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EVALUATION OF SUBCLINICAL POSTERIOR SEGMENT INVOLVEMENT IN MUCOPOLYSACCHARIDOS TYPE II- HUNTER SYNDROME BY SPECTRAL-DOMAIN OPTICAL COHERENCE TOMOGRAPHY

Poster

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Purpose:

Mucopolysaccharidosis type II, also known as Hunter Syndrome, results in accumulation of dermatan sulfate and heparan sulfate. Patients with Hunter syndrome typically have clear corneas, but cystoid changes in the macula and clinically pigmentary retinopathy have been reported.

Methods:

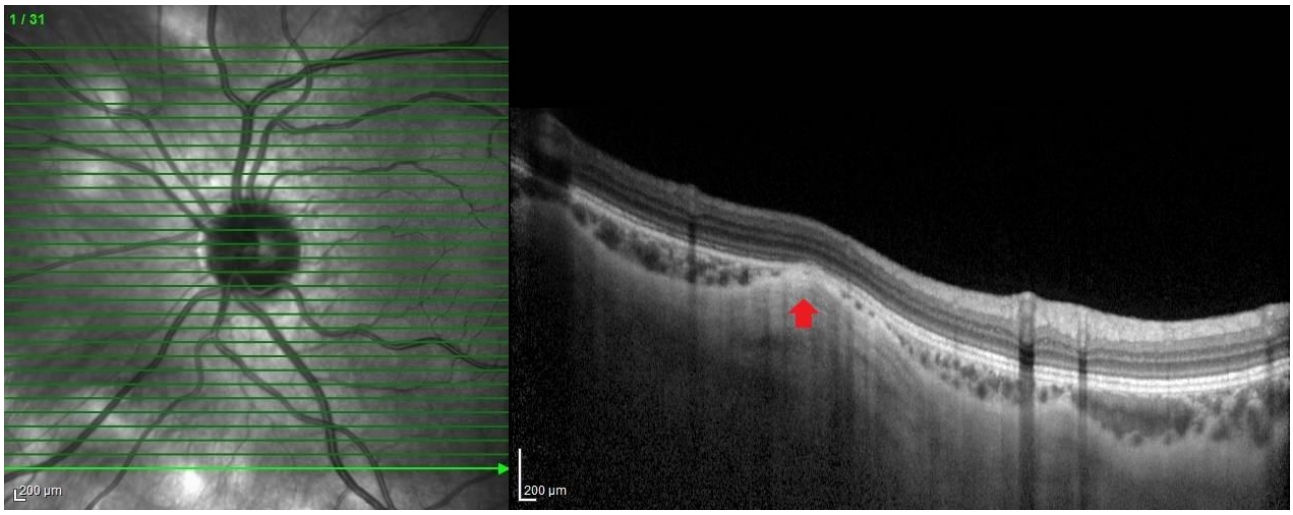
Ocular examination and spectral domain OCT (Heidelberg Spectralis) findings of an asymptomatic 12 year old Hunter case are presented.

Results:

A 12 year old male patient, diagnosed with Hunter syndrome at the age of 5 and followed up with enzyme replacement therapy, had bilateral visual acuity of 0.8 with normal anterior segments. Fundus examination revealed hypopigmented areas nasally. Optical coherence tomography (OCT) showed marked thickening of the external limiting membrane at the macula, when compared to the maculae of same aged children. EDI-OCT showed focal thickening of the sclera with prominent compression of the choroid at the site of nasal hypopigmented area, however the RPE seemed intact.

Conclusions:

In pre-OCT time, hypopigmentation of fundus in Hunter Syndrome was thought to be due to RPE atrophy. In our case, EDI-OCT revealed focal scleral thickening and choroidal compression with no overlying RPE changes. SD-OCT is an important aid in evaluating subclinical ocular changes in the early stages of Hunter syndrome.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