Symptomatic Rathke’s cleft cyst

Case report

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A case of a symptomatic Rathke’s cleft cyst in a 10-year-old boy is described. The importance of the intraoperative differentiation between this rare tumor and a craniopharyngioma is discussed.

Key Words • Ranchke’s cleft cyst • pituitary tumor • embryology • craniopharyngioma

Symptomatic Rathke’s cleft cysts may be confined to the sella turcica or extend upward into the suprasellar space. Like the more common cystic tumor of this region, the craniopharyngioma, they produce symptoms by compression of surrounding structures, most frequently the pituitary, hypothalamus, and optic nerves and chiasm. Although these two cystic lesions produce similar clinical pictures, their intraoperative differentiation is important because of differences in operative management. A case of Rathke’s cleft cyst is described, and its clinical, pathological, and surgical aspects are discussed.

Case Report

First Admission. This 10-year-old boy was first admitted in April, 1975, for evaluation of cessation in growth, increased thirst, and frequent urination; these symptoms had started approximately 3 years before. There were no visual problems, headaches, or symptoms of other neurological deficit. The patient had been normal except for occasional episodes of constipation. There was no family history of neurological disease. His mother and father were of normal height.

Vital signs were normal. His height was 110 cm (less than the third percentile). The general physical and neurological examinations were otherwise completely normal. Laboratory data collected during this admission revealed a normal complete blood count and serum electrolytes. The T4 was 8 μg/100 ml. Growth hormone in the resting state and after stimulation with insulin (0.5 units/kg) was decreased (highest level 2.2 ng/ml) and the blood-glucose level was depressed throughout the duration of the test (55 mg/100 ml at 45 min). A water-deprivation test demonstrated moderate impairment of urine concentrating ability (urine, 694 mOsm at 16 hrs). The bone age was 5½ to 6 years, consistent with his height. A brain scan and skull films were normal. Further work-up was planned, but the patient was taken home against medical advice.
FIG. 1. Left: Midline sagittal tomogram during pneumoencephalography. The tumor (T) outlined by gas in the suprasellar cisterns (arrowheads) is encroaching upon the interpeduncular cistern (arrow) and truncating the anterior recesses of the third ventricle (3). Right: Coronal tomogram during pneumoencephalography. The tumor is located between the internal carotid arteries (arrowheads) and the optic nerves (arrow). The lower part of the third ventricle (3) is effaced.

Second Admission. He was readmitted 2 months later. During the interval no new symptoms had developed, and his physical examination remained unchanged. Visual fields examined by perimetry showed no defects. The serum T4 on this admission, 2.8 μg/100 ml, was abnormal and significantly lower than it had been 2 months before. The T3 was 23.5%. A Hickey-Hare test demonstrated decreased urine concentrating ability during the infusion of hypertonic saline (222 mOsm after 120 min of a 3% saline infusion at 4 ml/min) and some response to exogenous pitressin (571 mOsm) suggesting diabetes insipidus of central origin. The serum cortisol level at 8 A.M. was 21 μg/100 ml, and 15 μg/100 ml at 4 P.M. Views of the optic foramina were normal. A pneumoencephalogram showed a 2-cm mass in the suprasellar region that distorted the anterior recesses of the third ventricle. The mass was not calcified (Fig. 1). Bilateral carotid arteriograms showed no definite abnormalities.

Operation. After these diagnostic studies, a right frontal craniotomy was made. A 2.5-cm dome-shaped cystic mass was exposed in the suprasellar region, between but not compressing the optic nerves (Fig. 2). Its opalescent mucinous contents were removed by suction. The capsule was dissected and removed piecemeal. It is probable that small fragments of the capsule attached to the hypothalamus were not removed. Following the operation the patient made an uneventful recovery. His

FIG. 2. Operative exposure shows the right frontal lobe retracted exposing the cyst, right optic nerve and chiasm, and the right internal carotid artery.

FIG. 3. Photomicrograph of the cyst wall shows a single layer of columnar epithelium resting on a hyalinized basement membrane. H & E, × 400.
visual fields remained intact, and his diabetes insipidus continued without change. At the time of discharge he was taking hydrocortisone, thyroid extract, and pitressin.

**Histological Examination.** The surgical specimen consisted of small nests of intact adenohypophyseal cells and a cyst wall lined in part by focally ciliated columnar epithelium (Fig. 3). No stellate cells or keratinized epithelium were present. The content of the cyst was an acellular gelatinous material that stained PAS-positive.

**Discussion**

Rathke's pouch forms during the third or fourth week of gestation as an outgrowth of the stomodeum. As the cells grow dorsally, the outgrowth elongates to form the craniopharyngeal duct. By the 11th week the proximal end of the duct obliterates, while the cranial portion comes in contact with the protrusion of the third ventricle, the infundibulum. The anterior wall of the pouch then proliferates to form the anterior lobe of the pituitary gland and pars tuberalis. The posterior wall becomes the pars intermedia. In a large percentage of human pituitaries the lumen is obliterated by epithelial infoldings. Retention of the lumen as a cleft was found by Shanklin\(^8\) in 13 of 100 autopsy specimens. In his cases the clefts were all small and asymptomatic; the largest measured 2 × 3 mm. Although formation or retention of these clefts was considered developmental, more were seen in adults than in children.

The histological appearance of the Rathke's cleft cyst is distinct from that of the craniopharyngioma. The wall consists of a single layer of cuboidal-to-columnar epithelium. It may also contain mucin-secreting cells and frequently ciliated epithelium is seen. The contents of the cyst are generally mucinous but in some cases clear or brown fluid has been found.\(^5,6,10\) The belief that these cysts are extensions or enlargements of Rathke's cleft is based on the observation that the cell lining of Rathke's cleft is similar in appearance. Shuangshoti, \textit{et al.},\(^10\) however, argued that many of these cysts may arise instead from neuroepithelial cells. In the cases they described they noted that this lesion closely resembles the colloid cyst of the third ventricle.

Craniopharyngiomas are partly cystic or microcystic epithelial tumors containing areas of stratified squamous epithelium resting on a prominent basement membrane. They are believed to originate from squamous epithelial rests presumed to be remnants of Rathke's pouch. An alternative view proposes that the cells of origin arise as a result of squamous metaplasia in cells of the adenohypophysis.\(^7\)

Histologically these two cystic lesions should then be easily distinguishable; separation on clinical grounds, however, may be difficult in some cases. Craniopharyngiomas seen in children are heavily calcified in at least 75% of cases, while in adults calcification is seen in only about half of the cases. The presence of calcification in a cystic suprasellar tumor strongly suggests that it is a craniopharyngioma, since calcification of a Rathke's cleft cyst probably does not occur.

When the lesion is not calcified, radiological differentiation is impossible, and at operation differentiation by gross inspection may be difficult. The distinction is important, however, because of differences in operative strategy. Although with new techniques radiation therapy has been found to be effective in suppressing growth of the craniopharyngioma,\(^4\) a vigorous attempt at total operative removal is generally recommended as the primary treatment for this tumor.\(^9,11\) Although this operation is frequently difficult to do without risk of injury to the hypothalamus, pituitary gland, stalk, and other surrounding structures, if it is successful, it insures a cure. Radiation therapy is then reserved for cases in which total removal could not be accomplished, or for those cases in which the tumor later regrows after what was mistakenly felt to be total removal. Cure of a Rathke's cleft cyst may also be accomplished by methods of less risk. Of the 32 cases of Rathke's cleft cyst reported in the literature, 18 have had operations\(^5,6\) and in only two cases has the cyst recurred. In both cases the initial operation consisted of drainage of the cyst and removal of only a small fragment of its wall. In one case the recurrence became symptomatic 23 years later\(^5\) and in the other after 3 years.\(^1\) Both cases responded favorably after reoperation. Fager and Carter\(^2\) reported the successful treatment of five cases of Rathke's cleft cyst, none of which had a total removal of the capsule or received radiation therapy. It is therefore recommended that in the uncalcified lesion the diagnosis of a Rathke's cleft cyst be made during operation by frozen

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section if necessary. The cyst contents and capsule are then removed, leaving any fragments of capsule that are firmly attached to vital structures or whose removal would require severe retraction of the brain. With the use of the operating microscope this procedure has little risk. Radiation therapy has no established role.

References


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