transsphenoidal approach, the tumors' volumes were limited (about 1 cm). As

endoscopic techniques evolve, lots of neurosurgeons are keen on the extended endoscopic endonasal approach to the suprasellar region. The handled lesions contained craniopharyngioma and meningioma. Although a few neurosurgeons have tried to use endoscopic endonasal approaches to treat vascular lesions in the sellar region,<sup>3)</sup> we think that such cases should proceed with caution, and suprasellar hemangioblastoma with large volume should not be handled by the endonasal approach. In our case, the significant arterial haemorrhage (600 ml) occurred during the removal. This may have eventually led to post-operative communicating hydrocephalus. We resisted removing the small piece of the tumor which was intimately related to the surrounding artery. The residual piece was located lateral to the right ICA and was difficult to expose. We recommend radiotherapy to her.<sup>11,16)</sup>

## Conclusion

Suprasellar hemangioblastoma is very rare. Any highly

vascular lesions located in the suprasellar region should alert the surgeon to the possibility of hemangioblastoma. An extended endoscopic transsphenoidal approach for large suprasellar hemangioblastoma is not the first choice of the treatment procedure.

### Conflicts of Interest Disclosure

The authors have no personal, financial or institutional interest in any of the drugs, materials, or devices in the article.

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