Morning glory syndrome associated with marked persistent hyperplastic primary vitreous and lens colobomas

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SUMMARY A case of morning glory syndrome with striking persistence of primary vitreous is presented. The hypothesis that the syndrome is an expression of abnormal closure of the embryonic fissure is substantiated by the coexistence of lens colobomas. Furthermore, the marked primary vitreous hyperplasia shows the way in which persistence of primary vitreous influences the clinical expression with respect to optic pit and colobomas of the optic nerve.

Many associations between morning glory syndrome (MGS) and other ocular and non-ocular abnormalities in the same and in the fellow eye have been reported.1-22 Particular forms have been described, such as acute morning glory,23 contractile morning glory,17,24-30 and bilateral morning glory with normal visual acuity.31 Occasionally morning glory syndrome and primitive vitreal persistence are found in association,32-34 and some authors consider the terms morning glory and persistence of primary vitreous to the optic disk to be interchangeable.35

We report a clinical case of MG syndrome associated with pronounced persistence of primary vitreous and lens colobomas in the upper and lower nasal sectors. We believe no similar cases have hitherto been reported.

Case report

A 17-year-old girl was referred with reduced vision in the right eye from birth. It had been attributed to congenital cataract. Routine eye examination showed a visual acuity of 20/2400 in the right eye, which was not improved by lenses. The lens showed a posterior polar cataract and there were colobomas at 2 and 4 o'clock (Fig. 1). There was persistence of primary vitreous, with the ciliary body tightly stretched towards the centre of the pupillary field owing to the traction exerted on it by a whitish mass which represented the primary vitreous (Fig. 2). The pupillary reflexes were normal, and strabismus and nystagmus were absent. The fundus was examined with difficulty, but it showed (Fig. 3) a peripapillary staphyloma, at the centre of which was a greyish-white tissue mass from which straight, narrow retinal vessels originated and ran from the disc to the periphery of the retina. It was not possible to distinguish between veins and arteries.

A slightly raised pigmented ring could be observed round the staphyloma. Owing to the opacity of the

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Fig. 1 RE: posterior polar cataract and colobomas of the lens at 2 and 4 o'clock (arrowheads) are particularly evident.
Morning glory syndrome and primitive vitreal persistence

Fig. 2 RE: persistence of primary vitreous (arrowhead) with ciliary body tightly stretched towards the centre of the pupillary field (arrow).

Fig. 3 Typical MGS appearance: a residue of the hyaloid artery is clearly visible on the left.

dioptic media the macula could not be precisely localised. A remnant of the hyaloid artery in the form of a stalk protruded into the vitreous. No abnormalities were detected by gonioscopy.

The left eye was healthy, with VA 20/20.

Standardised echographic examination carried out according to Ossoinig\cite{1} revealed an axial eye length (AEL) within normal limits (anteroposterior=23 mm) and a lens of normal dimensions in the right eye.

B scan echography revealed a morning glory disc filled with acoustically dense material and also hyaloid artery residue.

Discussion

It is debatable whether MGS represents a single entity or whether it is one of many expressions of the same pathogenetic stimulus. We do not agree with the hypothesis that it is a single entity caused by the persistence of the cavity extension of the optic cup in the optic stalk.\cite{17}

The present report is thought to corroborate the hypothesis that MGS represents an expression of the abnormal closure of the embryonic fissure (which is suggested by the presence of lens colobomas in the nasal sectors, particularly the inferior sector in the case here described), and we would stress the role of the persistence of primary vitreous in the determination of MGS.

In view of the clinical similarities, and considering the timetable of embryological development of the optic nerve, we believe that MGS represents the clinical form of a pathogenetic process, namely, the abnormal closure of the embryonic ocular fissure. This has a variety of clinical expressions ranging from MGS to optic pit,\cite{10,14,36} which can, however, be included in the family of optic nerve colobomas.\cite{13,34,37,38}

We suggest that the mass of whitish tissue which is ophthalmoscopically visible (and clearly demonstrable with B scan echography) on the abnormal MGS disc corresponds to a highly vascularised connective tissue in the area normally occupied by the physiological excavation deriving from the primitive hyaloid system.

Different degrees of the persistent hyperplastic primary vitreous process give rise to a spectrum of clinical expressions. They range from slight (which have been classified erroneously, in our opinion, as distinct clinical and embryological forms, peripapillary staphylomas\cite{19}) to conspicuous forms, with a noticeable mass of hyperplastic primary vitreous and with the classical appearance of ciliary body stretched towards the centre of the pupillary field, as described in our case. This range includes intermediate manifestations such as the persistence of lens tunica vascularis (Gass), the extension of vascular branches from the abnormal disc to the posterior lens surface,\cite{14,34} and the protrusion of fibrovascular tissue into the anterior vitreous, which does not reach the posterior capsule.\cite{44}

For these reasons, even though the pathogenesis of MGS is still not ascertained—whether there is a mesenchymal abnormality resulting in faulty closure of the posterior scleral wall or abnormal closure of the embryonic fissure, or both\cite{36} we support the unitary hypothesis that MGS, optic pit, and optic nerve colobomas are expressions of a single pathogenetic process. The persistence and hyperplasia of the
primary vitreous is a fundamental condition of the MG disc and has a great variety of clinical expressions, ranging from the mere presence of whitish tissue on the optic disc surface in the case of simple persistence of the hyaloid artery to more complex clinical pictures.

References


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