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Original Research Paper

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CRYSTALLINE RETINOPATHY WITH PERIFOVEAL TELANGIECTASIA – A CASE REPORT

Dr. Vishal Agrawal M.D., F.R.C.S. Professor, Department of Ophthalmology, SMS Medical College, Jaipur.

Dr. Anushree Sharma* D.N.B. Ex.Senior Resident, Department of Ophthalmology, SMS Medical College, Jai-pur. *Corresponding Author

ABSTRACT We report a case of Macular Telangiectasia type 2 with crystalline retinopathy in a 42 year old female complaining of gradual decrease of vision in both eyes for the past one year. Both eye fundus showed perifoveal refractile crystals with lamellar macular holes. A diagnosis of crystalline retinopathy was made. Other causes of crystals were ruled out based on history, systemic examination, multimodal imaging & laboratory work-up. Presence of dilated perifoveal deep capillary plexus, foveal cavitation & temporal leakage on FFA confirmed association with Macular Telangiectasia. To our knowledge, there is no thorough documentation of crystalline reti-nopathy reported in Indian population.

KEYWORDS : Retinal Crystals, Macular Telangiectasia, OCT Angiography

INTRODUCTION:

Crystalline retinopathies may be associated with different etiologies including genetic, toxic, degenerative, idiopathic, and iatrogenic causes.[1] Their location may be preretinal, intraretinal, or subretinal spaces or in the choroid and may be scattered in the macula and posterior pole or throughout the retinal periphery.[2] Diagnosis can be made with a thorough history, meticulous ocular and retinal examination, and multimodal imaging techniques.

Retinal crystals may be present in 46% of patients with Idiopathic Macular Telangiectasia type 2 (MacTel 2), 60% being bilateral.[3]

MacTel is a vascular anomaly of the retinal capillaries affecting the temporal juxtafoveal region of the macula.[4] Based on biomicroscopic and fluorescein angiographic features, it is classified into 2 main group,[5] Type 1 (aneurysmal telangiectasia) is a Coat disease variant with male predilection. Type 2 (perifoveal telangiectasia) is associated with bilateral occult telangiectatic abnormalities at the level of the deep retinal capillary plexus.[6] Progressive macular abnormalities may develop including reduced retinal transparency, foveal cavitation, fine retinal crystal deposition, blunted right angle venules, and pigment plaques.

Case Report:

A 45 years old woman presented with gradual progressive loss of vision for distance and near for the past 1 year. Her best corrected visual acuity was 20/80 both eyes (OU). Intraocular pressure & anterior segment biomicroscopy was within normal limits.

Fundus examination showed presence of golden yellow refractile crystals in the peri-foveal region in an annular arrangement, more numerous temporally. Clinically bi-lateral lamellar macular holes were seen. (Fig.1)

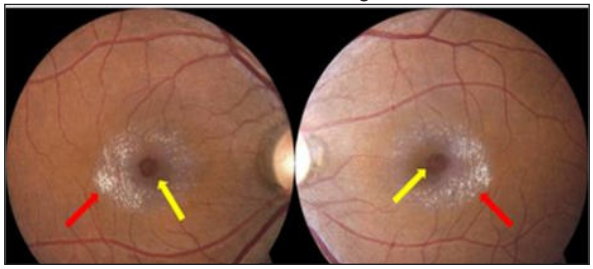


Figure 1 : Right(A) and Left(B) eye fundus showing retinal crystals (red arrow) and lamellar macular hole (yellow arrow)

On Optical Coherence Tomography (OCT), multiple punctate hyperreflective lesions were seen in the perifoveal nerve fibre layer. Foveal cavitaion was evident. (Fig.2)

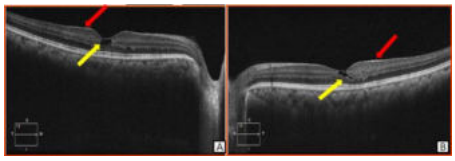


Figure 2: Right(A) and Left(B) eye OCT showing retinal crystals in nerve fibre layer (red arrow) and foveal cavitation (yellow arrow)

On Flourescein Angiography (FA), leakage was present in the temporal perifoveal region. (Fig.3)

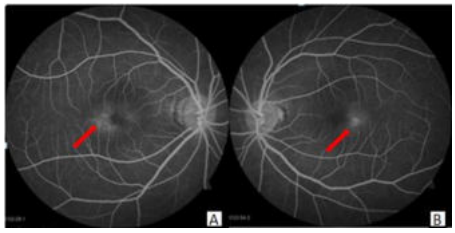
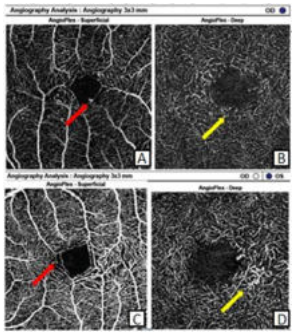


Figure 3 : Right(A) and Left(B) eye FA indicating temporal leakage from telangiectatic vessels (red arrow)

On Optical Coherence Tomography-Angiography (OCT-A) done with Cirrus Angio-plex(Zeiss), telangiectatic vessels were visualized in the perifoveal superficial and deep capillary plexus, more prominent temporally in the deep capillary plexus. (Fig.4)

Figure 4 : Both eye OCT-A showing dilated deep capillary plexus. (yellow arrow)(B,D) and less promi-nently dilated superficial capillary plexus. (red arrow)(A,C) . Changes are more marked in the left eye (C,D)



On Fundus Autoflourescence (FAF), there was no hyperautoflourescence Visual fields were normal.

Systemic examination was normal. There was no significant drug history. The pa-tient had no previous ocular pathology or surgery. A full lab work-up revealed no abnormality.

A diagnosis of MacTel 2 with crystalline retinopathy was made. Since there was no sub-foveal fluid on OCT, the patient was kept under observation. The findings remained stable on 3 and 6 month follow-up.

DISCUSSION :

Retinal crystals associated with Mac Tel 2 can be distinguished from drusen or hard exudates by their superficial location, color, size, and shape and varying from1-20 in number.[1] They can be found over or adjacent to the telangiectasis and don't correlate with the severity of the disease. The presence of crystals is noted to be associated with inner segment ellipsoid zone disruption and the number of crystals may correlate with the loss of retinal transparency, loss of macular pigment, and fluorescein leakage.[3]

Retinal examination may illustrate highly reflective, small white or golden yellow crystals distributed in an annular pattern of approximately 4000microns with central sparing of approximately 700microns.[3] A break corresponding to the horizontal raphe may be noted in the annular pattern of the crystals temporal to the fovea, with alignment of the crystals corresponding to the pattern of the nerve fiber layer.[3] Spectral domain OCT(SD-OCT) may demonstrate crystals within the nerve fiber layer. Cystic foveal cavitation is characteristic on SD-OCT, sometimes associated with foveal atrophy, and reduced foveal and parafoveal retinal thickness.[4] Focal atrophy of the foveolar retina may develop and lead to a lamellar or even a full thickness macular hole.

Crystals are not detected on FA or Indo Cyanine Green (ICG) imaging, although leakage from the telangiectatic macular capillaries is a characteristic feature on FA.[1]

Little is known of the origin and chemical composition of the crystals. The hypothesis that they may be composed of a retinoid originating in the visual cycle is based on their distribution and reflective properties, and the correlation between the number of crystals and presence of a break in the ellipsoid zone. [3] Muller cells play an important role in metabolism of retinoid. Histopathologically, Powner and coworkers identified a depletion of Müller cells and presence of vascular abnormalities in the areas of macular pigment loss in MacTel 2.[7]

Sjogren-Larrson syndrome (SLS) may share metabolic pathways with MacTel 2 based on similar ocular manifestations like arrangement of the crystals along the nerve fibers, central cystoid cavitation, and loss of luteal pigment.[8] Müller cell dysfunction is suggested to be associated with the retinal features of tamoxifen retinopathy as well, which include perifoveal retinal crystals and foveal cavitation.[8] Crystals in MacTel 2 are estimated to be larger than ganglion cell axon diameters and are located around the nerve fiber bundles, possibly within Müller cell foot-plates, similar to crystals in SLS and tamoxifen crystals.[3]

Clinical features of MacTel 2 include decreased visual acuity, metamorphopsia, paracentral scotoma, difficulty in reading, decreased visual quality.

No approved therapeutic modality is proven to be effective for the nonproliferative stage of Mac Tel 2. The proliferative stage of MacTel 2 may benefit from intravitreal injection of anti-Vascular Endothelial Growth Factor therapies.[9] Early treatment and a small size of the neovascular membrane may be associated with a better visual outcome.

CONCLUSION:

Crystalline Retinopathies have a diverse group of etiologies and manifestations. No treatment is required except for the underlying condition. To our knowledge, there is no documentation of crystalline retinopathy reported in Indian population .Our case report aims at providing better understanding of the same.

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