Pupillary ruff hypertrophy with secondary open-angle glaucoma

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DESCRIPTION

We present a case of a woman in her early 50s, who presented with complaints of diminution of vision in the left eye (L/E) which was gradual, painless and progressive. On clinical examination, visual acuity was 6/6 and 6/12 in the right eye (R/E) and L/E, respectively. Intraocular pressure (IOP) was 16 and 22 mm Hg in the R/E and L/E, respectively (on eye drop Travoprost HS and brimonidine+timolol combination). On slit lamp examination, pupillary ruff hypertrophy was present in both eyes but much more severe in the L/E than in the R/E (figures 1A and 2A). We also observed that hypertrophied pigment clumps were detached from the pupillary border and deposited at various levels on the anterior surface of the iris (figures 1A and 2A). On gonioscopy, we noticed detached pigment deposits in angle in the L/E (figure 2C) with open angles in three quadrants in both eyes. The R/E angle was normal (figure 1C). Optic disc evaluation showed 0.9 and 0.6 cup to disc ratio (CDR) in the L/E and R/E, respectively. The patient had increased IOP, CDR and advanced glaucoma only in the L/E. This was an unusual presentation of unilateral advanced glaucoma with pupillary ruff hypertrophy and pigment deposits on the iris surface and angle. We further investigated the patient and did anterior segment-optical coherence tomography to rule out the cystic nature of pigment deposits, which revealed multiple solid lesions in the L/E (figure 2B) but only a single deposit on the iris surface in the R/E (figure 1B). Retinal nerve fibre layer optical coherence tomography (RNFL-OCT) was done which revealed severe thinning in the L/E but normal RNFL thickness in the R/E (figures 1D and 2D). Earlier we made a differential diagnosis of combined mechanism glaucoma as per the study by Sihota *et al*[1] in the L/E but R/E still had normal RNFL and non-glaucomatous cupping. We believe that the rise in IOP and development of glaucoma in the L/E, in this case, was due to multiple pigment deposits in angle and came with another differential diagnosis of 'pupillary ruff hypertrophy with secondary open-angle glaucoma'.

In a literature search, Bansal *et al* reported two cases of primary iris pigment epithelial hyperplasia with glaucoma. Both the cases in this report had a loss of normal iris architecture, pupillary abnormalities and extensive hyperplasia of the iris pigment epithelium. This iris pigment was irregularly overlayed on the iris stroma but did not reach the angle,[2] whereas in our case there was pupillary ruff hypertrophy and pigment deposits on the iris surface and angle. Another study by Ang *et al* found a positive correlation between asymmetric pupillary ruff changes with asymmetry in both IOP and CDR, which suggested that pupillary ruff atrophy is a risk factor for glaucoma.[3] Our case is an unusual presentation as pupillary ruff hypertrophy is associated with unilateral advanced glaucoma.

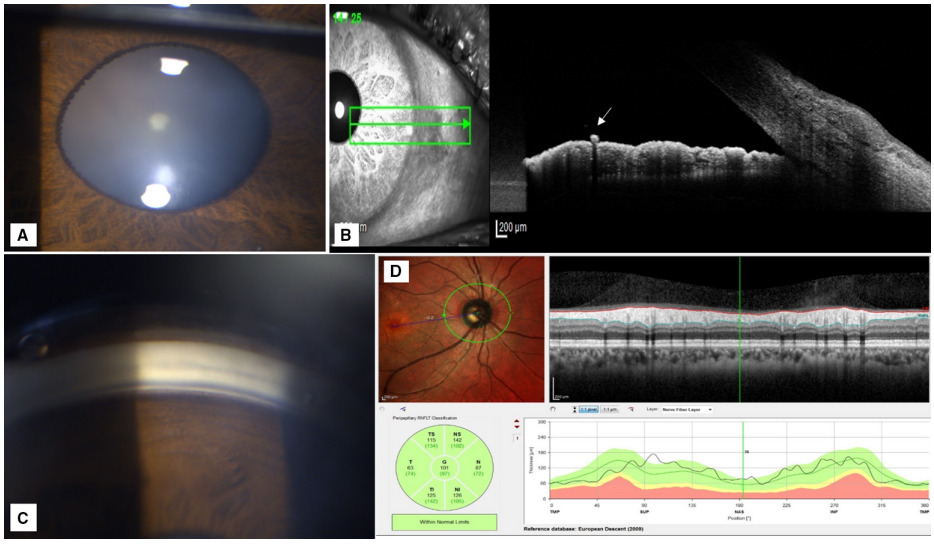

Figure 1 (A) Clinical photo of the right eye (R/E) showing mild pupillary ruff hypertrophy. (B) Anterior segment-optical coherence tomography of the R/E showing a single pigment deposit on the iris surface. (C) Gonioscopy of the R/E showing the open angle. (D) Retinal nerve fibre layer (RNFL) optical coherence tomography of the R/E showing normal RNFL thickness.

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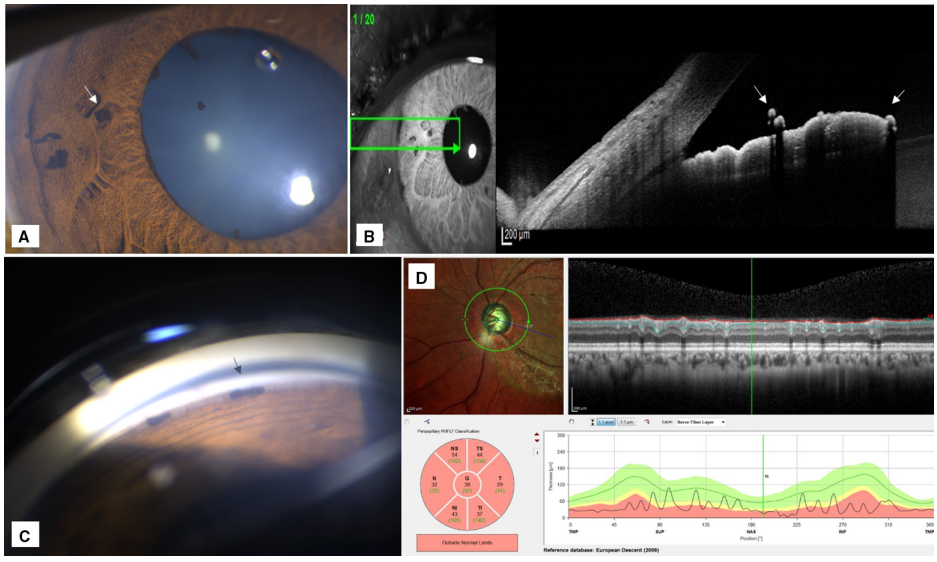

Figure 2 (A) Clinical photo of the left eye (L/E) showing severe pupillary ruff hypertrophy with detached pigment clumps deposited on the iris surface. (B) Anterior segment-optical coherence tomography shows multiple solid pigment deposits at the pupil and on the anterior surface of the iris. (C) Gonioscopy of the L/E showing large pigment deposits in angle. (D) Retinal nerve fibre layer (RNFL) optical coherence tomography of the L/E showing severe RNFL thinning.

Learning points

- There can be increased risk of glaucoma development in patients with pupillary ruff hypertrophy as seen in our case.
- Any such patient should be examined thoroughly at first visit only for early detection and treatment of glaucoma, also they should be kept on regular follow-up even if no glaucoma at presentation.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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