Juvenile Symptomatic Rathke’s Cleft Cyst
—Case Report—

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Abstract

A 14-year-old girl presented with a rare symptomatic Rathke’s cleft cyst manifesting as diabetes insipidus and growth retardation. Neuroimaging demonstrated the suprasellar cyst. Computed tomography showed the cyst as an isodense area with enhancement, and magnetic resonance imaging showed an hyperintense area on both T1- and T2-weighted images. Histological examination showed the cyst was consistent with Rathke’s cleft cyst. Symptomatic Rathke’s cleft cysts usually occur in middle-aged adults. Juvenile cases tend to present with diabetes insipidus, and the cyst content may include more mucopolysaccharides or hemosiderin degradation products.

Key words: Rathke’s cleft cyst, pediatric, brain neoplasms, pituitary gland, magnetic resonance imaging

Introduction

Rathke’s cleft cysts sometimes grow large enough to compress the surrounding structure, and become symptomatic. Most of the over 200 cases of symptomatic Rathke’s cleft cyst have occurred in middle-aged adults.13,17 Juvenile cases of symptomatic Rathke’s cleft cyst are rare, with only 10%17,19 of all symptomatic cases occurring in children under 16 years old.2-6,11,13-17 Clinicaloradiological studies of symptomatic Rathke’s cleft cyst have been reported in adults,1,3,5,10,13-15,17 but few in juvenile cases.1,8

We report a juvenile case of Rathke’s cleft cyst and clarify the clinical and radiographic features.

Case Report

A 14-year-old girl was admitted to Kitasato University Hospital because of growth retardation and diabetes insipidus. She stopped growing taller at the age of 10 years. Her menstruation started at the age of 12 years and occurred regularly thereafter. General physical and neurological examinations including visual functions on admission were normal. Laboratory tests of pituitary function showed thyroid stimulating hormone 2.43 μU/ml, growth hormone 0.14 ng/ml, adenocorticotropic hormone 33 pg/ml, triiodothyronine (T3) 143 ng/dl, thyroxine (T4) 7.5 μg/dl, free T3 3.2 pg/ml, free T4 1.0 ng/dl, cortisol 15.6 μg/dl, antidiuretic hormone (ADH) <1 pg/ml, and somatomedin-C 190 ng/ml (normal range 385-744 ng/ml). Suppression and stimulation tests of the hypothalamic-hypophyseal system revealed no abnormality except for mild hyporesponsiveness of growth hormone secretion. A water stress test revealed the pattern of diabetes insipidus. Skull radiography including the sella turcica showed no abnormalities.

Computed tomography (CT) revealed an isodense mass located in the intrasellar through the suprasellar region (Fig. 1 left), with part of the capsule enhanced by contrast medium (Fig. 1 right). Magnetic resonance (MR) imaging showed the mass as hyperintense on both T1- and T2-weighted images (Fig. 2).

A trans-sphenoidal surgery was performed. At the operation, the cyst contained yellow and mucinous fluid, and the cyst wall was partially removed. Histological examination of the cyst wall found ciliated columnar epithelium with basal cells which showed slight squamous metaplasia. A few goblet cells were

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observed in the epithelium. The histological diagnosis was consistent with Rathke's cleft cyst (Fig. 3).

During 2 years of follow-up, no regrowth of the cyst was observed. Laboratory tests after surgery showed the baseline values of the anterior pituitary hormones were within the normal range. However, the patient continues to receive hormone replacement for diabetes insipidus.

**Discussion**

The clinical and radiographic features of 13 juvenile cases with the diagnosis confirmed by pathological examination and details of features are summarized in Table 1.1,3,6,7,13-17

The clinical symptoms of Rathke's cleft cyst for all ages are anterior pituitary dysfunction (69.4%), visual disturbance (55.8%), headache (49.0%) and diabetes insipidus (12.9%).17 The clinical symptoms of the 13 juvenile cases were anterior pituitary dysfunction including growth retardation (62%), headache (62%), diabetes insipidus (46%), and visual disturbance (38%). Thus, the clinical presentation differs for adult and juvenile cases, with the latter tending to show diabetes insipidus more frequently, and visual disturbance less frequently. The cause of these differences is not yet clear. Possibly, ADH secretion in juveniles is more vulnerable than in adults.

Radiographic features of symptomatic Rathke's cleft cyst have been reported by several authors.1,3,6,7,13,14,17 Generally, symptomatic Rathke's cleft cyst has various MR imaging characteristics,1,3,6,7,13,14,17,18 but two-fifths of cases show watery contents, as reflected by low intensity on T1-weighted images and high intensity on T2-weighted images. On the other hand, most juvenile cases appeared as iso- to high density on CT scans and/or high intensity on T1-weighted images, except for Case 6 in Table 1, in which it was difficult to distinguish between Rathke's cleft cyst and intrasellar arachnoid cyst. This variation in cyst fluid may indicate different cyst contents. The high density on CT scan and hyperintensity on T1-weighted MR images that juvenile symptomatic Rathke's cleft cysts contain more mucopolysaccharides or hemosiderin degradation products than adult cases, as reported previously.1,14

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## Table 1 Clinical and radiographic features of symptomatic Rathke’s cleft cyst in children

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Symptoms and hormone examination</th>
<th>Radiographic findings</th>
<th>Fluid</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Iseki et al. (1981)⁷</td>
<td>11/M</td>
<td>GR, VD, PH</td>
<td>high, CE (+)</td>
<td>ND</td>
</tr>
<tr>
<td>2</td>
<td>Tajika et al. (1982)⁸ ⁹</td>
<td>11/M</td>
<td>GR, VD, PH</td>
<td>mixed, CE (+)</td>
<td>ND</td>
</tr>
<tr>
<td>3</td>
<td>Barrow et al. (1985)¹⁰</td>
<td>13/M</td>
<td>VD, HA, PH, DI, GR</td>
<td>iso, CE (+)</td>
<td>ND</td>
</tr>
<tr>
<td>4</td>
<td>Towbin et al. (1987)¹¹¹</td>
<td>10/M</td>
<td>GR, HA, DI, PH</td>
<td>high, CE (−)</td>
<td>T₁: high, T₂: low</td>
</tr>
<tr>
<td>5</td>
<td>Voelker et al. (1991)¹²¹</td>
<td>15/F</td>
<td>GR, HA, PH</td>
<td>iso</td>
<td>ND</td>
</tr>
<tr>
<td>6</td>
<td>Crenshaw and Chew (1992)¹³</td>
<td>15/F</td>
<td>HA, VD</td>
<td>low</td>
<td>T₁: high, T₂: high, CE (−)</td>
</tr>
<tr>
<td>7</td>
<td>Christophe et al. (1993)¹⁴</td>
<td>13/M</td>
<td>DI, HA, GR</td>
<td>normal</td>
<td>T₁: high, T₂: high, CE (−)</td>
</tr>
<tr>
<td>8</td>
<td>¹⁵</td>
<td>12/F</td>
<td>HA, AN, PH</td>
<td>iso</td>
<td>T₁: high, T₂: high, CE (−)</td>
</tr>
<tr>
<td>9</td>
<td>Ito et al. (1993)¹⁶</td>
<td>11/F</td>
<td>HA</td>
<td>high</td>
<td>ND</td>
</tr>
<tr>
<td>10</td>
<td></td>
<td>14/M</td>
<td>HA</td>
<td>high</td>
<td>T₁: high</td>
</tr>
<tr>
<td>11</td>
<td>Nakajou et al. (1994)¹⁷</td>
<td>13/M</td>
<td>DI, GR</td>
<td>ND</td>
<td>T₁: high, T₂: high, CE (−)</td>
</tr>
<tr>
<td>12</td>
<td>Ersahim et al. (1995)¹⁸</td>
<td>9/F</td>
<td>DI</td>
<td>ND</td>
<td>T₁: high</td>
</tr>
<tr>
<td>13</td>
<td>Present case</td>
<td>14/F</td>
<td>DI, GR</td>
<td>iso, CE (+)</td>
<td>T₁: high, T₂: high</td>
</tr>
</tbody>
</table>


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**References**


14) Oka H, Kawano N, Suwa T, Yada K, Kan S, Kameya...


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