Bilateral morning glory syndrome associated with sphenoid encephalocele

Case report

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Morning glory syndrome is a congenital anomaly of the optic disc, first described by Kinder in 1970. The disc is usually enlarged and excavated, with an elevated peripheral ring. White tissue in the center of the optic disc obscures retinal blood vessels. This anomaly was termed "morning glory syndrome" since it resembles the flower of that name.

Encephalocele is an uncommon, but not rare, brain anomaly and usually involves the occipital and parietal regions. However, basal encephaloceles in the sphenoid or ethmoid regions have rarely been reported. In this report, we describe an unusual case of morning glory syndrome associated with sphenoid encephalocele.

Case Report

This 22-day-old baby boy was referred to the Department of Neurological Surgery at Wakayama Medical College for evaluation of a large head size and dyspnea. He weighed 3180 gm at birth, after an uncomplicated 40 weeks' gestation and normal delivery.

Examination. Median cleft lip and palate were detected at birth, and a soft-tissue mass was observed in the oral cavity, causing breathing difficulty. Both optic discs were enlarged and excavated, with white glial tissue in the disc center (Fig. 1). A diagnosis of morning glory syndrome was made. The patient did not follow light with his eyes, but his spontaneous eye movement was normal. Skull roentgenograms revealed a bone defect in the sphenoid region. Computerized tomography and magnetic resonance (MR) imaging showed ventricular enlargement, agenesis of the corpus callosum, and a sphenoid meningocele. A mass with low intensity on T1-weighted and high intensity on T2-weighted MR imaging extended through the bone defect in the sphenoid region and into the retropharyngeal region and posterior oral cavity (Fig. 2 left).

Operation. Surgical repair of the sphenoid encephalocele to treat the dyspnea was attempted when the patient was 50 days of age. A bifrontal craniotomy was performed, and the sphenoid region was explored through a subdural approach. A bone defect was identified in the median part of the sphenoid plate, through which the arachnoid membrane was continuous with the oral cavity, creating a basal encephalocele. Both elongated optic nerves ran anterior to the bone defect after leaving the optic canal and formed the optic chiasm in the normal position. Since it was impossible to explore the entire bone defect and divide the stalk of
the encephalocele, the wall of the encephalocele was compressed digitally through the oral cavity and was sutured to the dura mater of the bone defect.

Postoperative Course The dyspnea improved postoperatively and no cerebrospinal fluid leakage was observed. Postoperative MR imaging showed that the encephalocele protruding into the oral cavity had decreased in size (Fig. 2 right). At follow-up study when the patient was 3 years 9 months of age, he was completely blind. The pupils were equal but neither eye showed reaction to light. Extraocular movement was normal.

Discussion

Five cases, including the present case, of morning glory syndrome associated with brain anomalies have been reported (Table 1).\(^1\),\(^2\),\(^3\),\(^4\),\(^5\) Four patients were male and one was female, with ages ranging from 22 days to 7 years. All five cases were associated with basal encephalocele; two had a cleft lip and agenesis of the corpus callosum. The combination of morning glory syndrome with basal encephalocele suggests a mechanism for the pathogenesis of morning glory syndrome. The hard palate is derived embryologically from the maxillary process arising from the first branchial arch. The palatine process originating from the maxillary process fuses completely with the nasal septum, derived from the frontal prominence, at about 60 days’ gestation. This produces the hard palate. Failure of the two palatine processes and nasal septum to fuse completely at 6 to 9 weeks’ gestation results in cleft lip and palate. At this developmental stage, neither the internal layer of the retina (ganglion cell layer) nor the optic nerves are well differentiated. During the 7th week of gestation, the axons of the retinal ganglion cells start to form the optic nerve, which is completely developed at 27 weeks’ gestation.\(^6\) If a sphenoid encephalocele prevents complete fusion of the palates, which precedes the formation of the optic nerve, then development of the optic nerve may be abnormal. In the present case, traction of the optic nerves drew them into the sphenoid bone defect. The fact that this anomaly occurs in combination with ethmoid or sphenoid encephalocele suggests the following. During development, a bone defect secondary to a basal encephalocele develops in the sphenoid or ethmoid bone. The optic nerves attached to the encephalocele become elongated and are drawn into the encephalocele. This results in the abnormal development of the optic nerves and the formation of white glial tissue in the center of the optic nerves.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Sex, Age</th>
<th>Patient Characteristics</th>
<th>Brain Anomaly</th>
<th>Affected Eye</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pollock, et al., 1968</td>
<td>M, 7 yrs</td>
<td>basal encephalocele</td>
<td></td>
<td>right</td>
</tr>
<tr>
<td>Caprioli &amp; Lesser, 1983</td>
<td>F, 17 mos</td>
<td>sphenoid encephalocele, cleft lip and palate</td>
<td>left</td>
<td></td>
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<tr>
<td>Nawratzki, et al., 1985</td>
<td>M, 10 mos</td>
<td>basal encephalocele, agenesis of corpus callosum, autism</td>
<td>both</td>
<td></td>
</tr>
<tr>
<td>Takezawa, et al., 1987</td>
<td>M, 58 mos</td>
<td>sphenoid encephalocele, pituitary dwarfism</td>
<td>left</td>
<td></td>
</tr>
<tr>
<td>Itakura, et al., 1992</td>
<td>M, 22 days</td>
<td>sphenoid encephalocele, cleft lip &amp; palate, agenesis of corpus callosum</td>
<td>both</td>
<td></td>
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</tbody>
</table>
Morning glory syndrome with encephalocele

References

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