Case Collection

Morning Glory Optic Disc Syndrome

Rana SSJB[1]

Abstract

Morning glory optic disc syndrome refers to an uncommon specific optic nerve appearance of an excavated disc defect which resembles the morning glory flower and hence the name. Diagnosis of this syndrome usually occurs in the first few years of life and the child is at risk of acquired visual loss. Reporting four such cases detected in a span of five months i.e. from January to May in the year 2006. Out of the four cases, one is bilaterally blind, two uniocular blind and the vision has not yet been affected in one of the cases. The psychosocial trauma of the child's blindness to the family and society is definitely devastating. For the child the long term burden of disability is tremendous.

Case Reports

Case – 1

A 13 – years – old girl was referred to this department by her class teacher as she had difficulty in seeing the letters written on the blackboard from the back benches. She had no other complaints. Her visual acuity was 6/18 in both eyes corrected to 6/6 by -0.25 D.Cyl. at 90° in both eyes. Fundus examination under mydriasis revealed the morning glory optic disc syndrome in the left eye, while the right eye showed no abnormality. She was cautioned accordingly, prescribed spectacles and asked to come for regular follow up.

Case – 2

16 – year – old girl with a history of deteriorating vision in both eyes since about 2 years. Her visual acuity was 3/60 in the right eye and 1/60 in the left eye. No visual improvement in both eyes with refraction. Fundus evaluation under mydriasis showed morning glory optic disc syndrome in both eyes with old retinal detachments. The poor visual prognosis was explained.

Case – 3

20-years – old girl was referred to this hospital from Bhaktapur. Her visual acuity was 6/6 in the right eye and 6/60 in the left eye. The vision in the left eye did not improve with refraction. Fundus evaluation under mydriasis showed morning glory optic disc syndrome with macular capture in the left eye. The right fundus revealed a cup disc ratio of 0.7:1. The patient was asked to come for regular follow up.

Case – 4

12 – year – old girl accidentally discovered that she could not see with her left eye. Dust had entered her right eye and due to the ocular irritation she closed that eye and to her horror she realised that she could not see clear enough with her left eye. She visited this department and on ocular examination the visual acuity in right eye was 6/6 and in the left eye 6/60 with no improvement on refraction. Fundus evaluation under mydriasis showed morning glory optic disc syndrome in the left eye. The poor visual prognosis was explained and the patient asked to come for regular follow up.

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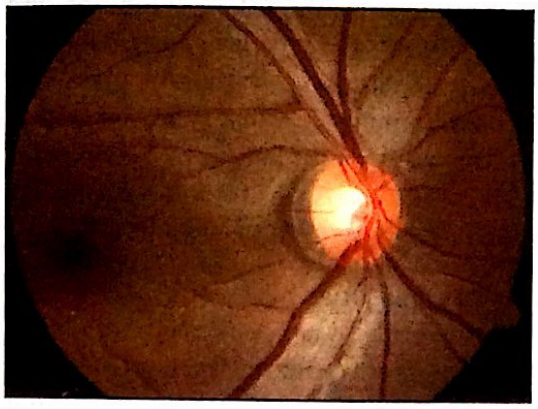
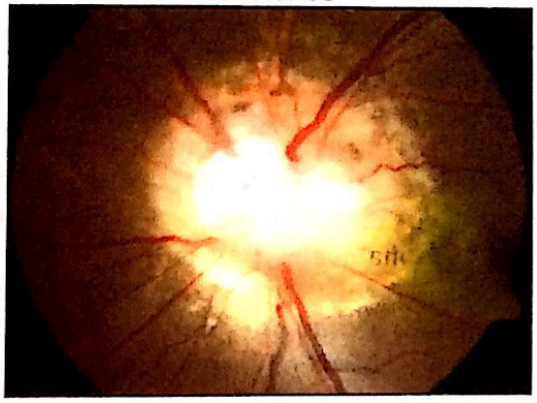
Morning Glory Optic Disc Syndrome

Case 1 : Age - 13 yrs.

6-1-2006 OD

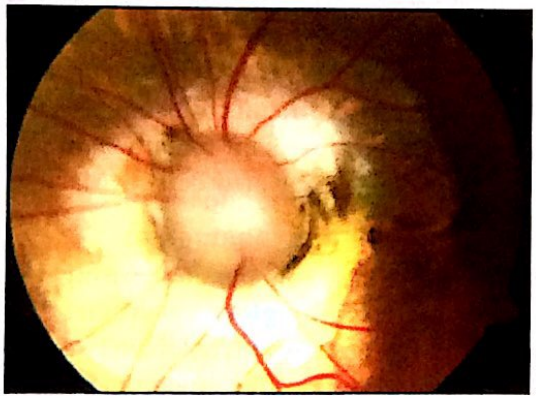


6-1-2006 OS



Case 2 : Age - 16 yrs.

23-02-2006 OS



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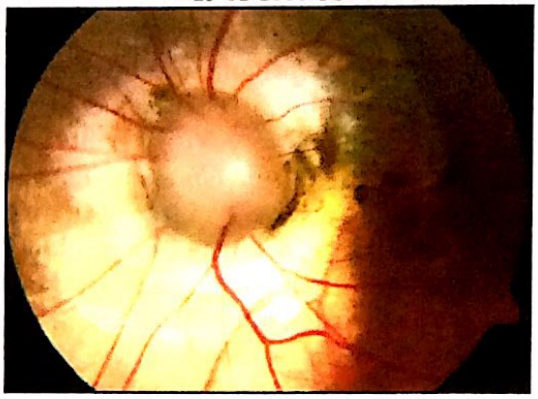
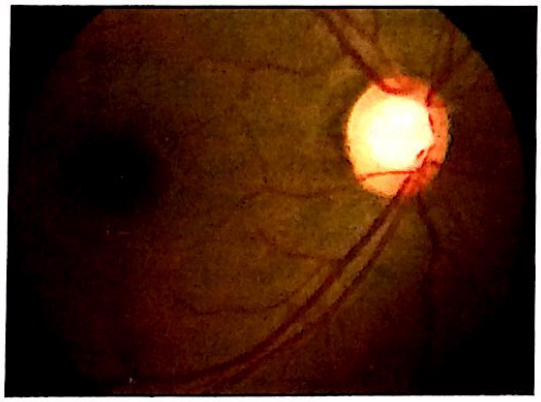
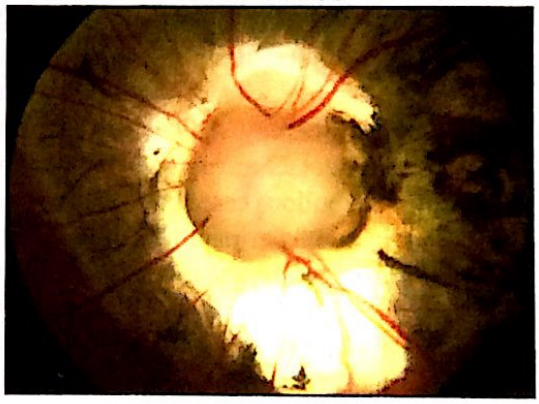


Case 3 : Age - 20 yrs.

21-03-2006 OD

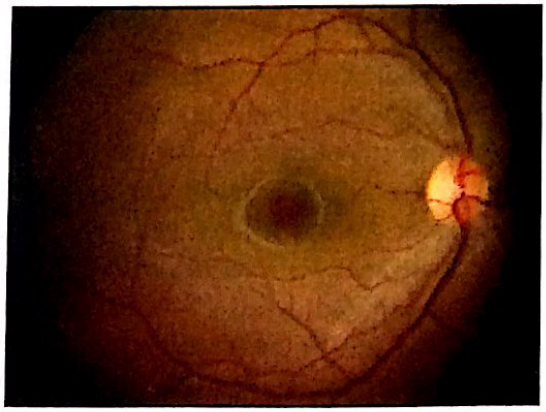


21-03-2006 OS



Case 4 : Age - 12 yrs.

31-03-2006 OD



21-03-2006 OS

Discussion

All the four patients were girls. Although initially there are some reports of a predominance in female persons, there probably in no significant difference in male versus female.[1] None of the cases were associated with systemic defects, such as hypertelorism, cleft lip and palate, renal anomalies and encephaloceles.[2] Descriptions of ocular anomalies associated with morning glory syndrome also abound, including strabismus cataract, nystagmus, ciliary body cyst, lens coloboma, eyelid hemangioma, aniridia, and drusens.[3] Diagnosis usually occurs in the first few years of life, with the majority of children presenting with strabismus. There dose not seen to be any predilection to abnormalities in pregnancy, birth weight or prematurity and there is no connection with a significant family history. Visual acuity can range the entire spectrum from 6/6 to questionable light perception, although 90% of patients have 6/60 or worse. The poor vision may be a result of retinal anomalies but vision could be grossly impaired even without retinal detachment. Rhegmatogenous retinal detachment is found in 30% of cases. In many of the cases the retinal detachment is connected to the optic disc.[4] The retinal detachment most typically appears during the first and second decades of life. The origin of the sub retinal fluid is unclear.[5] The detachments can be shallow or bullous. Particularly when bullous, they may not respond well to therapy. Advocated treatment for shallow retinal detachments can be laser therapy alone, but more bullous cases pars plana vitrectomy in combination with peripapillary laser therapy and internal drainage of subretinal fluid may be necessary to flatten the retina.[6] Optic nerve sheath decompression also has been reported to treat this form of retinal detachment, but its exact role is uncertain.[5] However the cases seen in this department reported quite late in life and no ocular therapy could be instituted. Abnormal vascular changes and subretinal neovascular membrane formation are variably associated. Fortunately the condition is commonly unilateral but it can also be bilateral.

This syndrome was first described by Handeman in 1929 but it was not until 1970 that is 40 years later that the term morning glory syndrome was coined by Kindler. There is some variation about how morning glory disc syndrome is distinguished from the other congenital excavated optic disc defects of optic disc pit and coloboma. Some authors use the term optic disc coloboma and morning glory syndrome interchangeably. Although mistakenly referred to as a variant of optic disc coloboma, the morning glory disc syndrome is truly a distinct anomaly, as evidenced by its sporadic occurrence, its lack of association with iris or retinal coloboma, and its systemic associations.[7]

Morning glory optic disc syndrome is characterised ophthalmoscopically by an enlarged orange or pink optic disc with either a central excavation or situated within of funnel shaped area of excavation, a central tuft of dysplastic white retina, a peripapillary subretinal fibroses and straightened retinal vessels that are often sheathed and emanate for the edge of the disc. The retinal blood vessels appear increased in number. It is often difficult to distinguish the arteries from the veins. The macula may be incorporated into the excavated defect (Macular capture).[7]

There is some disagreement concerning the pathogenesis of morning glory syndrome. The association in a few cases of lens colobomas argues for an abnormal closure of the embryonic ocular fissure. As such, it further supports the idea that morning glory disc syndrome is a variation of a pathological process that includes optic nerve colobomas and optic pits.[8] Pedler[9] suggested that there my be abnormal fusion of just the posterior sclera, with subsequent herniation of the disc and peripapillary retina. Another suggestion was put forth by Dempster and associates,[10] who proposed that it is rather an error in mesodermal differentiation, or at least disturbance in the relative speed of differentiation of the mesoderm versus the ectoderm, that results in abnormal closure of the posterior scleral wall and lamina cribrosa. This would allow herniation of retinal and neural tissue and of the optic nerve head, resulting in the excavation seen.

The visual potential of the eye, including the stage of macular development, should be a critical factor in determining whether treatment; be it surgical or amblyopia treatment; be pursued aggressively. Though many of the cases have good surgical reattachment outcomes, the final visual acuities are fairly poor, ranging from 6/60 to light perception. Clearly, therapy me be better directed if the pathogenesis of this rare disc anomaly can be more clearly established.

References

1. Traboulsi EI, Lee O'Neill JF. The spectrum in the morphology of the so-called "morning glory disc anomaly." J Pediatr Ophtholmol Strabismus 1988;25(2):93 – 98.

2. Eustis HS, Sanders MR, Zimmerman T. Morning glory syndrome in children. Association with endocrine and central nervous system anomalies. Arch Ophthalmol 1994;112:204 – 207

3. Hope-Ross M, Johnston SS. The morning glory syndrome associated with sphenoethmoidal encephalocele. Ophthalmic Pardiatr Genet 1990;2(2):147 – 153

4. Beyer WB, Quincex RM, Osher RM. Morning glory syndrome. A functional analysis including fluorescein angiography ultrasonography and computerized tomograpgy. Ophthalmology 1982;100: 1361 – 7

5. Irbine AR, Crawfore JB. The pathogenesis of retinal detachment with morning glory disc syndrome and optic pit. Retina 1986;6: 146 – 50

6. Brown GC, Brown MM. Treatment of RD associated with congenital excavated defects of the optic disc. Ophthalmic Surgery 1995;26:11 – 15.

7. Pollock S. The morning glory disc anomaly. Contractile movement classification and embryogenesis. Doc Ophthalmolol 1987;65:442 – 53.

8. Cennamo G, Liguori G, Pezone A, Iaccarino G. Morning glory syndrome associated with marked persistent hyperplastic primary vitrous and lens colobomas. Br J Ophthalmol 1989;73:684 – 686.

9. Pedler C. Unusual coloboma of the optic nerve entrance Br J Ophthalmol 1961;45:803 – 807.

10. Dempster AG, Lee WR, Forrester JV, McCreath GTM. The morning glory syndrome – a mesodermal defect? Ophthalmologica 1983;187:222 – 230.