CASE REPORT

Intracranial Approach to Suprasellar Rathke's Cleft Cyst

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ABSTRACT—The occurrence of a symptomatic Rathke's cleft cyst without extension into the sella turcica is rare. The purpose of this report is to present such a case, with its clinical manifestation, diagnostic imaging characteristics, operative approach, pathology, and review of the literature.

Rathke's cleft cysts are epithelial cysts believed to be derived from remnants of Rathke's pouch. These cysts are incidentally found in 2 to 26% of routine autopsy series.^{1,2} They are small, asymptomatic, and usually located in the pars intermedia or distalis.

The symptomatic cysts are larger and are usually intrasellar in origin.² However, a third of these cysts have a suprasellar component. They may compress the surrounding neural structures and produce symptoms such as visual disturbances, hypothalamic dysfunction, and headache. Unusual presentations include obstructive hydrocephalus, aseptic meningitis, and intracranial abscess.¹⁻⁷

This report describes a suprasellar Rathke's cleft cyst with a normal sella turcica presenting with visual disturbances and headache. This case illustrates the clinical, radiological and pathological characteristics of an exclusively suprasellar Rathke's cleft cyst. The surgical treatment of this uncommon lesion is presented.

eye over her temporal field. She subsequently noted cephalgia. Two days later, she awakened with transient blurring of the left eye with predominant involvement of her temporal field. She developed transient nausea and vomiting. She was seen by her primary physician. A computed tomography (CT) scan of the head revealed a suprasellar mass measuring approximately 12 mm in diameter associated with upward displacement and compression of the optic chiasm (Fig. 1). A subsequent magnetic resonance imaging (MRI) scan exhibited a 10-mm anterior suprasellar mass with minimal rim enhancement and mild compression of the optic chiasm (Fig. 2). Her general physical, neurological, and ophthalmological examinations were essentially within normal limits. In view of the patient's age, as well as the location and size of the lesion, operative intervention was recommended to the patient after discussing the indications, alternatives, possible benefits, realistic expectations, and risks of surgery. She concurred with proceeding with surgery.

CASE REPORT

A twenty-nine-year-old, right-handed female noted an episode of ocular-like migraine headache of 5 hr duration. Initially, she noted transient blurring of the left

OPERATION

A right frontal craniotomy was performed. The anterior limit of the bone flap included the superior orbital

Figure 1. Coronal CT scan of the head demonstrating suprasellar location of a mass without contrast enhancement.

rim. A subfrontal, intradural approach disclosed a suprasellar, cystic mass compressing the optic chiasm. Anteriorly, it was located between the optic nerves, without adherence. The capsule of the tumor was incised on its dorsal aspect. Thick, white mucinous fluid was removed using suction and micro curettes. The thin fibrous capsule was also removed using suction and biopsy forceps. The posterior and right lateral portions of the cyst wall were firmly attached to the infundibular stalk of the pituitary and were not removed. The operation was terminated once an adequate decompression of the surrounding structures was achieved and tissue for histologic confirmation was sent to pathology. Radical removal of the remaining residual tissue of the lesion

was not attempted. An intraoperative frozen section of the fluid and part of the tumor was consistent with the diagnosis of a Rathke's cleft cyst.

POSTOPERATIVE COURSE

The patient's postoperative course was denoted by transient diabetes insipidus, which responded readily to the administration of desmopressin acetate (DDAVP). Otherwise, her clinical course was entirely benign with resolution of her visual symptomatology and complaints of headache. A follow up MR scan reviewed postoperative changes and an intact infundibulum (Fig. 3).

PATHOLOGY

The surgical specimen consisted of a cyst wall characterized by loose fibrous tissue and lined by a single layer of columnar epithelial cells. Ciliated cells were recognized. Sections of the cyst contents showed acellular mucoid material (Fig. 4).

DISCUSSION

Symptomatic Rathke's cleft cysts are a rare clinical occurrence.^{1,2} Rathke's cleft cysts are believed to be derived from the remnants of the Rathke's pouch.⁸ The lumen between the anterior and intermediate portion of Rathke's cleft is obliterated by epithelial infoldings in most pituitary glands. If this lumen is not obliterated, a

Figure 2. (A) MRI scan coronal view of suprasellar mass with mild rim enhancement, cystic appearance, and compression of optic chiasm. (B) Sagittal view of lesion. Note suprasellar location without intrasellar extension.

Figure 3. Postoperative MR scan of the head confirming removal of suprasellar Rathke's cleft cyst. Increased signal of the infundibulum represents postoperative inflammatory changes.

Rathke's cleft cyst lined with columnar or cuboidal epithelial cells may develop.^{1,2,8,9,12} They are common incidental findings at autopsy, varying between 2 to 26% in reported series.⁸ Rarely, they may cause symptoms. When they do, it is because of significant size with resultant compression of adjacent neural structures.

Most of the Rathke's cleft cysts reported in the literature are intrasellar, with or without a suprasellar component. Entirely suprasellar Rathke's cleft cysts are unusual. Only 11 such instances have previously been reported in the literature. The case presented in this paper represents an additional, entirely suprasellar, cyst with a normal sella turcica.^{2,4,9} Preoperative endocrine function tests were normal. In view of these features, an intracranial, rather than transsphenoidal, operative approach was chosen.

The age distribution of reported symptomatic Rathke's cleft cysts, range from 4 to 72 years. Most of

Figure 4. Light microscopy H & E stained pathology specimen. Note single layer of columnar epithelial cells, some of which are ciliated.

the cases occur between 40 and 60 years of age.⁸ There appears to be no significant sexual predilection.^{2,8} The clinical presentation is usually generalized pituitary hypofunction with or without multiple endocrinopathies, visual disturbances, and/or headaches. Hypothalamic dysfunction, hydrocephalus, aseptic meningitis, and/or abscess formation have also been reported.¹⁻¹⁰

Rathke's cleft cysts are commonly located in the anterior and superior portion of the sella turcica or suprasellar cistern. CT and MRI scans reveal a well-defined margin, located in the midline, either in the anterior sella turcica or the anterior suprasellar cistern, or both.^{1,9,11,12}

The treatment of symptomatic cysts is surgical. A frontal craniotomy with a subfrontal intradural approach was performed in this case due to the purely suprasellar location of the cyst.^{1,2,6} Treatment should include full evacuation of the inner contents of the cyst. Removal of as much of the cyst wall as much as possible should be made.^{1,2,6} If an attempt toward radical removal of an adherent cyst wall endangers the hypothalamus, pituitary stalk, or optic chiasm, subtotal removal is preferable.¹ Radiation therapy has no role in the treatment of these benign lesions.¹ Recurrence of a Rathke's cleft cyst after drainage and partial resection has been reported, but is uncommon.^{5,13}

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