Photo essay

Bilateral Congenital Ectropion Uveae and Glaucoma

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Abstract *The presence of congenital ectropion uveae (CEU) in a patient calls for a definitive work-up for underlying glaucoma as the latter if missed can have potentially blinding sequel. Most patients may not be aware of the presence of CEU as far as third to fourth decade and may have lost significant proportion of their visual fields at their first ophthalmic consultation. A case of bilateral CEU with bilateral glaucoma is described emphasizing this association.*

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VA 32-year-old lady presented for a routine ophthalmic examination. Best corrected visual acuity with -0.50 diopter spheres was 6/9 in both eyes (BE). Circumferential proliferation of the pigment epithelium of iris was seen anteriorly over the pupillary margin associated with its patchy atrophy and loss of iris pattern in BE. This was suggestive of a congenital ectropion uveae (CEU), a non progressive neural crest cell disorder as the eyes lacked secondary signs of inflammation or ischaemia (Figure A, B).[1] Further evaluation confirmed an underlying diagnosis of juvenile open angle glaucoma based on her intraocular pressure (IOP, Right eye-24 mm Hg and LE- 26 mm Hg), Central corneal thickness (right eye- 530µm, Left eye- 535 µm), open angles and anteriorly inserted iris on gonioscopy, demonstration of arcuate scotoma on visual field charting, absence of a pupillary membrane, and an enlarged cup to optic disc ratio of 0.6:1 BE. Target IOP was achieved with treatment with topical timolol and brimonidine eye drops that stabilized her visual fields. CEU is characteristically unilateral with ipsilateral glaucoma, iris hypoplasia and iridotrabecular dysgenesis. Literature also related CEU with ocular abnormalities as coloboma, telecanthus, thickened corneal nerves. Bilateral presentation of CEU with glaucoma is unusual and in any case only a few case reports exist in literature.[2] Most studies have proposed the association between appearances of progressive open-angle glaucoma due to angle dysgenesis which seem to progress over time.[3,4,5] Angle closure glaucoma is also described. Congenital ectropion uveae (CEU) is a marker for underlying glaucoma. Congenital defects, neurofibromatosis type1 and genetic diseases are related. Genetic predetermination has not yet been verified. Medical management is the first line of treatment and works well. Progressive glaucoma not controlled with pharmacotherapy needs surgery, often multiple. Differential diagnosis include iris flocculi (in bilateral cases, ACTA2 Mutation for Familial Thoracic Aortic Aneurysms and Dissections should be searched for), or bilateral pupillary margin epithelial pigmentary cysts. Early

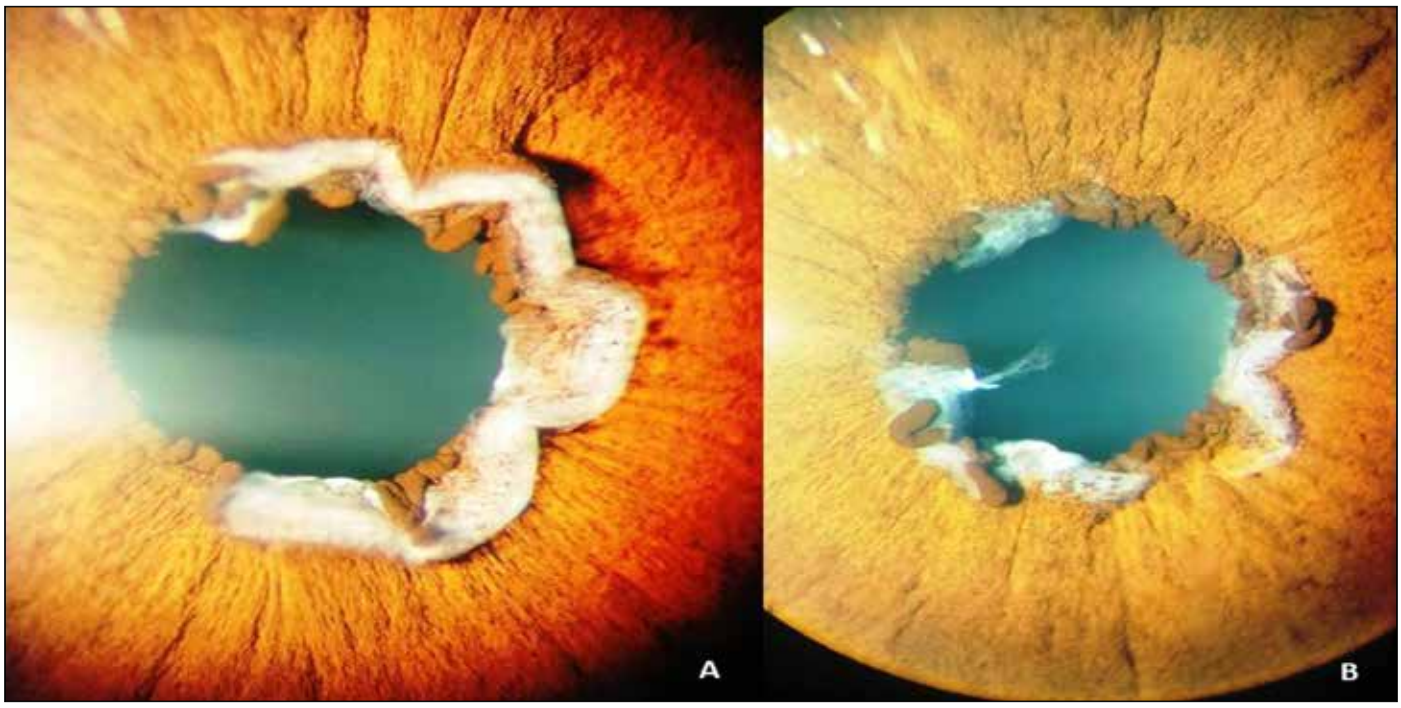

Figure 1: *Massive ectropion uveae visible as circumferential proliferation of pigment epithelium of the iris anteriorly over the margin of the pupil accompanied with white appearing patches of atrophy of this proliferating epithelium (Figure A {RE}, B {LE}). Loss of iris pattern with the absence of collarette was obvious (Figure A, B).*

recognition of CEU prevents blindness due to glaucoma in young patients and improves their prognosis.

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