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Symptomatic Rathke's Cleft Cyst (A Case Report)

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=Abstract=The first Korean case of a symptomatic Rathke's cleft cyst in a 14-year old boy is described. His chief complaint was headache of 2 months' duration and he had diabetes insipidus, hypopituitarism and decreased visual acuity on both sides. The computerized tomography scannng revealed an isodense small round mass at the suprasellar cistern with partial enhancement. The cyst was removed totally at the sacrifice of the pituitary stalk. The clinical, radiological and pathological findings are discussed with review of the literature.

Key words: ***Rathke's cleft cyst, Computerized tomography***

INTRODUCTION

Rathke's cleft cyst is a rare epithelial cyst which is confined to the sella turcica or extends upward into the supraseller area. There are controversies about the embryogenesis (Yoshida *et al.* 1977; Evans *et al.* 1979; Palma and Celli 1983; Okamoto *et al.* 1985), but it is widely believed that the cyst is derived from the remnant of Rathke's pouch. Small incidental Rathke's cleft cyst is found in 13% to 22% of normal pituitary gland between the pars distalis and the infundibular process at routine autopsies.

It produces symptoms by compression of the surrounding structures, most frequently the pituitary gland, hypothalamus, and the optic nerves and chiasm.

A case of Rathke's cleft cyst is described, and its clinical, radiological, pathological, and surgical aspects are discussed.

CASE REPORT

This 14-year old boy was admitted complaining of headachde of 2 months' duration. Ten months before the admission, he began to have polydipsia with polyuria subsequently controlled by the nasal spray of DDAVP (1-deamino-8-D-arginine vasopressin). Two months before admission, headache developed and he was admitted after brain computerized tomography (CT) scanning with the impression of germ cell tumor. Neurological examination on admission revealed visual acuity of 0.2/0.4, and bitemporal mild depression in visual fields. Hormonal studies showed mild depression of adrenal and thyroid functions, low growth hormone level and mild elevation of prolactin level. Serum α-fetoprotein and β-human chorionic gonadotrophin levels were within normal limits. Plain skull lateral X-ray showed slight ballooning of the sella (Fig.1). Brain CT checked 2 months before admission revealed an isodense small round mass at the suprasellar cistern with partial enhancement (Fig.2 & 3). On coronal CT scan, the mass was located in the sellar and suprasellar area (Fig.4).

Right frontotemporal craniotomy was done. A fusiform dilated cystic mass was seen under and behind the optic chiasm (Fig.5). Optic chiasm was not so remarkably compromised. The vascularized membranous cystic wall was composed mainly of longitudinally arranged spread vascular bundles reminiscent of pituitary stalk, and yellow or brown-green colored firm nodules in part (Fig.6). The containing cystic fluid, 2 cc in volume, was opaque brown and slightly greasy (Fig.7). Inner surface of the membranous capsule was smooth and glistening. The collapsed

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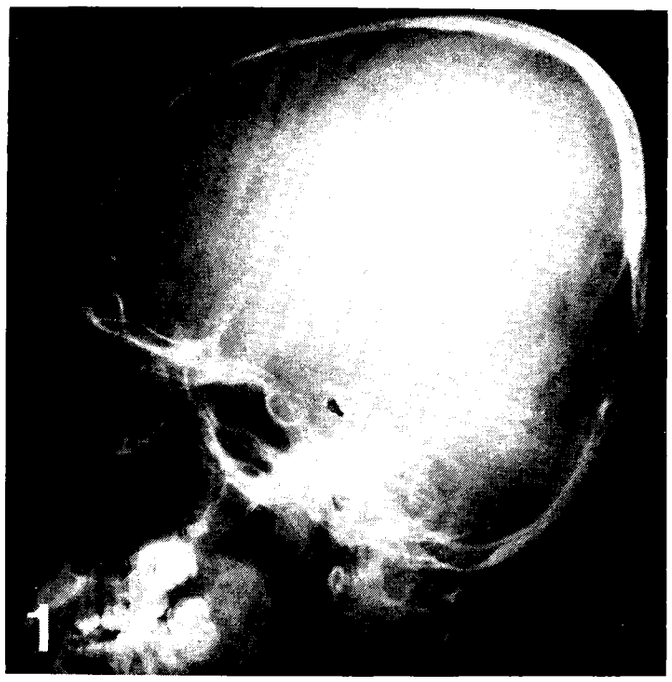
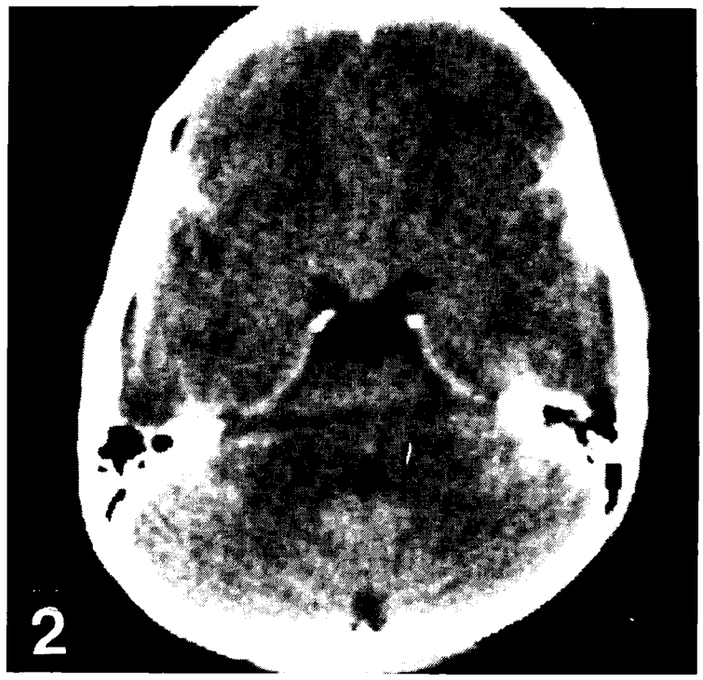
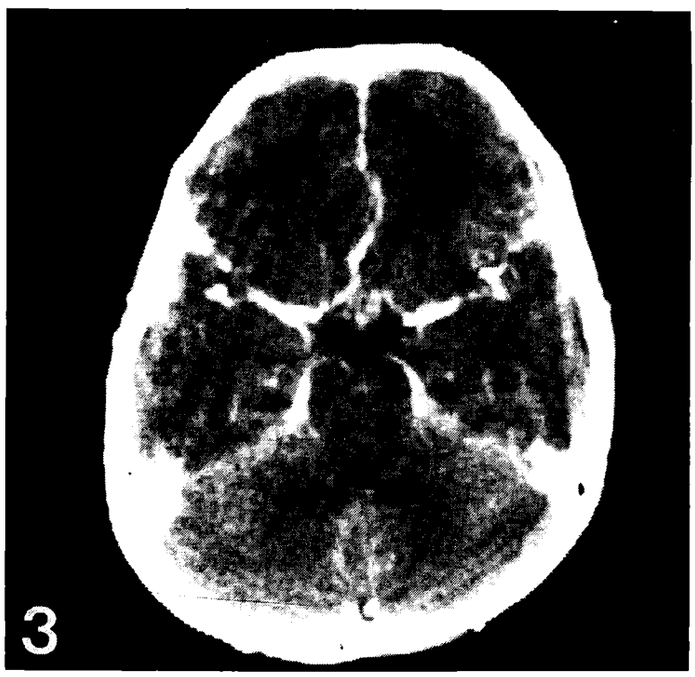






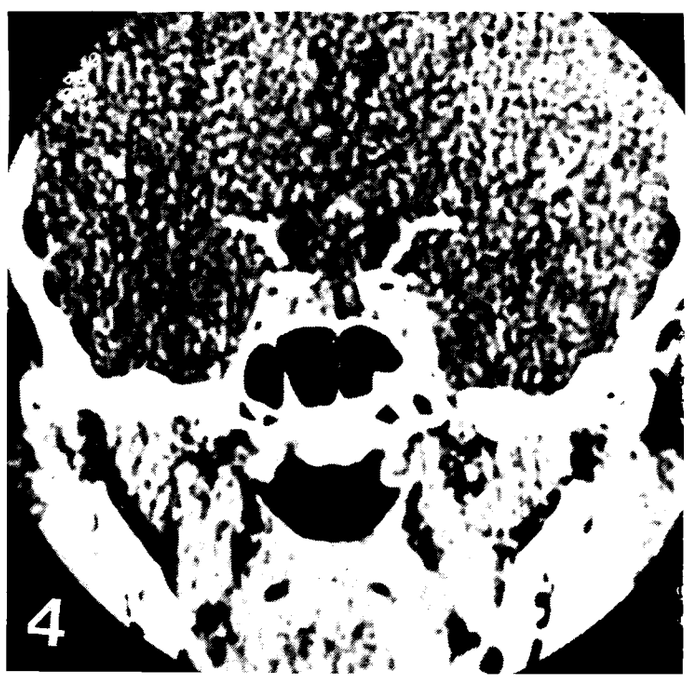


Fig. 1. Skull lateral x-ray showing slight ballooning of the sellar floor.
Fig. 2. Plain axial CT scan reveals a small isodense round mass located in the suprasellar cistern.
Fig. 3. Enhanced axial CT scan showing inhomogeneous partial enhancement.
Fig. 4. Enhanced coronal CT scan showing an intrasellar isodense, partly enhancing mass which extended into the suprasellar cistern.

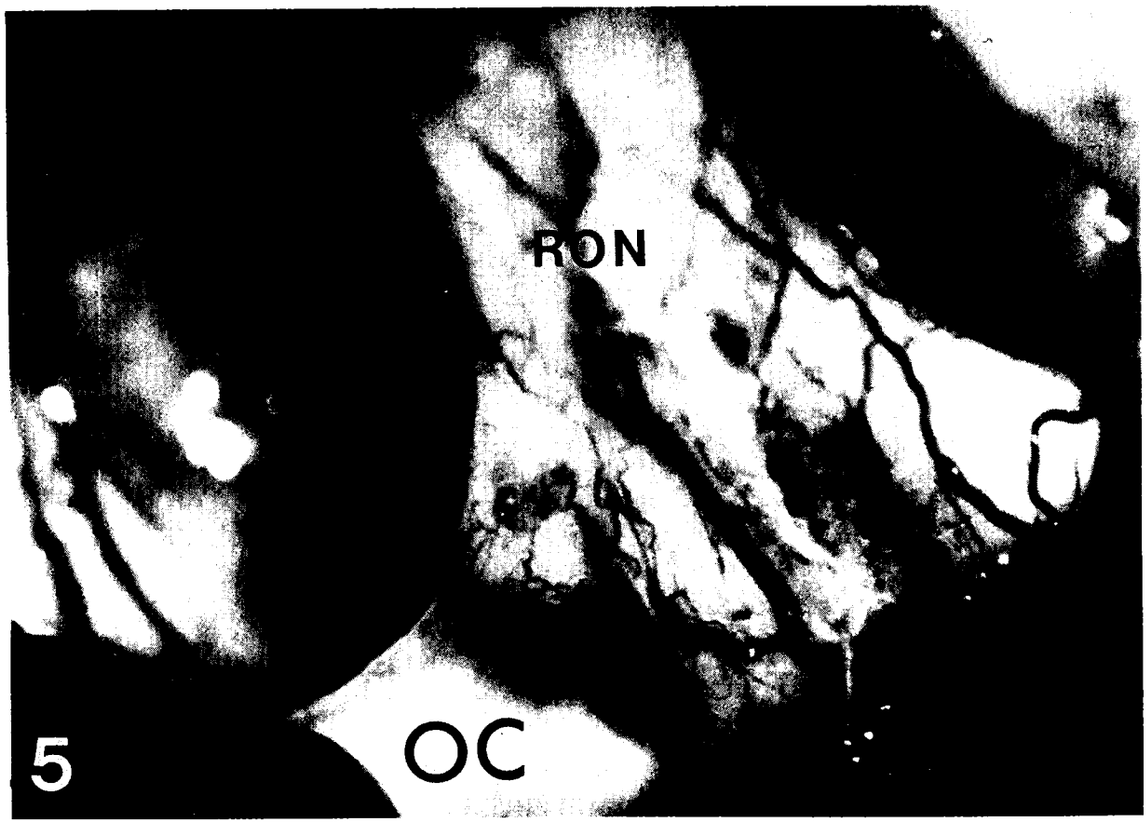
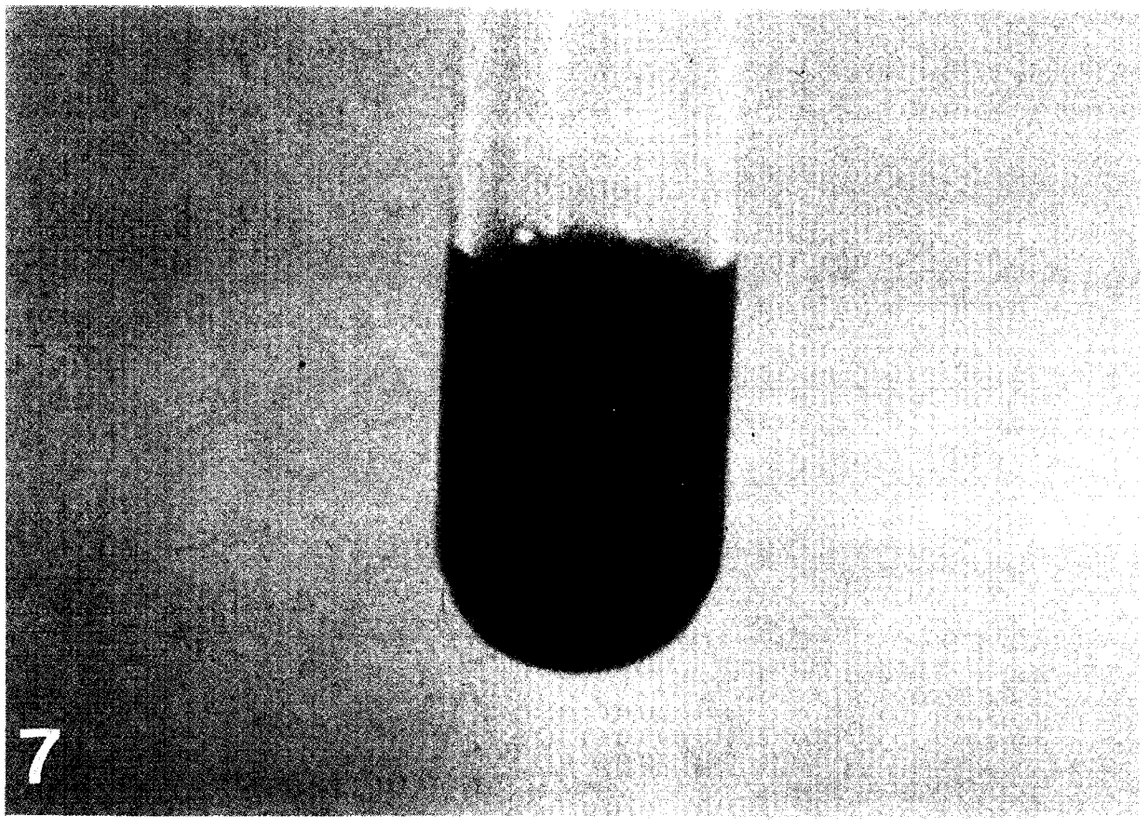


Fig. 5. Operative photomicrograph. The mass was located under the optic chiasm.
*-cyst, RON-right optic nerve, OC-optic chiasm

Fig. 6. Operative photomicrograph. The cystic mass was exposed after the retraction of the right optic nerve The surface was finely wrinkled with longitudinal vasculature.
*-cyst, RON-right optic nerve, ST-suction tip



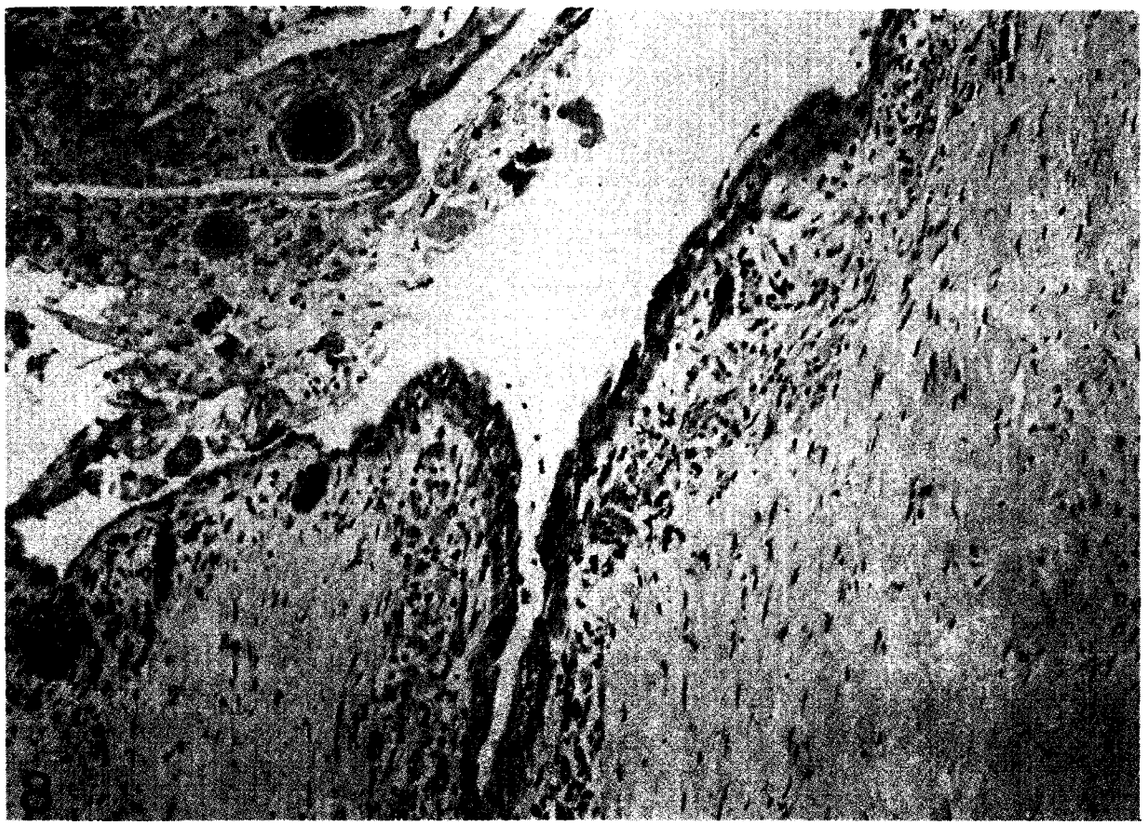


Fig. 7. Gross photograph of cystic fluid. Turbid brownish color was noted.

Fig. 8. Photomicrograph of the cyst wall consisting of fibrous connective tissue and stratified epithelium. The epithelium is focally attenuated and lies upon the inflamed collagen stroma. The lumen contains eosinophilic exudate with cholesterol clefts. H&E, ×100

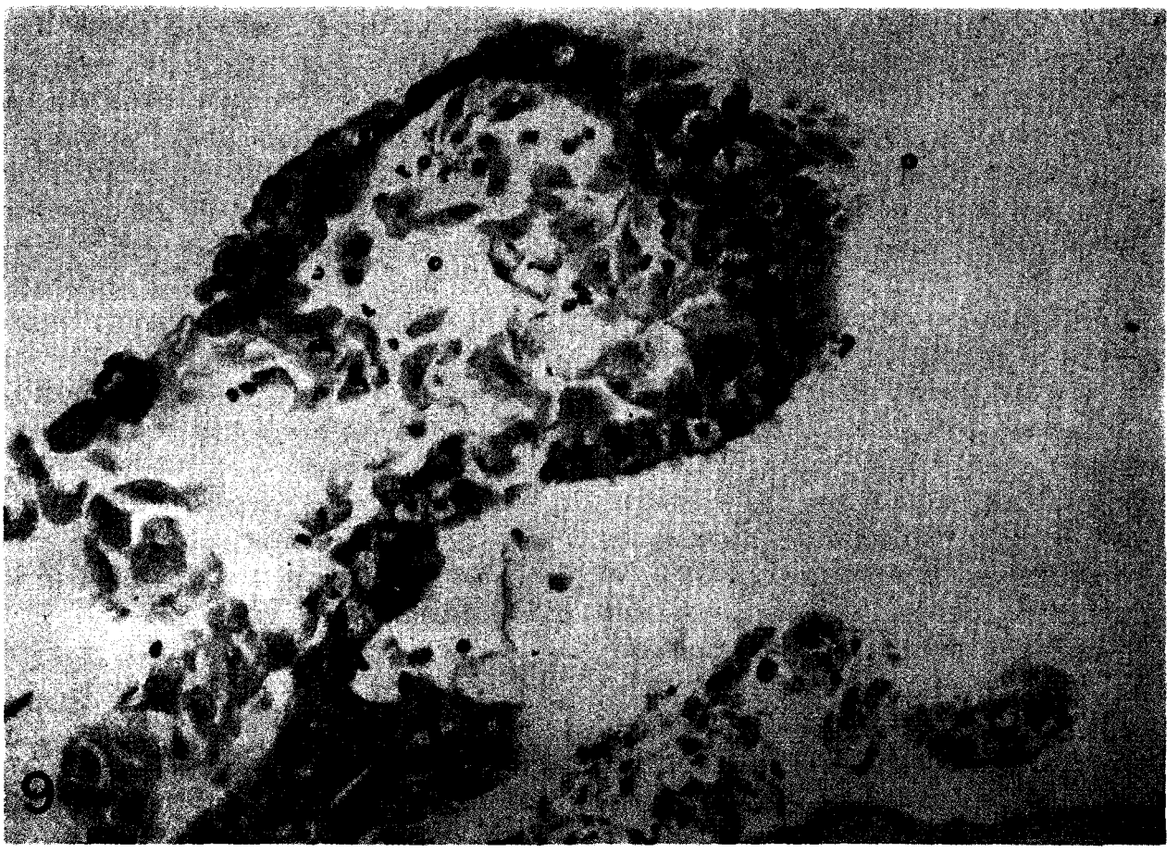


Fig. 9. Higher magnification of a portion of the cyst wall, showing pseudostratified columnar cells with occasional goblet cells. H&E, ×250

capsule was attached to the diaphragm sellae and the cavity extended into the pituitary gland. Posterior and superior end of the cavity was smoothly tapered into the pituitary stalk. Gross total removal of the cystic tumor including the capsule was done at the sacrifice of the pituitary stalk. Pathological findings showed an epithelial cyst lined by columnar and pseudostratified squamous epithelium compatible with Rathke's cleft cyst (Fig.8 & 9).

Postoperatively, the headache disappeared and the ophthalmological examination on postoperative 8th day revealed improved visual acuity (1.0/0.8) and normalized visual field. But diabetes insipidus, which was controlled with the nasal spray of DDAVP, persisted until the lastest follow-up, 12 months after the surgery. Combined pituitary function test performed 9 months after the surgery revealed panhypopituitarism with poor response to the provocation tests. The patient is on the replacement therapy of hydrocortisone, synthyroid, oxandrolone and DDAVP.

DISCUSSION

Symptomatic Rathke's cleft cyst is a rare clinical occurrence. Since the first symptomatic Rathke's cleft cyst was described by Goldzieher in 1913, fewer than 70 cases have been reported (Barrow *et al.* 1985). To our knowledge, this is the first case report of Rathke's cleft cyst in Korea.

In 1977, Yoshida *et al.* collected 34 cases including their case. According to their report, the age distribution was from 4 to 72 years and most cases occurred between 40 and 60 years of age without significant sex difference.

Rathke's cleft cyst is assumed to be a non-neoplastic remnant of Rathke's pouch, an outgrowth of the stomodeum beginning in the 4th week of gestation that eventually forms the pars distalis, pars tuberalis and the pars intermedia (Yoshida *et al.* 1977; Steinberg *et al.* 1982). They are usually intrasellar, but approximately one-third extend out of the sella turcica (Maggio *et al.* 1987), and rarely the cysts become symp-

tomatic with intrasellar location causing headache and hypopituitarism or with suprasellar extensions causing chiasmal compression, hypothalamic dysfunction, and obstructive hydrocephalus (Barrow *et al.* 1985). In present case, the cyst was located in the suprasellar area with small intrasellar extension and caused headache, mild chiasmal compression signs and hypopituitarism.

The CT appearance of a Rathke's cleft cyst in some cases is an intrasellar cystic mass, with suprasellar extension or enhancement (Martinez *et al.* 1979; Steinberg *et al.* 1982; Barrow *et al.* 1985; Okamoto *et al.* 1985; Maggio *et al.* 1987). Calcification is rarely present and the enhancement may represent inflammatory process or squamous metaplasia (Okamoto *et al.* 1985). In our case, the mass seemed nearly isodense and partly enhanced. The pathological specimen contained foci of the squamous mataplasia. Cystic pituitary adenoma, cystic craniopharyngioma, cysticercus cyst, arachnoid cyst, epidermoid cyst, and mucocele are included in the differential diagnosis (Steinberg *et al.* 1982; Barrow *et al.* 1985). Magnetic resonance imaging may provide useful information in planning the surgical approach (Iba-Zizen *et al.* 1984). However, it does not allow strict differential diagnosis from other lesions (Maggio *et al.* 1987).

Histologically, the cells lining the cyst are ciliated columnar or culboidal cells and the cyst can be lined by squamous epithelium (Russell and Rubinstein 1977; Yoshida *et al.* 1977; Kepes 1978). Rathke's cleft cyst can have solid components as pituitary adenomas (Trokoudes *et al.* 1978; Nishio *et al.* 1987), transitional cell tumors of the pituitary gland (Kepes 1978) or simply as tumor nodule (Yoshida *et al.* 1977). The fluid content of the Rathke's cleft cyst is often white mucoid, but my be clear, yellow, blue, green, thick, purulent, or even brown, like machine oil (Martinez *et al.* 1979; Yoshida *et al.* 1977). Rarely the Rathke's cleft cyst presents with recurrent aseptic meningitis as craniopharyngiomas. The cholesterol crystals, keratin and desquamated epithelial debris have been implicated as irritating substances (Patrick *et al.* 1974).

The recognition of a Rathke's cleft cyst at the time of operation and its differentiation from craniopharyngioma is very important (Eisenberg *et al.* 1976; Barrow *et al.* 1985), because the treatment and prognosis are quite different. Most reported cases have been treated by the frontal craniotomy or the transsphenoidal approach (Martinez *et al.* 1979; Baskin and Wilson 1984; Barrow *et al.* 1985). Some authors recommended aspiration of the cyst contents and only partial excision of the wall, although there are reported recurrences of Rathke's cleft cysts after less than radical removal (Raskind *et al.* 1968; Yoshida *et al.* 1977; Barrow *et al.* 1985). In present case, craniopharyngioma or teratoma could not be ruled out with the operative findings and the frozen biopsy report. So the cyst was removed totally.

Most of the symptomatic sellar and parasellar mass lesions should be operated on to make a definitive histological diagnosis and to institute proper treatment.

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=국문초록=

증상을 일으킨 Rathke씨열 낭종

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14세 남자 환아에서 증상을 일으킨 Rathke씨열 낭종 1례를 기술하였다. 한국에서는 최초로 보고되는 증례로서 2개월전부터 시작된 두통을 주소로 입원하였으며 요붕증, 뇌하수체 기능저하와 양안의 시력 장애를 보였다. 전산화단층촬영상 안상부에 부분적 대조강화를 보이는 등밀도의 작은 원형의 종괴음영을 보였다. 뇌하수체경과 함께 낭종을 완전 적출하였다.

본 질환의 임상증상, 방사선학적 및 병리학적 소견을 문헌고찰과 함께 기술하였다.