

# Pituitary Adenoma Accompanying Sphenoidal Meningoencephalocele with Clival Extension

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Encephaloceles are known as rare craniofacial developmental anomalies which are herniation of cranial contents through a skull defect. Sphenoidal meningoencephalocele represents an important cause of reversible visual loss. We report a rare case of pituitary adenoma accompanying sphenoidal meningoencephalocele with clival extension. Although the definitive diagnosis of cystic lesions in the sellar region before surgery is difficult, accurate diagnosis of these lesions is important to determine the type of treatment and predict prognostic outcome.

**KEY WORDS :** Sphenoidal meningoencephalocele · Pituitary adenoma · Craniofacial abnormalities.

## Introduction

Sphenoidal meningoencephalocele is a congenital anomaly that consists of herniation through a sphenoidal bony defect of an ependyma-lined sac filled with cerebrospinal fluid (CSF) and containing neurovascular tissue, reported only in 14 adult patients<sup>10,13</sup>. When the herniation consists of brain and meninges, it is called a meningoencephalocele; the presence of communication with ventricular portion characterizes a meningoencephalocystocele, while the absence of brain tissue is defined as a meningocele<sup>2,8,10</sup>.

We report a rare case of pituitary adenoma accompanying

sphenoidal meningoencephalocele with clival extension that is an important cause of reversible visual loss.

## Case Report

A 57-year-old man presented with headache, limitation of extraocular motion, and visual disturbance for 6 months. On neurological and ophthalmologic examination, there were a left abducens nerve palsy and left upper visual field defect. Preoperative pituitary hormonal laboratory findings revealed that cortisol level (2.7 ug/dl) was decreased mildly and other hormone level was normal range. A brain magnetic resonance (MR)



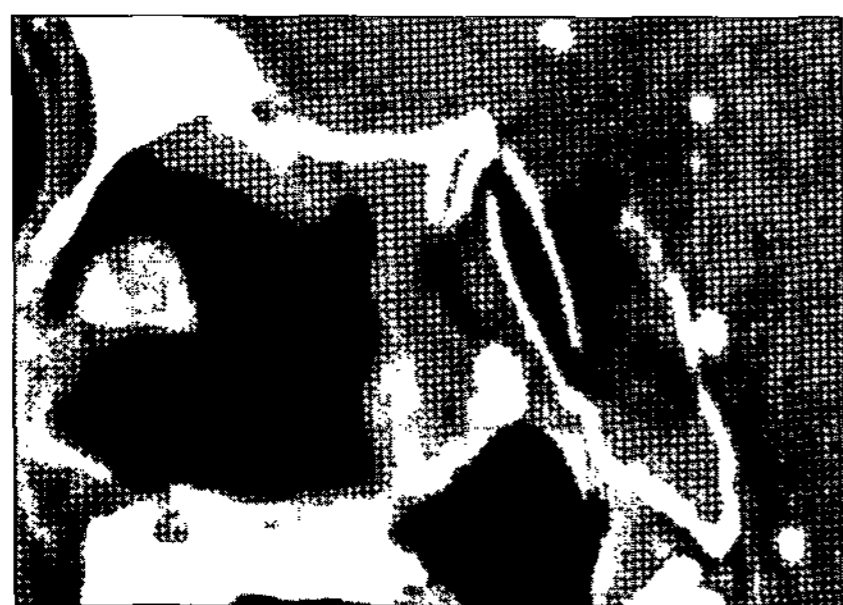
**Fig. 1.** Preoperative sagittal brain magnetic resonance (MR) images. A : T1-weighted MR image showing a sellar mass with hypointense cystic lesion. B : T2-weighted MR image showing a sellar mass with hyperintensity and sphenoidal extension. C : Gadolinium-enhanced MR image showing sellar tumor and cystic extension with neural content in the sphenoid sinus (arrow).

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**Fig. 2.** Intraoperative photograph demonstrating the cyst wall after dural opening by transsphenoidal approach.



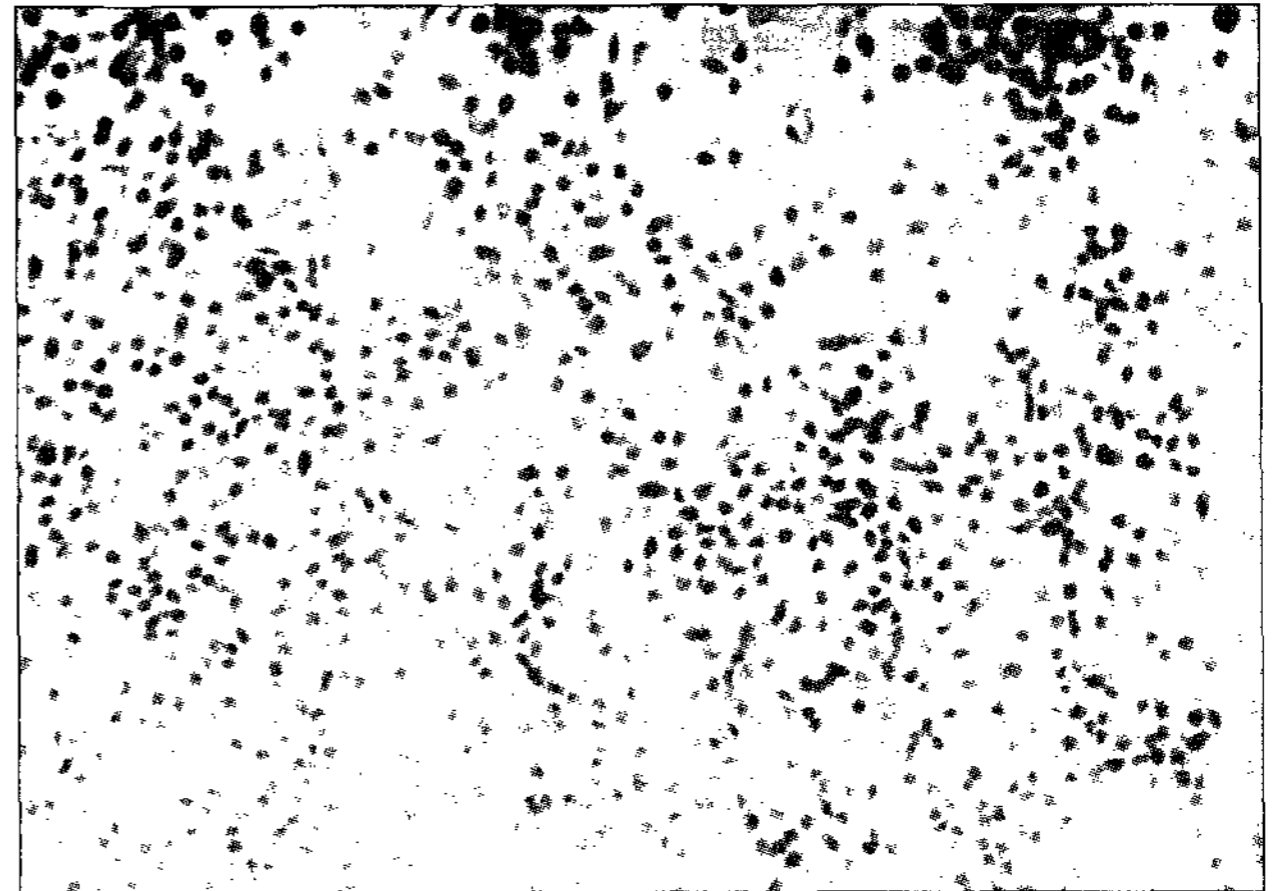
**Fig. 3.** Postoperative sagittal enhanced computed tomography scan showing total removal of sellar mass and well reconstruction of the sellar floor.

images showed a sellar mass with cystic portion, extended to sphenoid sinus and clivus. The cystic mass revealed different signal intensities, which were hypointense on T1-weighted image (WI) and hyperintense on T2WI (Fig. 1).

The patient underwent total removal of pituitary mass with cystic portion via transseptal, transsphenoidal approach. On intraoperative findings, the content of tumor mass was a mixture of hemorrhage and the cyst was covered by dura (Fig. 2). After removal of sellar mass with cystic portion, bony defect in the sphenoid sinus was reconstructed with fat graft and silicone plate. On postoperative neurological examination, visual disturbance and diplopia were improved and postoperative courses were uneventful. Postoperative sellar enhanced computed tomography scans showed total removal of sellar and cystic mass with the reconstructed bony defect in the sphenoid sinus (Fig. 3). Pathological examination of surgical specimen showed typical findings of pituitary adenoma (Fig. 4).

## Discussion

Cephaloceles occur in approximately one of every 3000 to 5000 live births<sup>2,7</sup>. There is no difference of incidence between males and females and one-third of patients have an associated anomalies<sup>1</sup>. Suwanwela et al.<sup>11</sup> proposed a classifi-



**Fig. 4.** Photomicrograph of the surgical specimen demonstrating typical findings of pituitary adenoma (H & E, original magnification  $\times 400$ ).

cation into frontal, occipital, parietal, and basal encephalocele. Basal meningoencephaloceles, which occur with an estimated incidence of one in every 35,000 live births have been further subdivided, depending on the location of the bone defect, into transthemoidal, sphenoidal, sphenoorbital, and transsphenoidal type<sup>7,11</sup>. Sphenoidal meningoencephalocele may encompass the third ventricle, hypothalamic-pituitary elements, anterior cerebral arteries, and optic apparatus<sup>10,13</sup>.

Pathogenesis of basal encephalocele have been controversial. Developments of this lesion might be caused by failure of ossification centers to fuse, leading to herniation of meninges and neural tissues<sup>11</sup>, and persistence of craniopharyngeal canal, following downward displacement of intracranial contents<sup>8</sup>. The majority of transsphenoidal meningoencephaloceles were diagnosed during the first year of life due to manifestations such as respiratory distress caused by epipharyngeal obstruction, cranial midline defects with cleft lip or cleft palate, hypertelorism, optic malformations with anophthalmia, retinal abnormalities, optic nerve hypoplasia, or unexplained bouts of recurrent meningitis<sup>3,5,6,10,12,13</sup>.

Headache is the most common clinical symptom and progressive visual disturbance, bilateral hemianopsia, and pituitary hypofunction are shown when the mass was involved in parasellar area<sup>4</sup>. The clinical presentation of a patients with sphenoidal encephalocele is in part dependent on age. With characteristic facial anomalies, the diagnosis is usually made in infancy or early childhood<sup>10,13</sup>. Respiratory difficulties due to epipharyngeal obstruction may lead to the prompt evaluation in most cases. Presentations in adults are rare, as congenital facial anomalies as subtle or absent and diagnosis may be delayed until CSF rhinorrhea occurs, visual field defect become evident, hormonal deficiency is noted, or an epipharyngeal mass is discovered. This case was also diagnosed by the visual disturbance. Hypothalamic-pituitary dysfunction was often

found in patient with transsphenoidal encephalocele. Morioka et al.<sup>9)</sup> reviewed 15 reported patients of transsphenoidal encephalocele with hormonal deficiency, and described that the deficiency of growth hormone and antidiuretic hormone was most frequently found.

Surgical indications and approaches for sphenoidal encephaloceles have been remained controversial<sup>10)</sup>. Surgical intervention is indicated for rhinorrhea or meningitis, and for progressive visual defects ascribed to the lesion<sup>10,13)</sup>. Although transsphenoidal encephaloceles have been treated by the transcranial or the transpalatal or transsphenoidal route, the optimal mode of treatment has not yet been established. MR imaging provides the most accurate road map for these delicate interventions. The transcranial approach was associated with high postoperative rates of morbidity, mortality, and hypothalamic dysfunction. In the transpalatal approach, the closure of the mucosal layer and the reconstruction of the skull base were also difficult. In our case, the transsphenoidal approach was used and skull base was reconstructed with silicone plate.

## Conclusion

A case of pituitary adenoma accompanying sphenoidal meningoencephalocele is extremely rare and we should consider this entity to diagnosis the sellar tumor with cystic extension. Meticulous reconstruction of skull base is also important to prevent CSF rhinorrhea.

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