Gyrate Atrophy: A Photo Essay of Progressive Chorioretinal Degeneration

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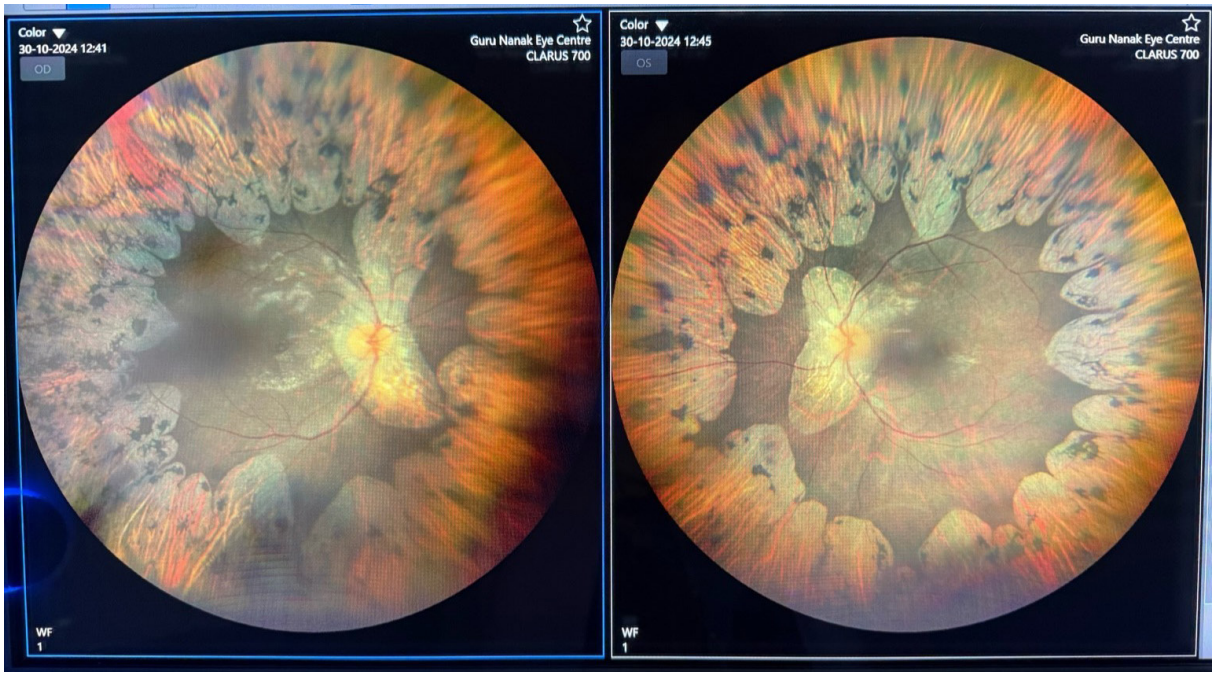
Abstract

Introduction: Gyrate atrophy is a rare autosomal recessive chorioretinal dystrophy caused by a deficiency of the mitochondrial enzyme ornithine aminotransferase, leading to elevated plasma ornithine levels and progressive vision loss. This photo essay presents a series of fundus images demonstrating the classical features of gyrate atrophy, including scalloped areas of chorioretinal atrophy and their centrifugal progression.

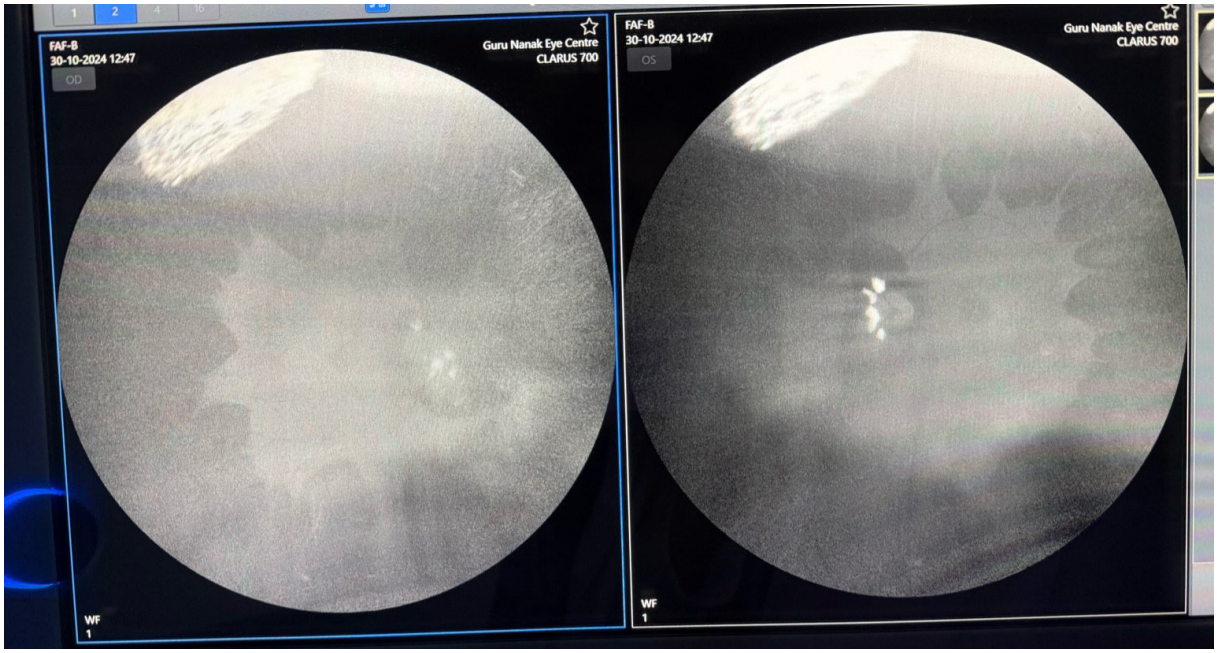
Case Summary: A 22year old presented with progressive night blindness and peripheral vision loss. His sister also has similar complaints. BCVA in both eyes were finger counting at 2 meters, the intraocular pressure in right eye was 9 and the left eye was 11mmHg. Anterior segment examination in both eyes were within normal limits. Fundus examination revealed both eye normal optic disc with peripapillary atrophy, foveal reflex dull and an attached retina. There were bilaterally symmetrical, sharply demarcated areas of chorioretinal atrophy beginning in the mid-periphery with gradual centripetal spread. OCT in both eyes reveal distorted foveal contour with CMT 428 micrometer in right and 515 micrometer in left eye and ERM. Fundus autofluorescence reveal well defined areas of hypo autofluorescence corresponding to regions of complete RPE and choriocapillaris atrophy with macular sparing. The FAF pattern is bilateral and symmetrical.Plasma ornithine levels were significantly elevated, confirming the diagnosis of gyrate atrophy.

Photographic Documentation:

- Right eye – Fundus photograph: Shows multiple sharply demarcated, scalloped areas of chorioretinal atrophy predominantly in the mid-periphery with sparing of the central macula.
- Left eye – Fundus photograph: Similar to the right eye, revealing symmetrical distribution of lesions.



- Wide-field composite image: Highlights the full extent of peripheral degeneration.
- OCT scan: Demonstrates
- FFA



Discussion: Gyrate atrophy is characterized by the progressive loss of choriocapillaris, RPE, and photoreceptors. Patients typically present in the first two decades of life with nyctalopia and field constriction. Diagnosis is confirmed by elevated plasma ornithine levels and genetic testing (mutations in the OAT gene). Treatment with vitamin B6 supplementation (in responsive patients) and an arginine-restricted, low-protein diet can delay progression. Fundus photography remains a vital tool in documenting disease extent and progression.

Conclusion: This photo essay illustrates the classical fundus findings of gyrate atrophy and underscores the importance of early recognition for appropriate management. Periodic fundus imaging plays a crucial role in monitoring disease progression and therapeutic response.

Acknowledgement

Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given their consent for images and clinical information to be reported in the journal. The patients understand that names and initials will not be published and efforts will be made to conceal their identity.

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