

Computerized Tomography of Two Patients with Morning Glory Syndrome

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ABSTRACT

Morning glory syndrome (MGS), an uncommon optic disc anomaly, is characterized by a funnel-shaped, excavated optic disc surrounded by chorioretinal pigmentary disturbance. Generally, it is an isolated ocular abnormality. The authors describe two patients in whom MGS developed in association with brain abnormalities. In both cases, there was enlargement of the optic nerve that showed increased radiodensity similar to that of sclera and cavum vergae in the brain cavity present in computerized tomography (CT). To our knowledge, the coexistence of MGS, cavum vergae and an enlarged retrobulbar optic nerve showing increased radiodensity have not been previously reported. The findings suggest that MGS may be based on a developmental anomaly involving the brain, and the enlarged optic nerve may be associated with sclera because of the isodensity in CT.

Key words: *Morning glory syndrome, Cavum vergae, Enlarged retrobulbar optic nerve, Cerebral midline anomaly*

Morning glory syndrome is a rare congenital eye disorder that is characterized by an enlarged area of the optic disc with a funnel-shaped excavation, fluffy grayish glial tissue anterior to the optic disc, elevated annulus of chorioretinal pigment disturbance around the optic disc, and abnormalities of the retinal vessels. Kindler⁵⁾ introduced the term morning glory syndrome, because the ophthalmoscopic appearance of this anomaly closely resembles that of the morning glory flower. The anomaly is usually limited to the eye, and the retrobulbar optic nerve and brain appear normal. MGS is thought to develop from Bergmeister's papilla that arises in the 6-week-old fetus. Besides the congenital anomaly involving the optic disc, a few reports^{3,6,7)} have described the intracranial changes observed on CT in this syndrome. Cavum vergae, a large cavity between two lobulus of the median wall in the lateral ventricles which arises 12-19 weeks after conception, is a congenital cerebral, midline, anomaly. This cavity is easily diagnosed by CT. We present two patients with MGS who exhibited cavum vergae and an enlarged retrobulbar optic nerve that showed increased radiodensity.

CASE REPORTS

Case 1

A 42-year-old Japanese man was referred to the JR West Hiroshima Hospital in July 1989 with a sudden deterioration of vision in the right eye. There was no family history of ocular defects. His visual acuity was : right, 20/250; left, 20/12. The intraocular pressure, in each eye, determined by applanation tonometry, was 14 mmHg. The results of examinations of the left eye were normal. The pupils were briskly reactive and there was no afferent pupillary defect. The right optic disc appeared enlarged and excavated with a central area of glial tissue surrounded by a large annulus of light, variably pigmented, peripapillary tissue. A fluffy grayish-white mass of tissue was also present anterior to the optic disc. The head of the optic nerve was surrounded by a large area of retinochoroidal atrophy speckled with small amounts of pigment. The exit of the retinal vessels was radially oriented from the peripheral aspect of the disc (Fig. 1). Fluorescein angiography demonstrated that the fovea was touched by retinochoroidal degeneration (Fig. 1).

CT of the orbits showed a funnel-shaped excavation of the right optic disc. The right thickened retrobulbar optic nerve showed a radiodensity similar to the sclera (radiodensity

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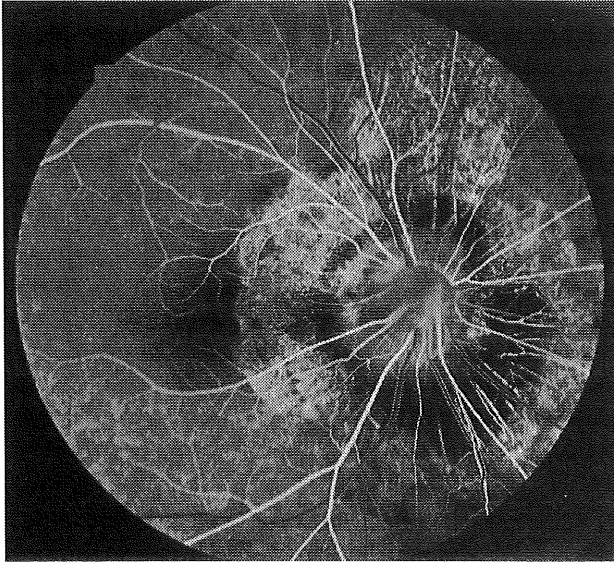


Fig. 1. Fluorescein angiography in case 1. The exit of the retinal vessels was radially oriented from the peripheral aspect of the disc, and the fovea was touched by retinochoroidal degeneration.

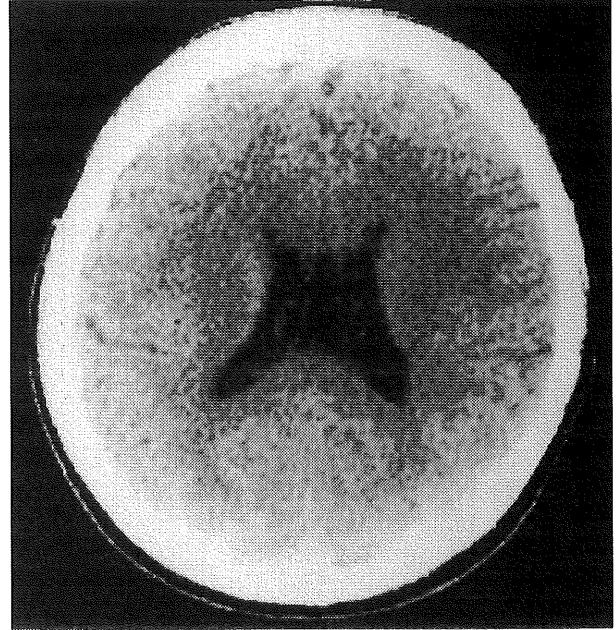


Fig. 3. Computerized tomography in case 1. Axial view of the brain showing cavum vergae.

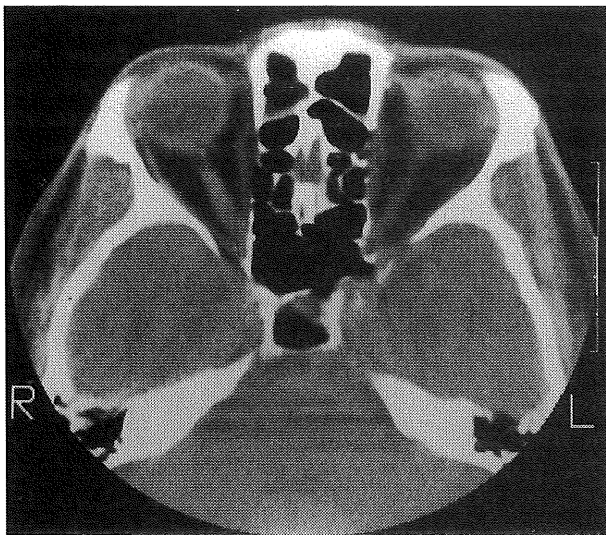


Fig. 2. Computerized tomography in case 1 (Axial view). The retrobulbar optic nerve is thickened with a radiodensity similar to that of the sclera which shows an increase in radiodensity with contrast.

with contrast medium: right 58.7 Hounsfield unit (H.U.), left 6.4 H.U., sclera 61.7 H.U.) (Fig. 2). The diagnosis was MGS. Cavum vergae was identified in the intracranial cavity (Fig. 3).

Case 2

A 14-month-old Japanese girl was referred to the JR West Hiroshima Hospital in November 1991 for an inward deviation of the left eye. Gestation, delivery, perinatal course and family

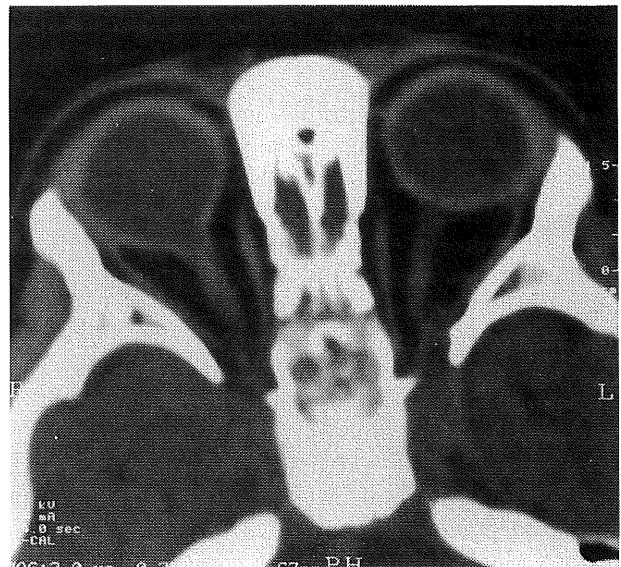


Fig. 4. Computerized tomography in case 2 (Axial view). Findings are similar to those of case 1 (Fig. 1).

history were all unremarkable. Her right pupil was normally responsive to light. The area of the right optic disc was deeply excavated and contained white tissue in the center that was surrounded by an elevated peripapillary white ring which extended into the central portion of the macula. Thus, the right eye showed typical features of MGS. The results of examinations of the left eye were normal.

CT examination demonstrated a posterior glob-

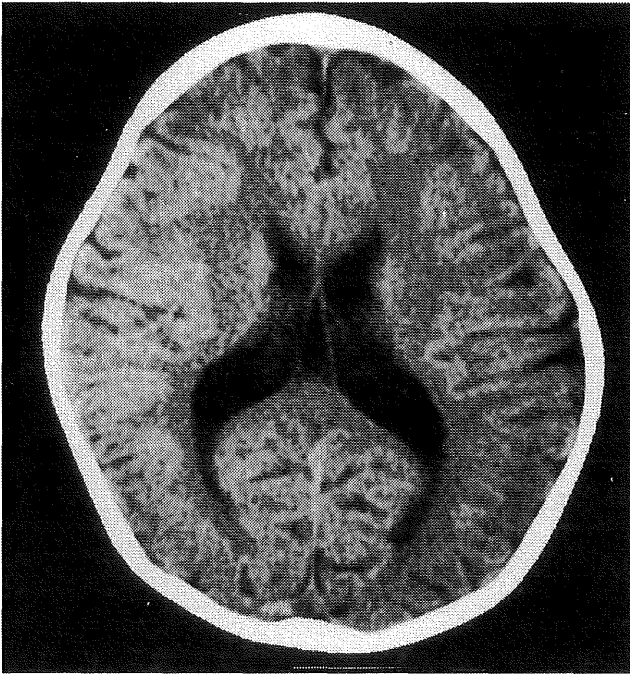


Fig. 5. Computerized tomography in case 2. Axial view of the brain showing cavum vergae.

al defect with a large excavation of the optic disc. The lesion consisted of a thickened retrobulbar optic nerve that showed an increase in radiodensity with contrast medium (right 47.6 H.U., left 20.4 H.U.) (Fig. 4). This radiodensity (47.6 H.U.) resembled that seen in the sclera (47.8 H.U.). The diagnosis was MGS. CT examination of the brain showed cavum vergae (Fig. 5).

DISCUSSION

The findings of examination of the optic disc in these two cases corresponded to the well-known clinical manifestations of MGS⁵.

Concerning the ocular and orbital findings observed on CT imaging, a thickened retrobulbar optic nerve⁴, a shallow excavation deformity of the globe¹, and the presence of hypodense matter within and surrounding the optic nerve⁴ have all been previously reported in MGS. CT findings in the intracranial cavity in this syndrome have been discussed by various authors^{3,6,7}. Koenig and associates⁶ described a patient with MGS who showed remnants of the primitive hyaloid vasculature, a midline cleft lip and palate, agenesis of the corpus callosum, and a sphenoidal encephalocele on CT. They thus suggested that MGS may be part of the syndrome of midline cranioencephalic dysraphism. The association of MGS with midline clefting abnormalities can be attributed to a common developmental defect in embryogenesis. Beyer et al reported a patient with MGS and trans-sphenoidal encephalocele, and considered a combination of ectodermal and

mesodermal dysgenesis to be involved in its pathogenesis³. Mafee et al⁷ described one patient with MGS whose brain CT showed a bifrontal mass, a symmetrical enlargement of the lateral ventricles including the temporal horn, and mild hydrocephalus with a large cisterna magna and pontine and suprasellar cisterns.

In our patients, CT demonstrated an enlarged retrobulbar optic nerve that showed increased radiodensity, similar to that of sclera, as compared to that of fellow eye. This is in contrast to Giuffrè⁴, who reported that the hypodense matter located inside and around the optic nerve in the CT finding of his case might be adipose tissue migrated into the optic nerve. This isodensity in the retrobulbar optic nerve and sclera suggest that the high dense matter may consist of a thickness of connective tissue rather than a replacement of fatty tissue².

Considering previous reports and findings in the present patients, it seems likely that MGS is associate with a variety of brain anomalies such as the cavum vergae reported for the first time in our two patients. The increased thickness of the retrobulbar optic nerve and cavum vergae in these patients is interesting in association with the embryologic origin of MGS. We recommend that patients with MGS have general physical examination and image diagnosis such as CT or magnetic resonance imaging (MRI).

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