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Short Communication

Cone Rod Dystrophy

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Cone rod dystrophies (CRDs) (prevalence 1/40,000) are inherited retinal dystrophies that belong to the group of pigmentary retinopathies. CRDs are characterized by retinal pigment deposits visible on fundus examination, predominantly localized to the macular region. In contrast to typical retinitis pigmentosa (RP), also called the rod cone dystrophies (RCDs) resulting from the primary loss in rod photoreceptors and later followed by the secondary loss in cone photoreceptors, CRDs reflect the opposite sequence of events. CRD is characterized by primary cone involvement, or, sometimes, by concomitant loss of both cones and rods that explains the predominant symptoms of CRDs: decreased visual acuity, color vision defects, photoaversion and decreased sensitivity in the central visual field, later followed by progressive loss in peripheral vision and night blindness.

History

A 21-year-old male patient presented with progressive loss of vision over the past years. The patient had history of reduced vision in dim light with a specific loss in the central field. He had no known precipitating factor and no family history of similar symptoms. The patient had been prescribed glasses in the past but they did not provide much improvement in vision.

Examination

On examination, the unaided visual acuity in right and left eye was 1/60 and 2/60 respectively. Dilated retinoscopy was performed and we found some degree of improvement with refraction. Anterior Segment examination appeared normal and the papillary reactions were brisk. Details of the refraction and anterior segment evaluation are listed in table 1.

	Right eye	Left eye
Unaided VA	1/60	2/60
Pinhole	1/60, NIF	2/60, NIF
Subjective Acceptance	-8.00/-2.00*170	-6.00/-2.00*180
BCVA	5/60, N36@15cm	5/60, N36@15cm
Lids	Flat	Flat
Conjunctiva	Quiet	Quiet
Cornea	Clear, nystagmus	Clear, nystagmus
AC	Clear, no cells and flares	Clear, no cells and flares
Pupil	PERRLA	PERRLA
Lens	Clear	Clear

Table 1

Patient was referred to Retina Specialist. VEP was performed, dilated fundus examination was performed and OCT, Fundus Photography was advised to him.

VEP revealed low amplitude pattern and flash VER in both eyes.

Figure 1

OCT examination revealed.

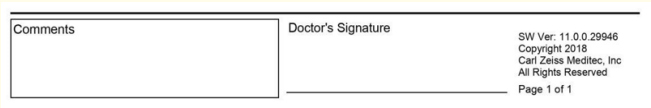

Figure 2

Fundus examination revealed.

Figure 3

The patient was diagnosed to have Hereditary Maculopathy with Cone Rod Dystrophy in both the eyes and explained that the visual prognosis was guarded. In view of no definite treatment for the condition, the patient was referred to the LOW VISION CLINIC for assessment. While assessing we observed the patient was depressed and hopeless due to his condition. He left his study too 3 years ago.

Low vision history

Disability in distance and near task

- Difficulty seeing TV.
- Able to recognize face.
- Able to read text.

- Not able to see sign board.
- Able to pick up things from desk.

Disability in mobility

- Independent in known environment.
- Dependent in unknown environment.
- Not able to cross road independently.

Glare problem

On assessment we found a fair vision improvement in both the eyes with the low vision aids.

Low vision device trial

Distance

- Monocular Spectacle Mounted Telescope = No Improvement due to nystagmus.
- SEE TV 2.1X Binocular Telescope = 6/18**.**

Near

- Hand held Illuminated Magnifier = N18 @ 20 cm ENGLISH
- Video Magnifier (5") = N10 @ 20 cm ENGLISH.

SEE TV 2.1X Binocular Spectacle was advised for the patient. Training on using the device was too given.