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Case report

Pigment dispersion syndrome presenting as endothelial dystrophy: An atypical presentation

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Abstract

Purpose: To describe an atypical presentation of pigment dispersion syndrome (PDS) with diffuse, homogeneous pigment deposition on the corneal endothelium and its management.

Methods: A 44-year-old female was referred to a cornea clinic as a case of endothelial dystrophy. Slit-lamp examination revealed bilateral, diffuse, and homogeneous pigment deposition on entire corneal endothelium without any iris transillumination defects. Intraocular pressure (IOP) at presentation were 18 mmHg OD and 16 mmHg OS. Gonioscopy showed dense, homogeneous pigment deposition in the angles. The optic nerve head examination revealed a cup disc ratio of 0.6:1–0.7:1 in both eyes with neuroretinal rim thinning.

Results: Peripheral Nd:YAG laser peripheral iridotomy (PI) was performed. On follow-up, a localized clear pigment free endothelial area was noted over the iridotomy sites bilaterally. IOP was well controlled within 12–14 mmHg with prostaglandin analogue at last follow-up of 24 months.

Conclusions: Diffuse homogeneous pigment dispersion on the endothelium may occur in atypical cases of PDS which may clear in the areas overlying the PI site and, therefore, should not be confused with endothelial disease. This case demonstrates the significance of a thorough clinical evaluation in cases with unusual presentation.

Keywords: Pigment dispersion syndrome; Atypical pigment dispersion syndrome; Endothelial dystrophy

Introduction

Pigment dispersion syndrome (PDS) is a bilateral disorder characterized by concave iris, Krukenberg's spindle, mid peripheral iris transillumination defects, and heavy pigment deposition in trabecular meshwork. Pigments released due to mechanical rubbing of posterior iris surface against the lens zonules are redistributed and deposited in the trabecular meshwork, thus obstructing the aqueous outflow and causing glaucoma. Krukenberg's spindle is a localized vertical collection of pigment granules, most commonly on inferior corneal endothelium.[1,2] It is unusual for the pigments to get dispersed homogeneously over the entire corneal endothelium in a case of PDS. Herein, we describe a case of PDS with unusual presentation of diffuse pigment deposition on the endothelium and misdiagnosed as an endothelial dystrophy.

Case report

A 44-year-old female was referred to the cornea clinic as a case of endothelial dystrophy diagnosed from outside. There

Conflict of interest: None.

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was neither any family history nor any symptoms of pain or redness or previous trauma. Her best corrected visual acuity (BCVA) at presentation was 20/20 bilaterally.

Slit-lamp examination revealed bilateral, diffuse, and homogeneous pigment deposition on corneal endothelium but more so inferiorly (Fig. 1, top). However, no iris transillumination defects were noted. Subsequently, she was referred to the glaucoma clinic for further evaluation. Intraocular pressure (IOP) at presentation was 18 mmHg OD and 16 mmHg OS. The central corneal thickness was 562 μm in the right eye and 564 μm in the left eye. Gonioscopy showed angles with dense, homogeneous pigment deposition in trabecular meshwork. On dynamic gonioscopy, concave iris configuration was appreciated (Fig. 1, bottom). The optic nerve head examination revealed a cup disc ratio of 0.6:1–0.7:1 with neuroretinal rim thinning bilaterally.

Endothelial dystrophy and copper-related toxicity were the two primary differential diagnosis. Specular microscopy showed a cell count of >3000/mm^2 in both eyes. Anterior segment optical coherence tomography (ASOCT) and ultrasound biomicroscopy (UBM) showed relatively narrow angles with slightly concave iris configuration in supine position (Fig. 2, top left and right). Humphrey visual field examination revealed no field defect. Confocal microscopy of the endothelium showed multiple hyper-reflective spots suggestive of pigments (Fig. 2, bottom left). Laboratory examination revealed normal serum ceruloplasmin (24.40 mg/dl), urine copper, and liver function tests. Based on the above findings, a clinical diagnosis of PDS was made, and peripheral Nd:YAG laser peripheral iridotomy (PI) was performed.

On follow-up, a localized clear pigment free endothelial area was noted over the iridotomy sites bilaterally, which further strengthened our diagnosis (Fig. 3). Confocal microscopy revealed fewer hyper-reflective spots over the iridotomy site (Fig. 2, bottom right). The IOP was well controlled between 12 and 14 mmHg at 24 months follow-up on a prostaglandin analogue.

Discussion

Literature abounds with several reports on the types of atypical presentations of PDS like asymmetric involvement, childhood presentation, masquerading as acute anterior uveitis and as uveal melanoma.[3–6] However, diffuse pigment dispersion on the corneal endothelium is not encountered commonly.

PDS is characterized by a structural disturbance in the iris pigment epithelium that leads to dispersion of the iris pigment (melanin granules) and its deposition on various structures within the eye. According to Campbell, the mechanical rubbing leads to loss of pigments in "predisposed individuals".[7] This may be attributed to basic abnormality[7] or congenital defect[8,9] of the pigment epithelium of the iris, and ciliary body,[8,9] or to the reverse pupillary block mechanism in which the concave iris drapes over the lens and acts as a "flap valve," preventing aqueous in the anterior chamber from returning to the posterior chamber.[10] The liberated pigment is deposited on lens zonules, anterior and posterior lens surfaces, iris, cornea, and trabecular meshwork. The deposited pigment contributes to damage to the trabecular meshwork over time, impeding the outflow of aqueous humor. This buildup of aqueous humor may lead to elevated IOP and associated glaucomatous optic

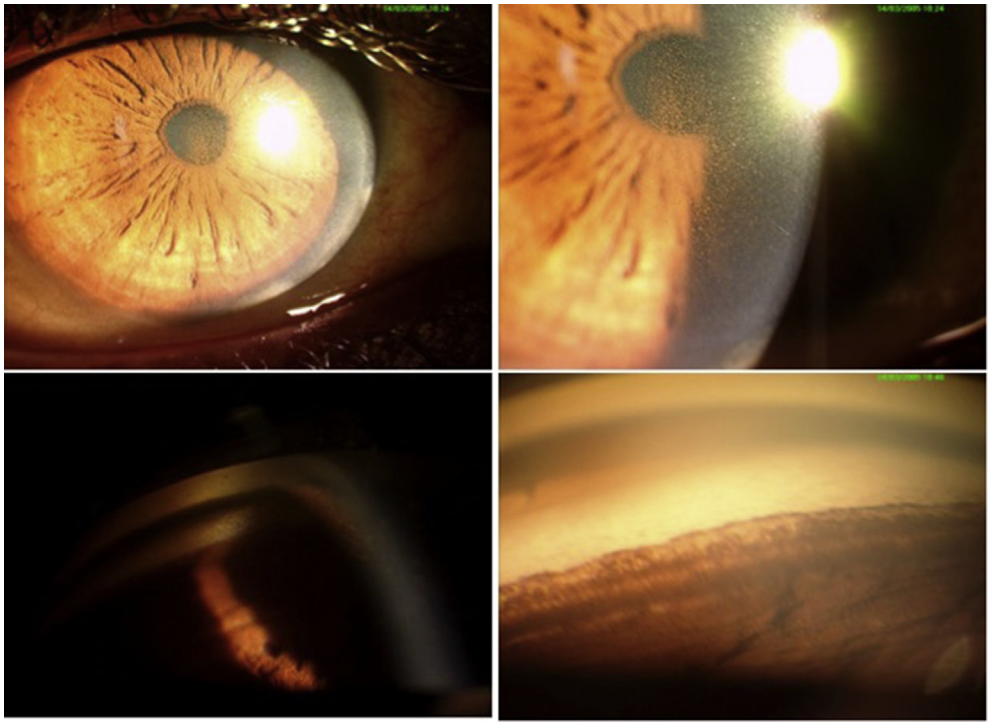

Fig. 1. (Top) Clinical photographs showing bilateral diffuse pigment deposition on corneal endothelium on diffuse and slit illumination; (Bottom) Gonioscopy showing dense, homogeneous pigment deposition in trabecular meshwork (4+) on slit and diffuse illumination with concave iris configuration.

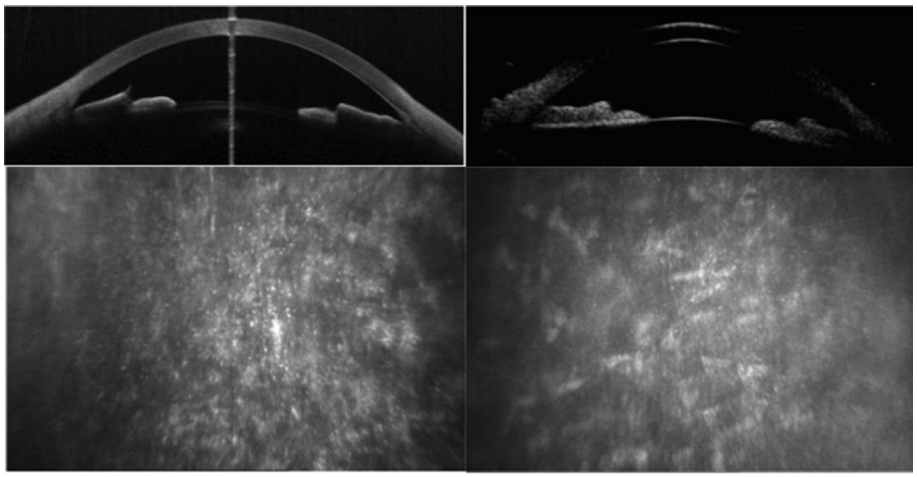

Fig. 2. (Top left) Anterior segment optical coherence tomography (ASOCT), and (Top right) ultrasound biomicroscopy (UBM) showing relatively narrow angles with slight concave iris configuration; (Bottom left) Confocal microscopy of the endothelium reveals hyper-reflective spots suggestive of pigments; (Bottom right) Reduction in pigments noted over the iridotomy site.

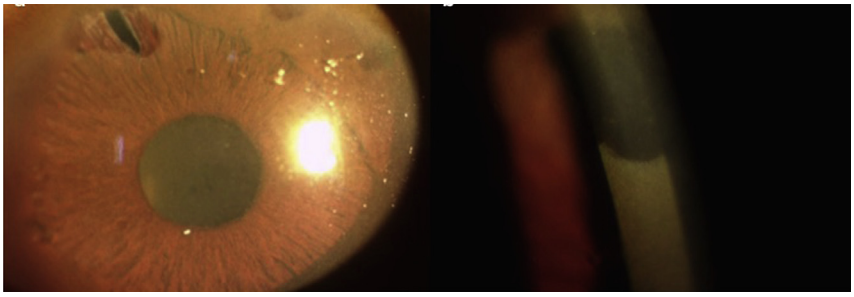

Fig. 3. Clinical photographs showing a localized clear pigment-free endothelial area over the iridotomy sites on diffuse and slit illumination.

neuropathy.[11–14] Our case had a concave iris configuration evident on dynamic gonioscopy as well as UBM that likely led to pigment dispersion. Although the pigment was diffusely dispersed on the endothelium, it was more evident inferiorly.

Peripheral laser iridotomy eliminates the posterior bowing of iris by creating an opening in the iris tissue to allow drainage of fluid from the posterior chamber to the anterior chamber and vice versa. Equalizing the pressure within the eye alleviates the friction that leads to pigment dispersion and prevents visual field deterioration. Exercise and accommodation increase the concavity of iris in patients with PDS.[15] PI abolishes the changes in the iris profile that is normally seen with accommodation in patients with PDS.[15] We therefore performed a PI in our patient.

Our case did not have typical mid-peripheral iris transillumination defects or Krukenberg's spindle. A diffuse pigment deposition led to a misdiagnosis of endothelial dystrophy. But based on a high index of clinical suspicion, the case was thoroughly evaluated to rule out various differentials and to arrive at a diagnosis of PDS.

The presentation of PDS with diffuse pigment deposition on corneal endothelium is rare and can be confused with endothelial dystrophies. This case demonstrates the significance of comprehensive evaluation in cases with unusual presentation.

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