PSEUDODOUBLING OF OPTIC DISC WITH BILATERAL SERPIGINOUS CHOROIDITIS AND MACULAR CHOROIDAL NEOVASCULARISATION- A CASE REPORT

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PRESENTATION OF CASE

We present here a case of pseudodoubling of the optic disk- a spectacular and rare clinical presentation. Of additional interest is that this case also includes another rare presentation, which is serpiginous choroiditis associated with choroidal neovascularisation. Patient was given a course of oral prednisolone and planned for intravitreal anti-VEGF. Poor visual prognosis has been explained.

A 46 years old man came to us with gradually decreasing vision in both eyes for last 8 months. On examination, his unaided visual acuity for distance was 3/60 in right eye and 3/60 in left eye. On pin hole, it reduced to 1/60 in both eyes. Pupil was normal size, but sluggishly reacting in both the eyes. Fundus examination revealed clear media and bilateral grey or creamy yellow placoid lesions in peripapillary region and extending centrifugally with finger like or serpentine projections. The lesions also show retinochoroidal degeneration and pigmentation involving the macula (Figure 1, 2). Left eye had signs of inciting active lesion inferior to the disc.

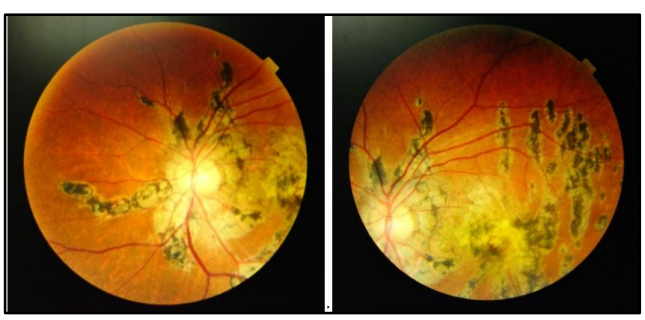

Figure 1. Left Eye Serpiginous Choroiditis Involving the Macula

In the right eye, there was a large dull white circular patch of size 1DD in between optic disc and fovea. Blood vessels were feeding its centre and it seemed that vessels are arising from it. A large flame-shaped haemorrhage was also present superiorly resembling splinter haemorrhage of disc. It resembled another optic disc and was more confusing when seen isolated (Figure 3). There was no associated vasculitis or disc neovascularisation.

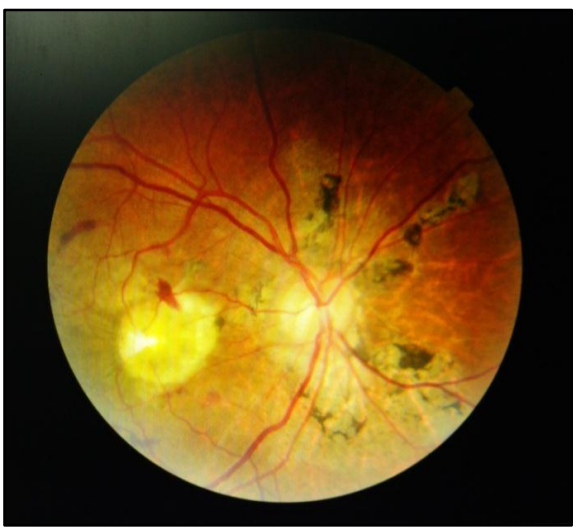

Figure 2. Right Eye Serpiginous Choroiditis

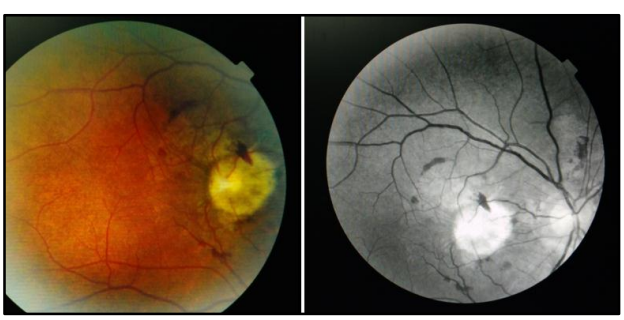

Figure 3. Right Eye Coloured Fundus and Red Free Photo of Pseudodoubling of Optic Disc with Adjacent Haemorrhage

Spectral Domain Optical Coherence Tomography Findings of Right Eye

HD 5 line raster of right eye (Figure 4) revealed macular oedema of 794 micron and dome-shaped elevation of RPE of 389 microns. So, the central ILM-RPE thickness was around 405 microns (794-389 micron). It was due to Choroidal Neovascularisation (CNV) invading the retinal layers. A traction band was present connecting to it. There was loss of IS-OS junction at several parts signifying photoreceptor loss. There was increased reflectance of the choroid and the deeper retinal layers.

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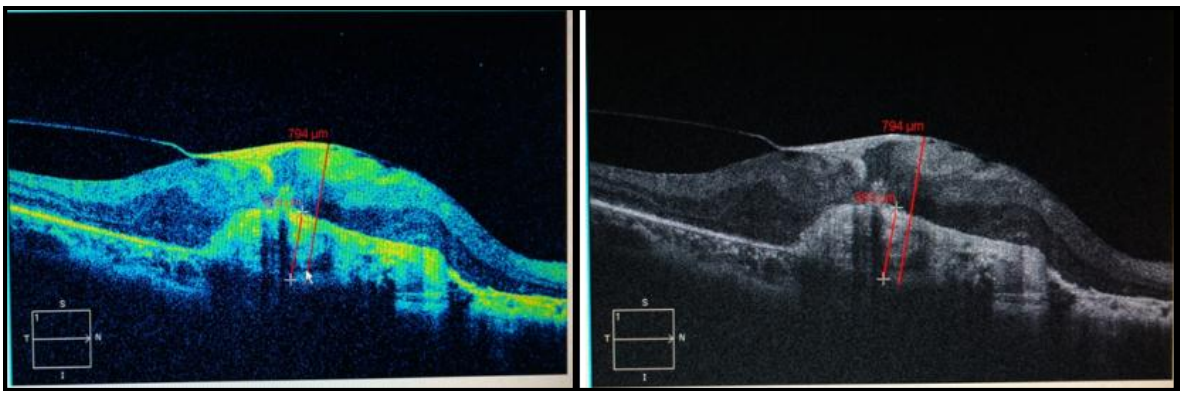

Figure 4. HD 5 Line Raster of Right Eye

Macular cube 512*128 analysis for macular thickness (Figure 5) revealed central subfield thickness (ILM-RPE) of 446 micron, thus nearly confirming the HD 5 line raster findings.

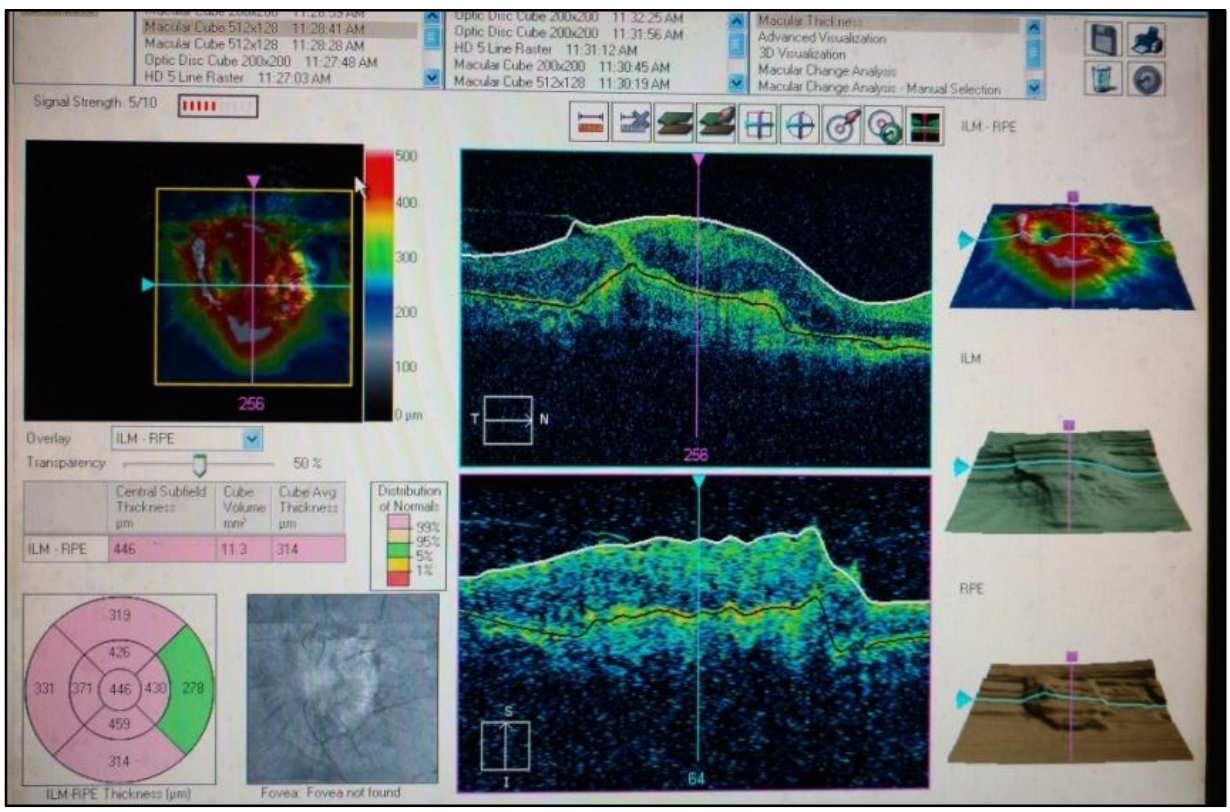

Figure 5. Macular Cube 512*128 Analysis- Macular Thickness

Macular cube 512*128 analysis 3D visualisation (Figure 6) progressive cut section showed macular oedema.

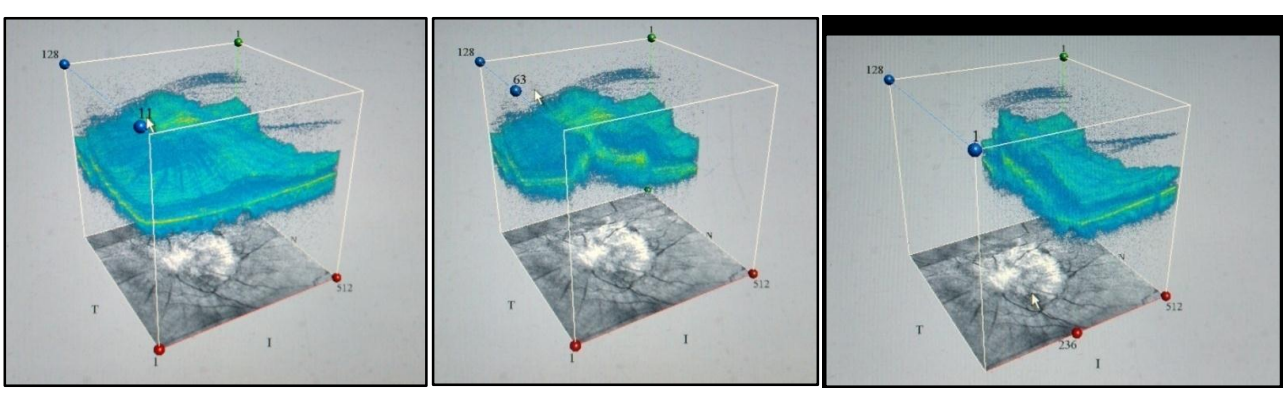

Figure 6. Macular Cube 512*128 Analysis 3D Visualisation

Spectral Domain Optical Coherence Tomography Findings of Left Eye

HD 5 line raster of right eye (Figure 7) revealed macular oedema of 342 microns with an impending complete posterior vitreous detachment. Now, it is attached to the macula leading to traction and causing oedema. The RPE was also not regular with small bumps, probably initial stage of choroidal neovascularisation.

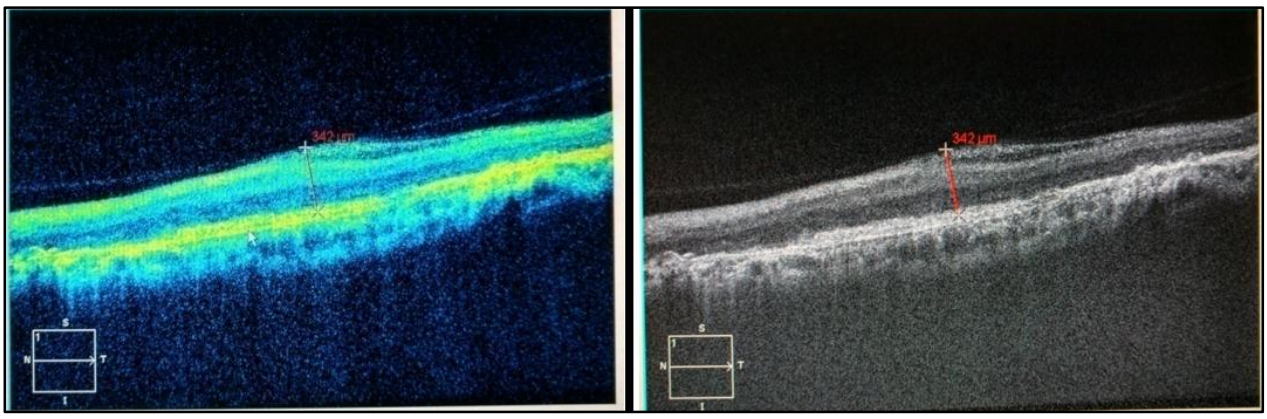

Figure 7. HD 5 Line Raster of Right Eye

Macular cube 512*128 analysis for macular thickness (Figure 8) revealed central subfield thickness (ILM-RPE) of 270 microns, progressive scarring at perifoveal area and relatively less scarring at inferior and nasal quadrant.

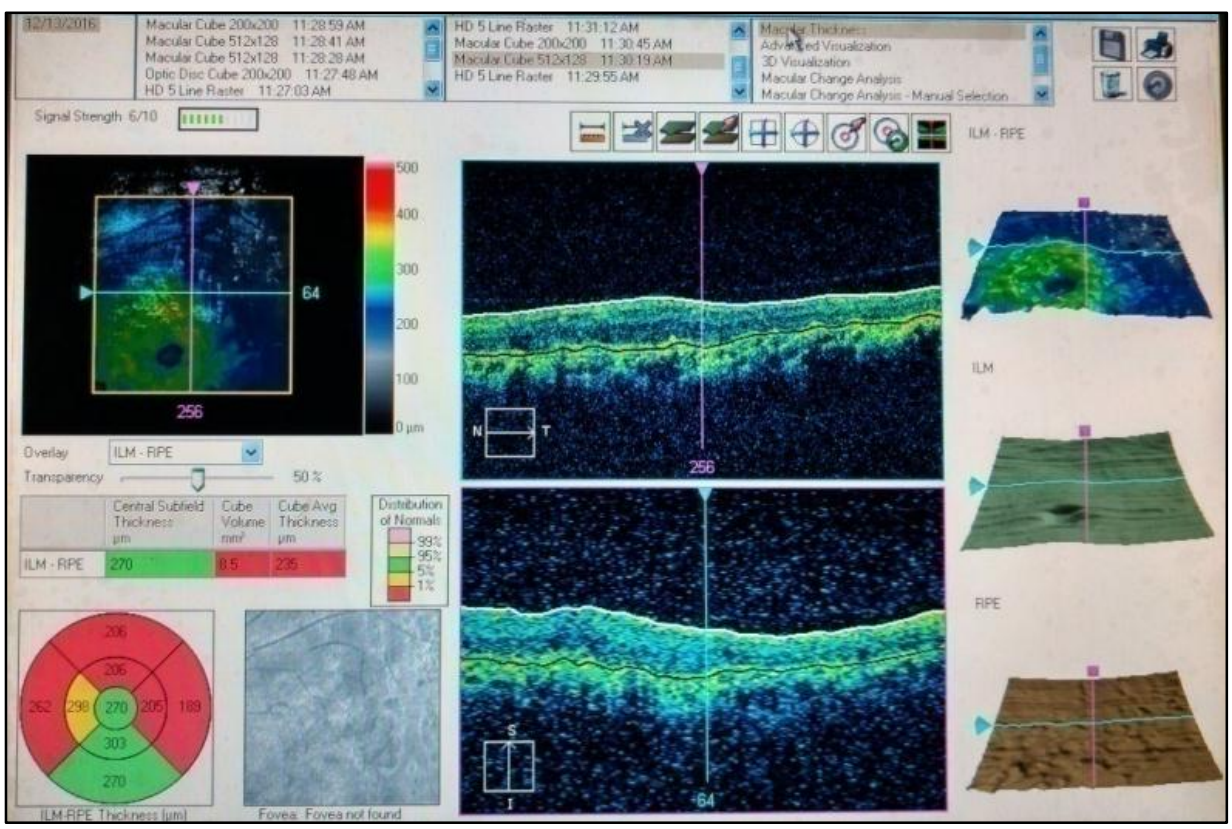

Figure 8. Macular Cube 512*128 Analysis for Macular Thickness

Macular cube 512*128 analysis 3D visualisation (Figure 9) progressive cut section showed relatively less scarring at inferior and nasal quadrant.

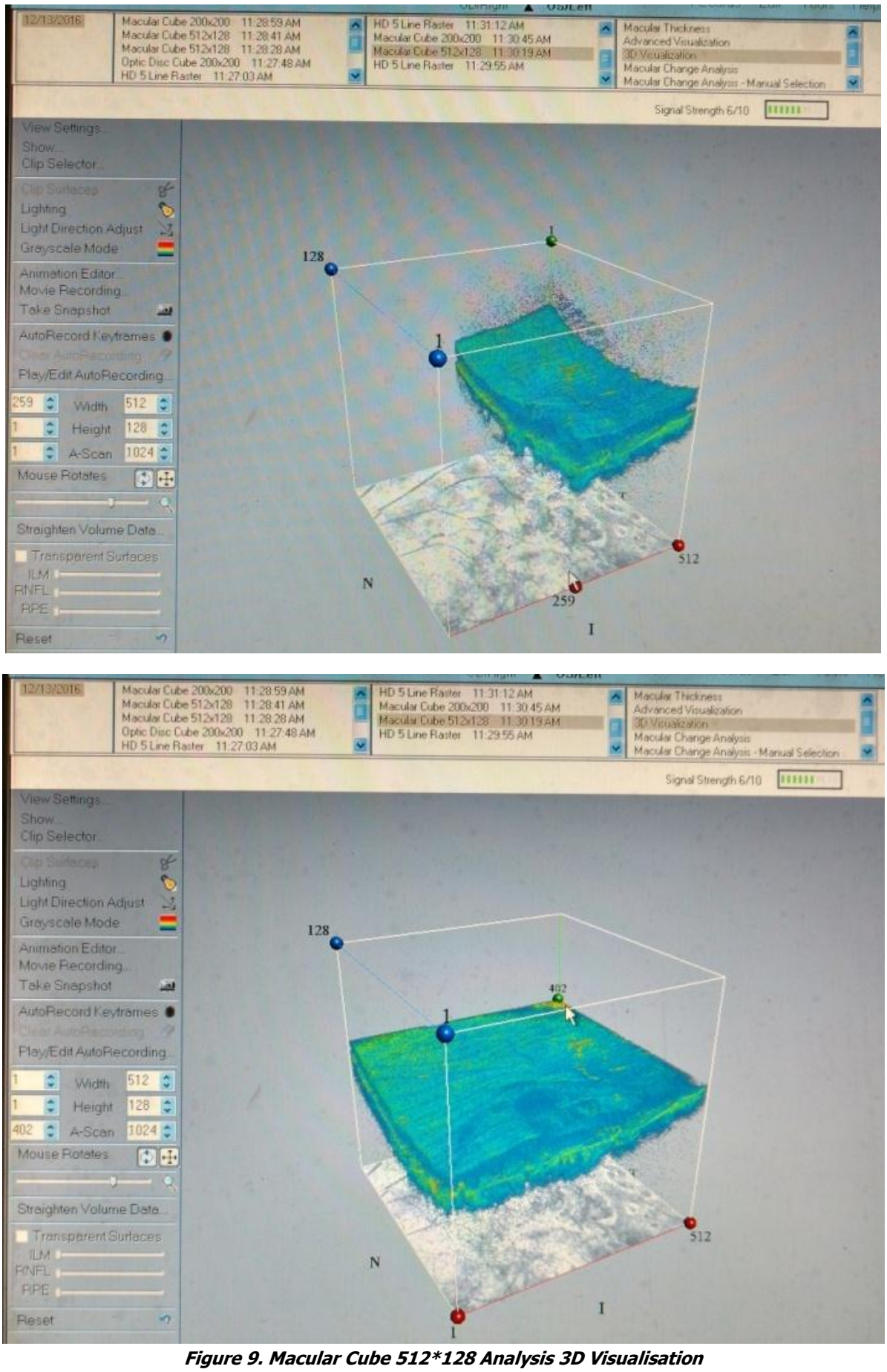

Figure 9. Macular Cube 512*128 Analysis 3D Visualisation

Visual field analysis of (central 30-2 threshold test) right eye (Figure 10) showed severely depressed sensitivity at optic disc and macular area. VFI of 71% and MD-9.21 dB with P<0.5% signifies large amount of RNFL loss and generalised depressed sensitivity and PSD 13.28 dB with P<0.5% shows a localised severely depressed area.

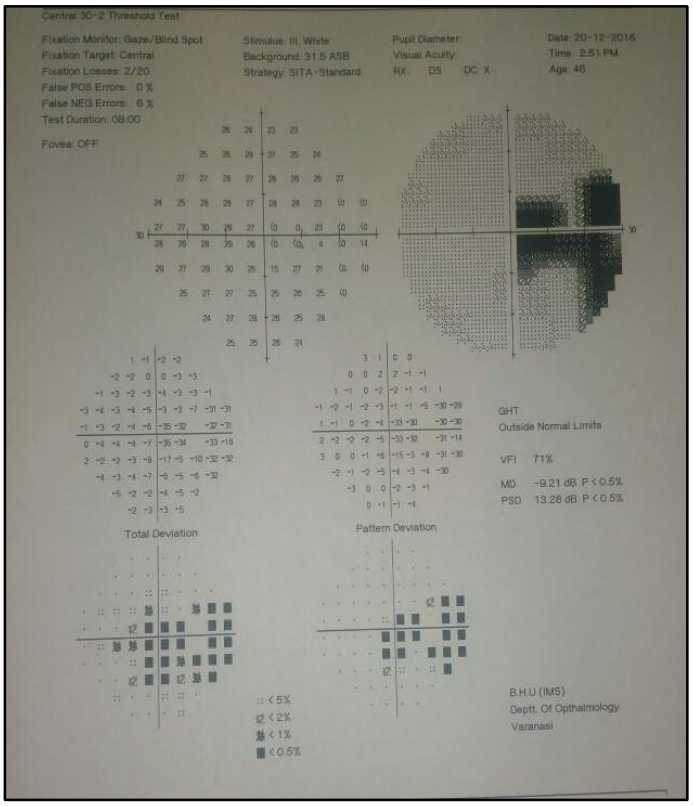

Figure 10. Visual Field Analysis of Right Eye

Visual field analysis (central 30-2 threshold test) of left eye (Figure 11) showed severely depressed sensitivity at optic disc and macular area. VFI of 65% and MD-10.09 dB with P<0.5% signifies large amount of RNFL loss and generalised depressed sensitivity and PSD 11.58 dB with P<0.5% shows a localised severely depressed area.

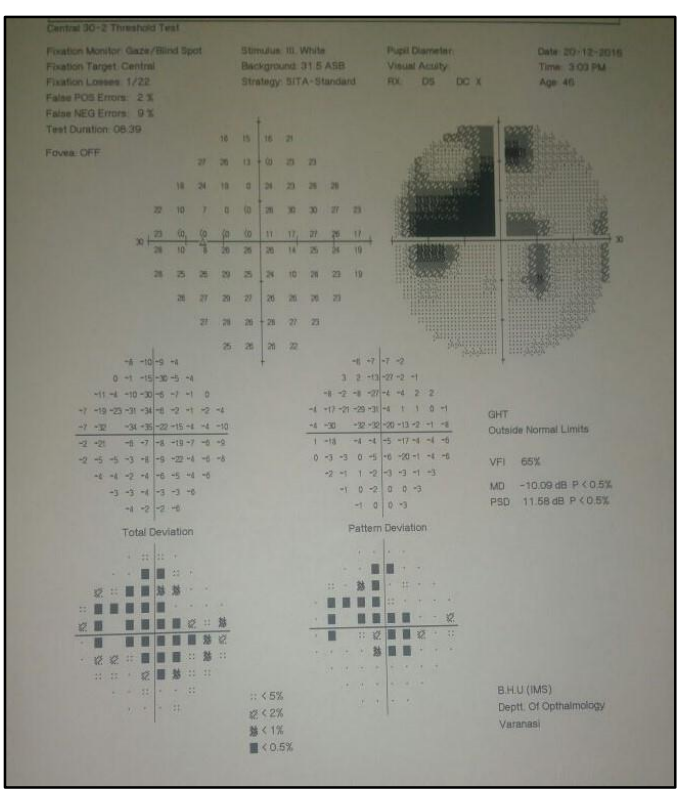

Figure 11. Visual Field Analysis of Left Eye

DIFFERENTIAL DIAGNOSES

1. Bilateral serpiginous choroiditis- Provisional diagnosis.
2. Acute posterior multifocal placoid pigment epitheliopathy- It occurs in younger age group and has bilateral presentation. CNV is a rare occurrence and visual prognosis is better.
3. Birdshot choroidopathy- It has a strong association with HLA A-29. The lesions are ovoid with nasal predominance and macula is spared usually.
4. Tuberculous choroiditis- It has multifocal lesions involving the periphery.
5. Toxoplasmosis.
6. Sarcoidosis.
7. Syphilis.

CLINICAL DIAGNOSIS

Right eye pseudodoubling of optic disc with bilateral serpiginous choroiditis and right eye macular choroidal neovascularisation with signs of inciting active lesion in left eye.

CLINICAL DISCUSSION

Pseudodoubling of the optic nerve head is a rare condition where a lesion resembling an optic disc is situated adjacent to the true optic disc.[1] This pseudoduplication of optic discs maybe caused by lesions such as peripapillary chorioretinal coloboma, optic disc coloboma or inflammatory foci (in our case). Rare cases of true duplication of optic discs with separation of optic nerve into two or more strands have been reported.[2] Pseudodoubling of the optic discs caused by lesions such as optic disc coloboma, peripapillary chorioretinal coloboma or inflammatory foci (in our case) are much more common.

Serpiginous Choroiditis (SC) is also known as helicoid peripapillary chorioretinal degeneration. It is a rare disease of unknown aetiology. It is usually bilateral, chronic and progressive condition. SC is a disease of healthy individuals. The onset is usually between the ages of 30 and 70 years. However, cases of SC have been reported in younger individuals.[3] The literature reflects a slight predilection for males.[4] The lesions involve the outer retina, RPE, choriocapillaris and large choroidal vessels. Patients are initially asymptomatic until the macula is involved and there is obvious decrease in visual acuity. The patient usually presents with unilateral decreased visual acuity due to asymmetrical involvement of the eyes. SC can present as two forms.

Classic type (80% cases), which starts with geographic patches of gray or creamy yellow placoid lesions in the peripapillary region. It generally progresses in a centrifugal manner with finger-like or serpentine projections.

Macular type is the second variant where the fovea is affected first. There is generally a poorer prognosis for the macular variant as the fovea is affected at the start. Vision loss may also be due to presence of Choroidal Neovascularisation (CNV).[5] CNV may affect 13-20% of eyes. SC has recurrences and therefore it is common to see lesions of different stages. Macular SC maybe misdiagnosed as macular degeneration, macular dystrophies or toxoplasmosis. Healing of individual lesions occur in 2 to 8 weeks. Generally, acute lesions are seen in one eye at a time.

Spectral domain OCT will show retinal atrophy with disruption of the photoreceptor layers in affected areas. There is thinning of the RPE and there is increased reflectance of the choroid and the deeper retinal layers. Disruption of the photoreceptor IS-OS junction is seen in active and inactive lesions. Steroids are mainstay of treatment.

DISCUSSION OF MANAGEMENT

Steroids are Mainstay of Treatment

Patient was prescribed oral prednisolone 60 mg/day with a tapering dose and was screened for TB (Mantoux test), septic foci (chest x-ray), syphilis (VDRL) and toxoplasma (IgG titre), which came negative. Poor visual prognosis has been explained to the patient and planned for intravitreal anti-VEGF (ranibizumab) injection to combat choroidal neovascularisation.

FINAL DIAGNOSIS

Right eye pseudodoubling of optic disc with bilateral serpiginous choroiditis and macular choroidal neovascularisation (re>>>le) with signs of inciting active lesion in left eye.

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