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RESEARCH ARTICLE

MORNING GLORY SYNDROME:A RARE ENTITY

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Abstract

Morning glory syndrome is a rare congenital optic disc anomaly that can cause visual impairment and may be linked to ocular and non-ocular abnormalities. It is characterized by a distinct fundoscopic appearance, which includes a large excavated disc with radial vessels and a central turf of glial tissue within a funnel-shaped excavation of the posterior fundus. In this case report, we present a female middle aged, with this uncommon congenital optic disc anomaly.

Introduction:-

It is a rare congenital optic disc abnormality [1]. It was first described as an unusual congenital disc anomaly as "morning glory syndrome" because of its resemblance to the morning glory flower [2]. An expanded, funnel-shaped excavation that includes the optic disc is its defining feature. Peripapillary chorioretinal pigmentary alterations surround the larger disc, which has an orange or pink hue. It is typically unilateral; it can be bilateral too but more often unilateral cases are seen [3]. The visual prognosis is usually not very good. Retinal detachments are also seen to occur in 20%-30% of individuals. It is hypothesized that there is contact between the subretinal region and the subarachnoid area within the posterior excavation, however this communication is not well understood [4].

Case Report

A 30-year-old lady presented to our opdwith complaint of diminished vision in both the eye since childhood. The visual acuity of the right eye was measured at 3/60, whereas the left eye had a visual acuity of 6/60.

She had no previous record of hallux valgus or facial clefts. Additionally, there was no familial background of ocular disease or congenital anomalies. Upon examination, no facial abnormalities were observed and the neurologic assessment showed normal results.Her birth history and family history were not eventful. Her best corrected visual acuity for distance with cycloplegic refraction was −0.75DS, −2.5 DC × 180°, 6/24 for Right eye; and -0.50DS,6/12 for left eye.

No nystagmus was observed, no strabismus was seen (orthophoric) withnormal ocular motility. The anterior segment examination yielded no notable findings in the eye. Fundoscopy was done of both eyes and it revealed a large excavated disc with a central tuft of glial tissue (whitish tissue) and peripapillary chorioretinal pigmentation. The blood vessels were observed to be supernumerary and arranged radially from the disc, foveal reflex was dull and the peripheral retina was intact and did not exhibit any signs of retinal detachment. (see Fig. 1). Intraocular pressures measured were found to be 14mmHg and 15mmHg in the right and left eyerespectively. We also did USG B-scan which showed an axial length of 25.7mm in the right eye and 23.3mm in the left eye.

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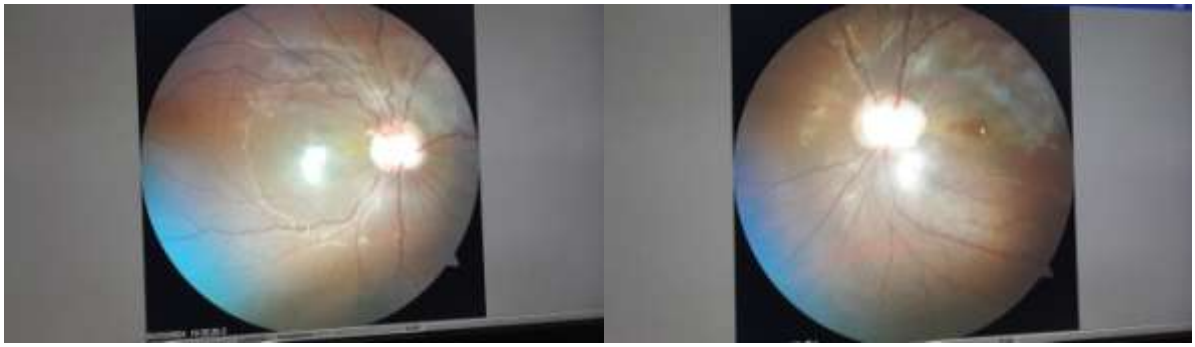

Fig.1:- Fundus picture showing A) excavation of the optic disc with peripapillary pigmentation and a white tuft of glial tissue overlying the central portion of the disc and the increased number of blood vessels arising from the periphery of the disc B) similar findings seen in left eye.

Morning glory syndrome can also lead to many complications such as cataract, secondary glaucoma, corneal oedema, retinal detachment, strabismus, nystagmus and accordinglythey were given further treatment.

If the patient had cataract, then the patient was advised for lensectomy, or if there is retinal detachment one would go for PPV, some patients also have cataract with retinal detachment, then they had lensectomy with vitrectomy.

Evaluation

The evaluation of the optic disc abnormality is aided by optical coherence tomography (OCT). Research has demonstrated that the retina is pulled centripetally by a distinct epiretinal membrane [8]. Early diagnosis and assessment of potential sub-retinal fluid are greatly aided by OCT, which also provides information on the pathophysiology and related clinical characteristics. An expanded optic disc and cup with increased RNFL thickness temporally was revealed by Srinivasan et al.'s[9] OCT examination (quantitative) of a case with isolated MGDA. This violation of the inferior, superior, nasal, and temporal (ISNT) rule and subnormal macular thickness was seen.

Optimizing visual acuity is crucial in preventing amblyopia as there is no such treatment for morning glory syndrome. To rule out CNS involvement, including basal encephalocele, Moyamoya, and other structural or vascular abnormalities, brain imaging (MRI, MRA, and/or CTA) should be performed [10].

Discussion:-

This particular congenital anomaly of the optic disc was initially documented by Handmann.[5] Subsequent reports followed by Kindler [2] who named the anomaly 'morning glory' due to its resemblance to flowers belonging to the genus Ipomoea, which are akin to the convolvulus found in British hedgerows. In Kindler's case it was accompanied by a hemorrhage at the periphery of the malformed optic disc, which eventually resolved. Other instances were described in which morning glory discs were associated with significant ocular and neurological congenital abnormalities, such as basal encephalocoele,[6] choroidal coloboma,[7] microphthalmos,[8] and persistent hyperplastic primary vitreous.[11]

Morning glory syndrome is a type of congenital optic disc anomaly, along with optic disc coloboma and peripapillary staphyloma. Unlike optic disc coloboma and peripapillary staphyloma, morning glory syndrome is not hereditary,[12] and its associated genetic defects have not yet been established. While a case of morning glory syndrome has been reported in a patient with Down syndrome, (Altun et al., [13]) no other genetic abnormalities were detected aside from trisomy of chromosome 21, which is characteristic of Down syndrome. The exact cause of morning glory syndrome is not well understood. It is theorized that the excavation may result from either insufficient closure of the embryonic fissure, making it a variant of optic nerve coloboma, or from a primary mesenchymal abnormality that leads to a scleral defect, central glial tuft, and vascular abnormalities. [14]

Morning glory syndrome can be clinically diagnosed based on its distinctive appearance during fundoscopy. This involves observing a funnel-shaped excavation of the posterior fundus that encompasses and incorporates the optic disc. The optic disc itself is enlarged and excavated, exhibiting a central region of white glial tissue and peripapillary chorioretinal pigmentation. Additionally, the blood vessels present in this condition are supernumerary and radiate in a radial fashion from the disc's periphery, making it difficult to differentiate between arterioles and venules.

The visual acuity of eyes with morning glory syndrome can vary from no light perception to normal vision. However, in most cases, it is impaired.[14] Additionally, the refractive error in eyes with morning glory syndrome is commonly myopic.[15] In the case of our patient, they had poor vision, high myopic astigmatism, and impaired vision in both the eye affected by morning glory syndrome. It is worth noting that our patient did not receive any amblyopia treatment during childhood. Poor vision in patients with congenital structural abnormality may be due to a combination of functional amblyopia and the organic defect.[16] Functional amblyopia refers to poor vision caused by a lack of pattern vision or abnormal binocular interaction in an eye that appears normal upon physical examination.[17] On the other hand, organic amblyopia refers to poor vision caused by structural abnormalities in the eye or brain that are independent of sensory input, such as structural abnormalities in the visual pathway.[18] While organic amblyopia is traditionally considered irreversible, there have been reported cases of improved visual acuity with amblyopia therapy in children with morning glory syndrome. For instance, Cavazos-Adame et al.[19] reported a case of a 3.8-month-old child with unilateral morning glory syndrome whose visual acuity improved from counting fingers at 0.3 meters to 20/100 after 5 years of amblyopia therapy.

Ocular associations that have been reported with morning glory syndrome include nystagmus,strabismus, persistent hyperplastic primary vitreous (PHPV),retinal detachment, microphthalmia,cataract, optic nerve glioma and corneal leucoma.[15,20,21] Non-ocular associations of morning glory syndrome include basal encephalocele, capillary hemangiomas, corpus callosum agenesis,cerebrovascular anomalies such as moyamoya disease,carotid artery stenosis and pituitary gland abnormalities and facial abnormalities such as median cleft lip and palate. [10,15,22]

Optic disc coloboma is considered as a potential differential diagnosis for morning glory syndrome.[23] In morning glory syndrome, the entire disc appears excavated, in contrast to optic disc coloboma where the excavation is confined within the optic disc, typically in the inferior aspect, while the superior neuroretinal rim remains discernible [23]. Another possible differential diagnosis for our patient, who also had high myopia, is posterior staphyloma. In posterior staphyloma, there is excavation of the posterior fundus surrounding the optic disc, and the optic disc itself is usually flat, lacking a central tuft. The retinal vasculature in posterior staphyloma is typically normal [23,24]. However, given that our patient exhibited excavation of both the posterior fundus and the disc, along with the presence of a central tuft of tissue on the disc and abnormal retinal vasculature, posterior staphyloma is considered less likely in this case.

Conclusion:-

In conclusion, the morning glory syndrome is a rare congenital anomaly of the excavated optic disc that poses a threat to vision. This condition can be accompanied by both ocular and non-ocular manifestations. Toprevent deep amblyopia, organic congenital defects of the optic nerve must be diagnosed early and treated accordingly.

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