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Angio OCT of the Macula and Optic Disk in a Rare Case of Bilateral Morning Glory Disc Anomaly

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Abstract: To report a rare case of bilateral morning glory disc anomaly and to evaluate the Angio OCT characteristics of macula and the optic disk in a case of bilateral morning glory disk anomaly.
Keywords: Angio OCT, Morning Glory Disk Anomaly

INTRODUCTION

The term morning glory syndrome was given by Kindler in 1970 to a rare ocular anomaly characterised by a large staphylomatous optic disc covered by glial tissue [1]. The disc itself is enlarged, and orange or pink in color within a surrounding area of peripapillary chorioretinal pigmentary changes. Within the center of the disc is a white glial tuft. Similar to petals on a flower, the blood vessels are increased in number and curve as they emanate radially from the disc, rather than in the usual central branching pattern. They then straighten, and it is often difficult to distinguish the arteriolar from the venous circulation. There are also small peripapillary arteriovenous communications. Depending on the size of papillary chorioretinal involvement, the macula may be incorporated into the excavation, which is termed "macular capture" [2, 3].During the last few years many cases of morning glory syndrome have been described. These reports have contributed to the clarification of various clinical and pathological problems of the disease. However the Angio OCT have not been investigated extensively in these cases. Hence we are reporting a rare case of a bilateral morning glory syndrome with angio OCT characteristics of the macula and optic disc.

CASE REPORT

A 15 year old male referred to our hospital in the Retina Department for ophthalmologic examination. He noticed diminution of vision in the left eye since 1year. His birth history and family history were unremarkable. The ocular adnexa were within normal limits. Confrontation fields by finger counting were full in both eyes. Pupils were round and regular and the reflexes were normal to light and accommodation in both eyes with no relative afferent pupillary defect. Corrected visual acuity was 6/6 in the right eye and 6/36 in the left eye. The refractive error in the left eye was of (-2-50 D, 60). The anterior segment appeared normal with the slit lamp examination.

Ophthalmoscopy revealed an enlarged and excavated optic disc in his right eye. A tuft of whitish tissue was present in the centre of the disc. Peripapillary chorioretinal atrophic changes were noted inferior and around the optic disc. Retinal vessels are increased in number, emerged from under the central tissue and followed a relatively straight course to the periphery of the retina. The foveal reflex was absent in the right eye. The peripheral retina was intact without suggestion of retinal detachment. (figure1).

Enlarged and excavated optic disc was also noted in his left eye. A tuft of whitish tissue was present in the centre of the disc. Circular area of peripapillary pigmentary changes was around the optic disc in the left eye. Retinal blood vessels are increased in number followed a relatively straight course to the periphery of the retina. The foveal reflex was absent in the left eye also along with macular edema. No evidence of retinal detachment noted. (figure2)

Optovue Avanti AngioVue system technology for optical coherence tomography (OCT) angiography (Optovue, Inc., Freemont, CA), which runs on split-spectrum amplitude-decorrelation angiography (SSADA) algorithm was used for taking OCTA. Angio OCT retina 6×6 mm scan size protocol for macular OCTA and Angio OCT optic disk 4.5×4.5 mm protocol for OCTA of the optic disk was used.

Right eye OCTA of macula shows intact superficial and deep capillaries with normal appearing outer retina and choriocapillaries. However enface image shows altered reflectivity just nasal to the fovea which is corresponding with the macular retinoschisis area in the OCT. (figure 3).

Left eye OCTA of the retina shows intact superficial capillaries with loss of the deep capillaries with normal appearing outer retina and choriocapillaries. However enface image shows altered reflectivity in the macular area which is corresponding with the macular retinoschisis and sensory detachement in the OCT. (figure 4).

OCTA of the optic disk shows the horizontal oval configuration of the optic disk with vessels emerging out at the edge of the optic nerve head, presence of a dense radial peripapillary network in RE. En-face OCT of the optic disk shows hyper reflective centre surrounded by hypo reflective ring in right eye. Angio OCT of the optic at the level of choroid shows over exposure of the choroidal vessels which corresponds to the chorioretinal atrophy inferonasal to the optic disc in the fundus photo of the right eye. (figure5)

OCTA of the optic disk in the left eye shows large disc with vessels emerging at the edge of the disc in a more straighter course, and the presence of a radial peripapillary microvascular network. Left eye shows hyperfluorescence on wide field En-face OCT, which was surrounded by a hyporeflective ring, corresponded to a vascular tissue lesion. (figure6)

SD OCT of the optic disk in the both eyes shows the excavation and the preretinal membranous tissue and, in left eyes an anomalous communication in the retina between the subarachnoid space and the subretinal space. (figure7, figure8)

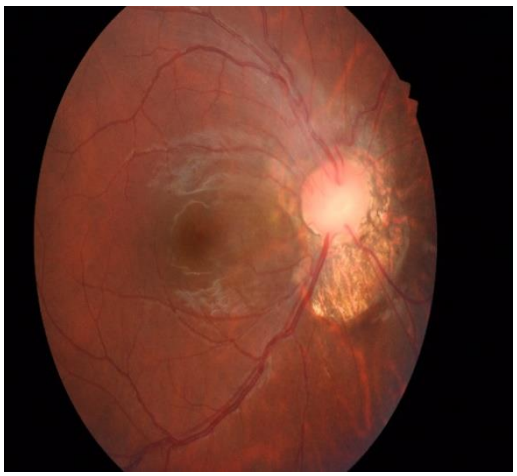

Fig-1: (right eye fundus photo)

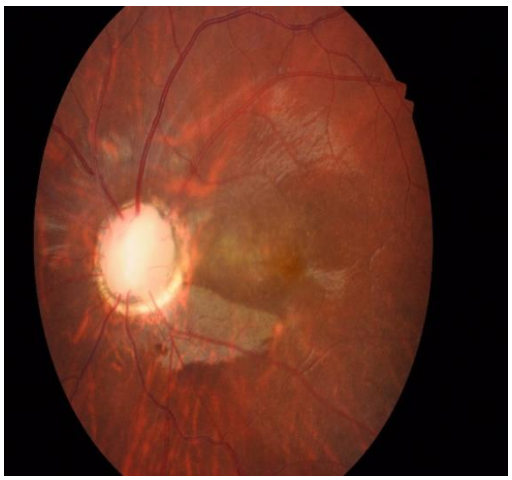

Fig-2: (left eye fundus photo)

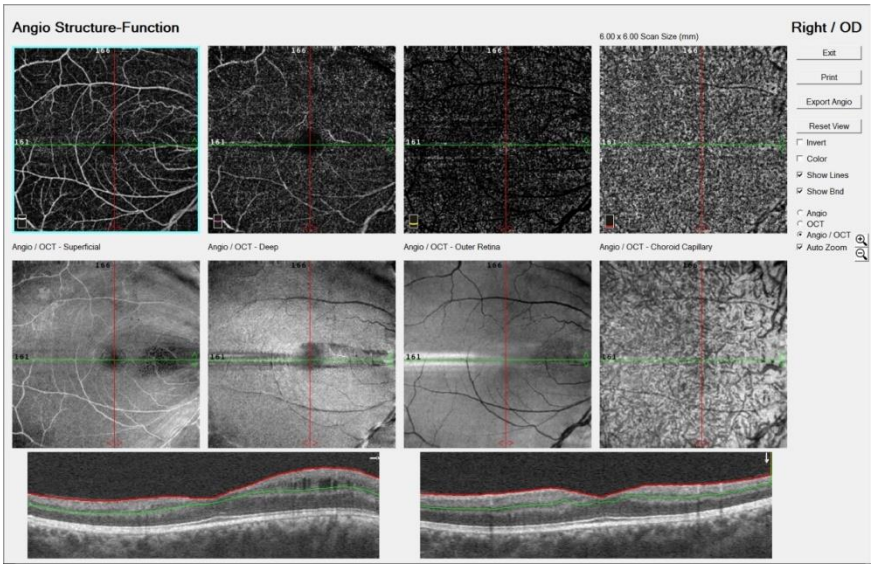

Fig-3: (Left eye Angio OCT Macula 6×6 mm scan)

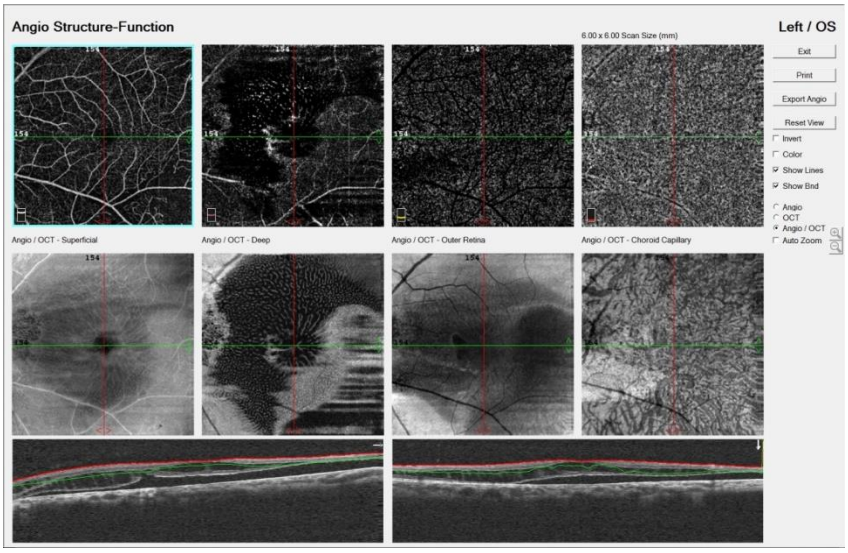

Fig-4: (Right eye Angio OCT Optic disk 4.5×4.5 mm)

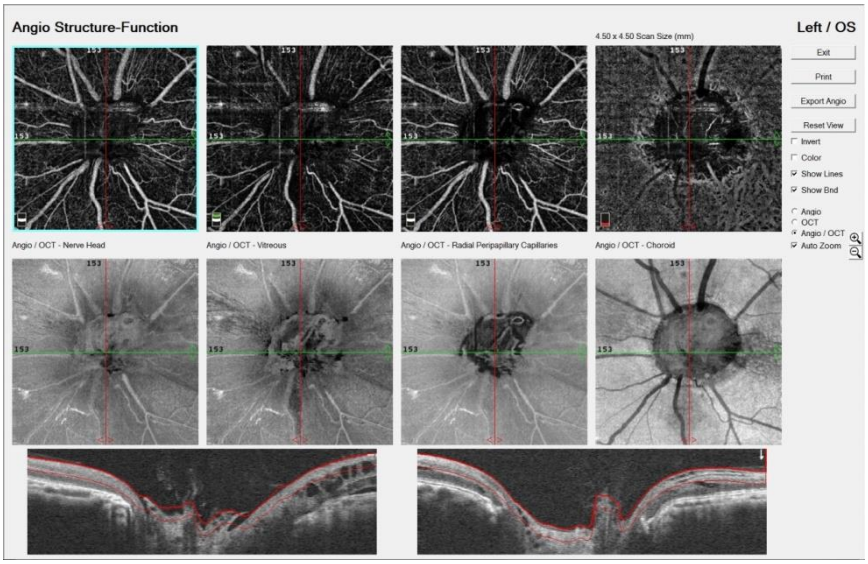

Fig-5: (Left eye Angio OCT Optic disk 4.5×4.5 mm)

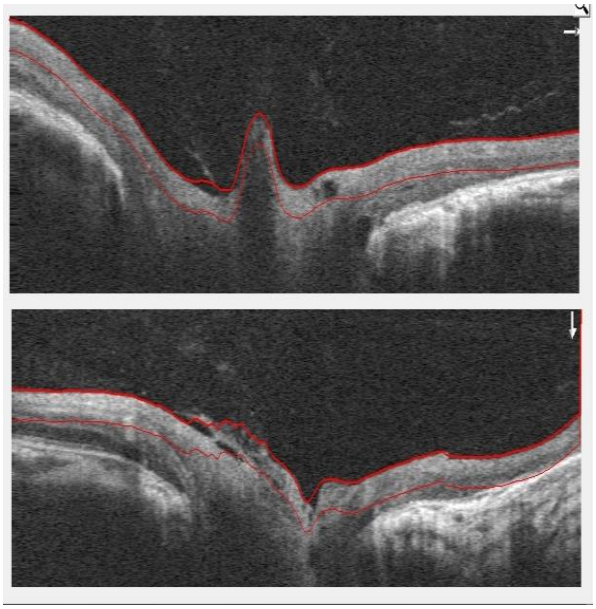

Fig-6: SD OCT of optic disk right eye

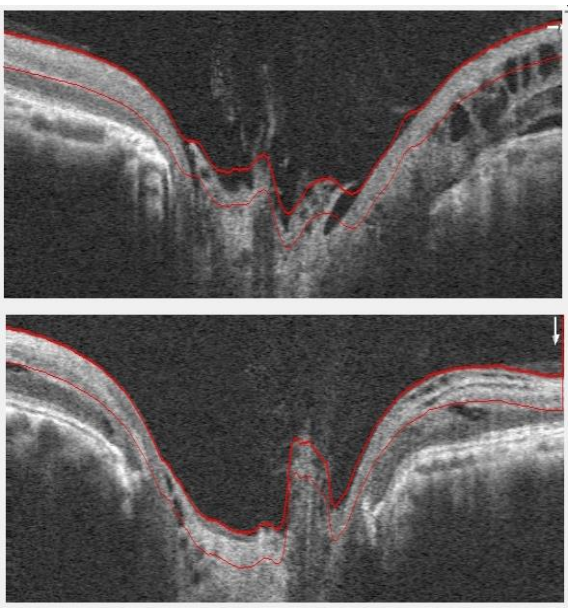

Fig-7: SD OCT of optic disk left eye

DISCUSSION

Morning Glory syndrome is a congenital anomaly of the optic disk in which there is a funnel shaped excavation of the posterior fundus incorporating the optic nerve, surrounded by an elevated annulus of chorioretinal pigment.

A central core of white glial tissue occupies the position of the normal optic cup, causing a white mass. When a picture is taken of the eye, this white mass stands out apart from the veins of the eyes, looking very much like the center of a Morning glory flower. Morning Glory Syndrome typically affects only one eye; but rare cases have been documented of bilateral Morning Glory Syndrome. Our case was also bilateral MGDA.

The retinoschisis and subretinal fluid in the left eye in our case could be due to slit-like retinal break at the edge of the excavated anomalous disc [4] or a retinal hole in tissue lying within the optic cup [5-7]. These provide a fluid pathway between the vitreous cavity and the subretinal space. The embryogenesis of MGS is poorly understood [8].Some has proposed that MGS results from the abnormal closure of the embryonic fissure [9, 10] More recent studies have suggested that MGS is a primary mesenchymal abnormality resulting in faulty closure of the posterior scleral wall and the poor development of the lamina cribrosa. To our knowledge, this is the second study to use angio-OCT to evaluate the radial peripapillary capillary network and first study to use angio OCT to

evaluate macula in MGDA in the attempt to shed light on the pathogenesis of these rare disease. In our case there is loss of deeper capillaries in angio OCT of the macula with intact superficial capillaries in the left eye. The clinical features and ancillary investigations establish the diagnosis of MGDA in our patient. Angio-OCT is a noninvasive diagnostic tool that could provide information about structural characteristics of the MGDA. The purpose of reporting this case is to highlight the possibility of a rare bilateral existence of MGDA and to report the Angio OCT characteristics of MGDA.

CONCLUSION

Noninvasive investigative modality like Angio OCT can be used as a diagnostic tool and to evaluate the structural characteristics of the MGDA.

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