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CASE REPORT

Bilateral Pigment Dispersion Syndrome (PDS) in a Young Female Patient

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Abstract

Introduction: Pigment dispersion syndrome (PDS) is characterized by pigment accumulation in the anterior chamber and a concave peripheral iris. Many PDS patients are not diagnosed until the disease has progressed to pigmentary glaucoma or other visual problems. Since glaucoma is the primary cause of permanent blindness globally, it is crucial to perform a thorough examination on patients with PDS to identify early indicators of pigmentary glaucoma (PG). **Case Presentation:** A 17-year-old female presented to the ophthalmology outpatient unit with eye pain and headache. The intraocular pressure (IOP) in the right eye was 30 mmHg, while in the left eye, it was 20.5 mmHg due to the peripheral iris' concavity and heavy pigmentation in the trabecular mesh in both eyes. The patient was diagnosed with pigment dispersion syndrome in both eyes and was given timolol maleate 0.5% eye drops. A follow-up examination revealed a decrease in the IOP and pain. **Conclusions:** Many young PDS patients go undiagnosed, and those with glaucoma are misdiagnosed as having juvenile onset glaucoma or primary open angle glaucoma. When high IOP is seen in young myopic patients, a thorough evaluation of the anterior segment is required. The patient should be aware of the progression of PG, and regular follow-up is recommended.

Keywords: pigment dispersion syndrome (PDS); pigmentary glaucoma (PG); trabecular pigmentation; myopia

Introduction

Pigment dispersion syndrome (PDS) is a secondary open-angle glaucoma defined by aberrant dispersion of the iris pigment epithelium and the accumulation of the iris pigment on anterior segment structures.[1],[2] The most frequent cause of PDS is pigment leakage from friction between zonules and the posterior iris brought on by a peripheral iris concavity.[3] The pigment subsequently accumulates in the trabecular meshwork, obstructing the aqueous humor outflow and causing cell atrophy.[4] The obstruction can lead to elevated intraocular pressure (IOP) and progress to pigmentary glaucoma (PG).

PDS is uncommon in African and Asian ethnicities and affects about 2.5% of the European Caucasian population.[5] Myopia, Caucasian race, male gender, and family history are among the risk factors for PDS.[1] According to Simcoe et al (2022) estimated 15% of PDS patients will proceed to PG within 15 years of their initial diagnosis, and up to 50% will do so during their lifespan.[2]

Many PDS patients are not detected until a late stage of the condition when PG or visual symptoms occur.[6] Therefore, it is important to do a comprehensive examination in patients with PDS to detect early signs of PG since glaucoma is the leading cause of irreversible blindness worldwide.[7] This case report will present a case of pigment dispersion syndrome in a 17-year-old female patient.

Case presentation

A 17-year-old female presented to the ophthalmology outpatient ward with complaints of eye pain and headache. The patient previously came to the emergency ward with diagnosed ocular hypertension in both eyes. The IOP in the right eye was 30 mmHg and in the left eye was 20.5 mmHg. She was treated with

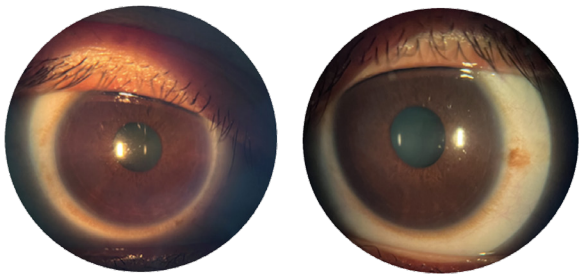


Figure 1. Anterior segment examination in both eyes is within normal limits.

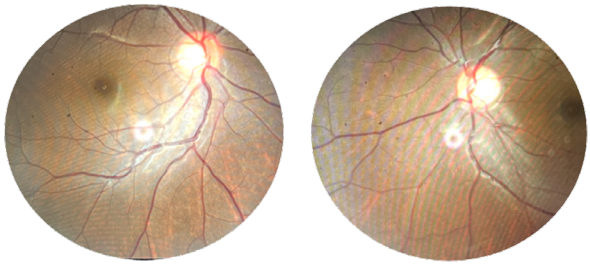


Figure 2. Posterior segment in both eyes. (A) The right eye showed a CDR of 0.6 and a tilted disc, and (B) The left eye showed a CDR of 0.5 with no glaucomatous optic neuropathy.

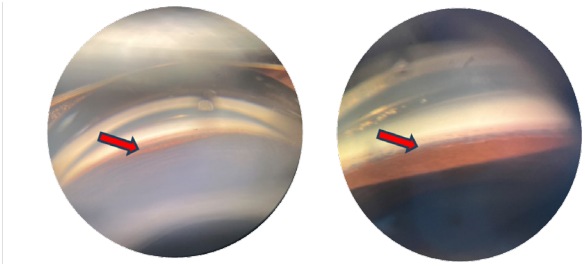


Figure 3. A gonioscopy examination in both eyes revealed heavy pigmentation in the trabecular meshwork (red arrow).

timolol maleate 0.5% eye drop every 12 hours in the right eye, carboxymethyl cellulose sodium eye drop every four hours in both eyes, acetazolamide 250 mg twice a day per oral, potassium chloride 600 mg (8 mEq K) once a day per oral, paracetamol 500 mg three times a day per oral and scheduled to follow up.

At the follow-up visit, she reported that the ocular pain and headache had been reduced. There is no blurry vision in both eyes and no nausea or vomiting. The pain usually occurs when exposed to light and improves with oral analgetic. There is no complaint of pain after exercise. There is no history of glaucoma or ocular hypertension in the family. There is no history of ocular surgery and trauma. She had a history of using glasses since the 5th grade of elementary school (S-7.50 in both eyes).

The best-corrected visual acuity (BCVA) in the right eye was 6/6 with correction S-7.25 C-1.00 A180. The BCVA in the left eye was 6/6 with correction S-6.75 C-0.75 A180. The IOP in the right eye was 18 mmHg and in the left eye was 17 mmHg. Anterior segment examination by slit lamp biomicroscopy in both eyes was within normal limits. The anterior chambers were profound, with Van Herick grade 4 in the right eye and Van Herick grade 3 in the left eye. There was no endothelial pigment in the cornea and no mid-peripheral iris transillumination defect (Figure 1).

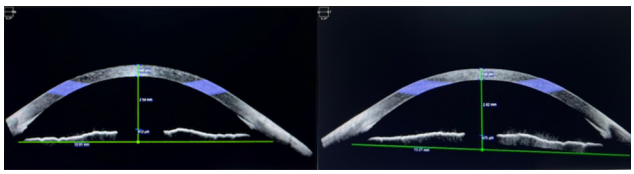


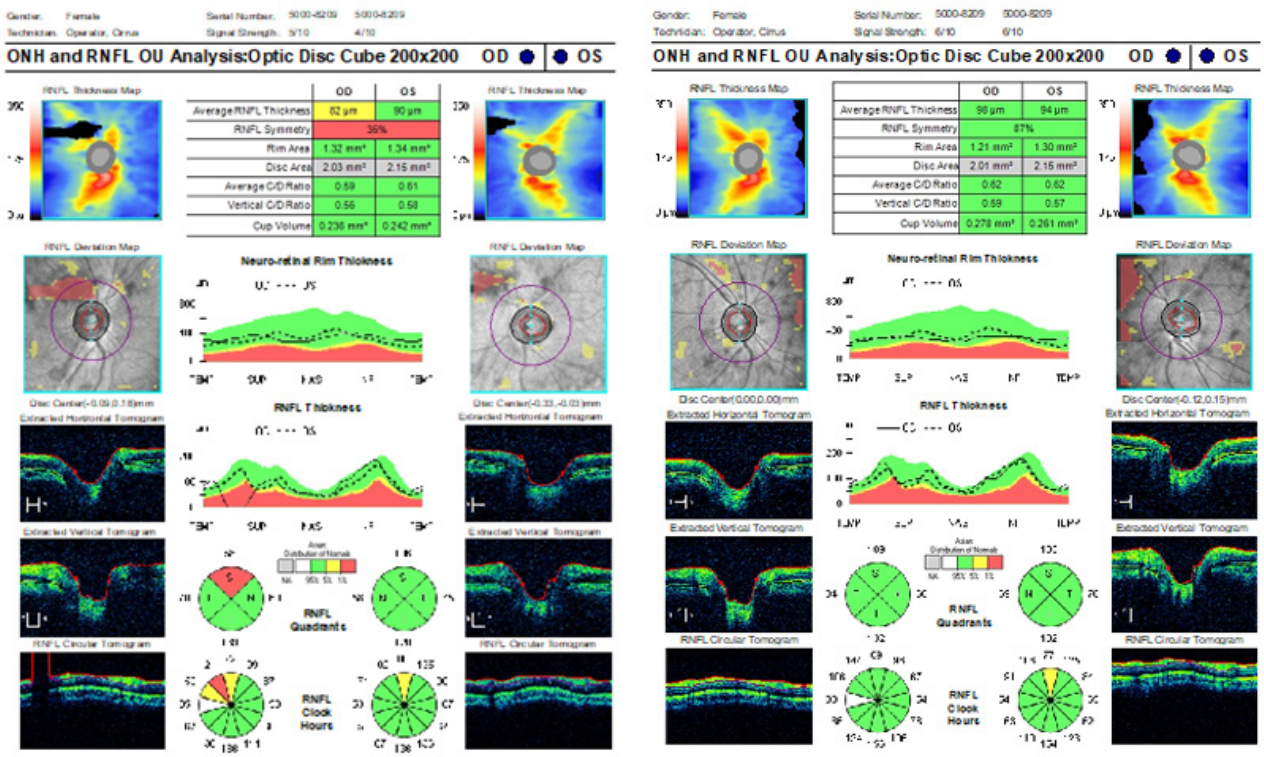
Figure 4. Anterior OCT captures concavity peripheral iris in (A) the right eye and (B) the left eye.

The funduscopic examination shows a normal fundus reflex in both eyes. The optic disc showed a cup-to-disc ratio (CDR) of 0.6 with a tilted disc in the right eye and a CDR of 0.5 in the left eye without signs of glaucomatous optic neuropathy in both eyes. The foveal reflex was good and the peripheral retina was healthy (Figure 2). We performed a gonioscopy and showed bilateral open angles with a heavy diffuse grade 4+ pigmentation in trabecular meshwork using Scheie's grading (Figure 3).

The retinal nerve fiber layer (RNFL) in the left eye (Figure 5A). Humphrey field analyzer (HFA) examination showed a general reduction of sensitivity in both eyes (Figure 6). The patient was diagnosed with pigment dispersion syndrome in both eyes and compound myopic astigmatism in both eyes. Due to increased IOP in the right eye accompanied by pain, we decided to continue antiglaucoma medication with timolol maleate 0.5% eye drop every 12 hours in the right eye, carboxymethyl cellulose sodium eye drop every four hours in both eyes and stop the oral acetazolamide and potassium chloride.

After a two-week follow-up, the pain and headache were getting better. The BCVA in both eyes was 6/6. The IOP in the right eye was 18 mmHg on therapy, and in the left eye was 17 mmHg. The examination was within normal limits from the anterior and posterior segments. We planned to continue the medication with timolol maleate 0.5% eye drop in the right eye every 12 hours and carboxymethyl cellulose sodium eye drop every six hours in both eyes. After a four-week follow-up examination, the pain was relieved. The BCVA in both eyes remained the same. The IOP in the right eye was 19 mmHg on therapy, and in the left eye was 17 mmHg. Anterior segment and posterior segment examination were within normal limits.

The medication continued with a timolol maleate 0.5% eye drop in the right eye every 12 hours and carboxymethyl cellulose sodium eye drop every six hours in both eyes. The patient was educated about her disease and the risk of pigmentary glaucoma. The patient was scheduled for a 6-month glaucoma progression evaluation with optical coherence tomography (OCT) and HFA. The OCT evaluation revealed normal RNFL in both eyes (Figure 5B). HFA evaluation showed no visual defect in both eyes (Figure 7).



A B

Figure 5. (A) The optic disc OCT showed thinning RNFL in the superior area in the right eye and normal RNFL in the left eye, and (B) The optic disc OCT evaluation showed normal RNFL in both eyes.

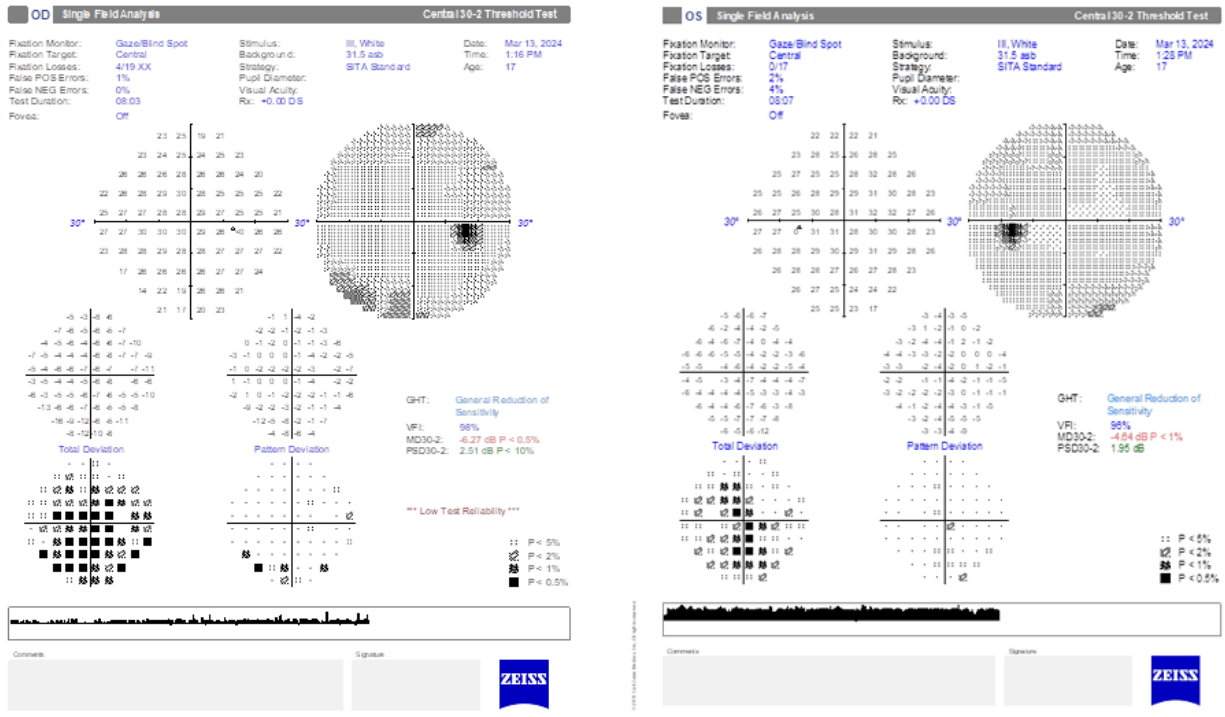


Figure 6. Humphrey Field Analyzer (HFA) examination showed a general sensitivity reduction in both eyes.

Discussion and conclusions

PDS is characterized by pigment accumulation in the anterior chamber and a concave peripheral iris. In addition, pigments on the corneal endothelium (Krukenberg spindle), increased trabecular meshwork pigmentation, and mid-periphery iris transillumination abnormalities are characteristics of this condition.[5] Males and myopic patients are more likely to have PDS, which is heritable in an autosomal dominant manner. In this case, our patient presented a 17-year-old female with characteristics of a concave peripheral iris and an increased pigmentation of the trabecular meshwork in both eyes. This patient has no Krukenberg spindle and mid-peripheral transillumination defects of the iris. Krukenberg spindles

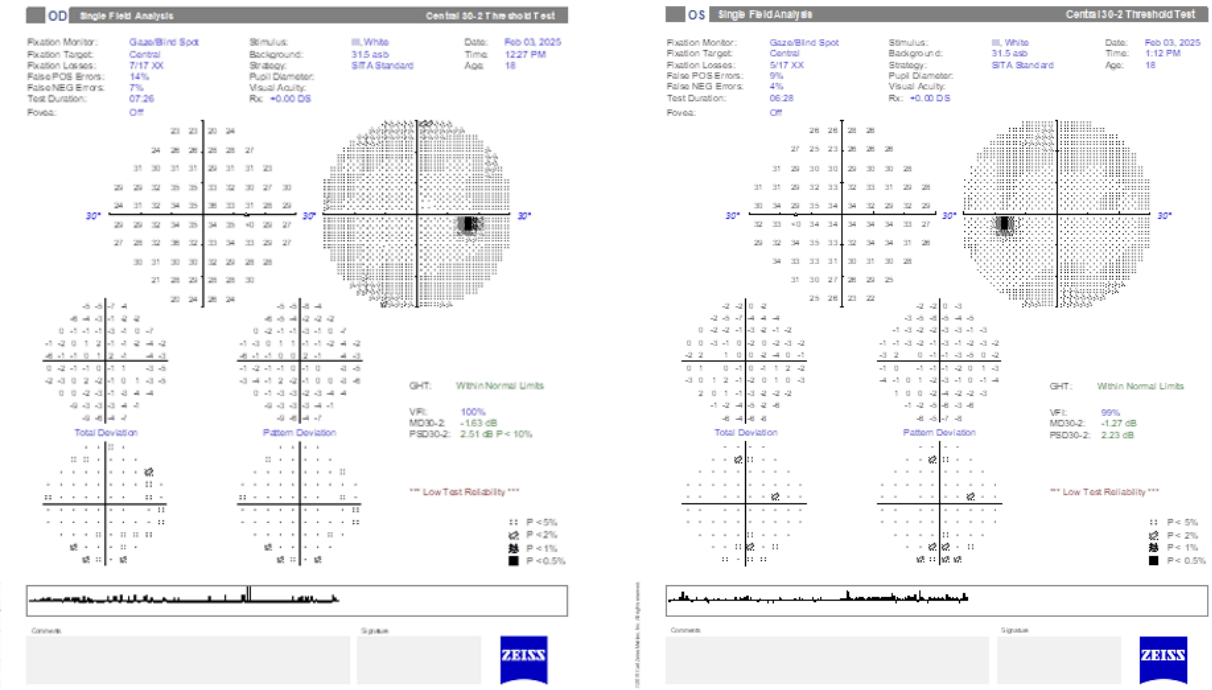


Figure 7. Humphrey Field Analyzer (HFA) evaluation showed no visual defect in both eyes.

and mid-peripheral iris transillumination abnormalities are typical, but not always present, features of this clinical entity.[8] According to a study by Siddiqui et al. [9], intensely pigmented trabecular meshwork was the only clinical symptom seen in 18% of patients with PDS. According to a study by Qing et al.[6], pigment granule dusting on the peripheral posterior lens surface and/or zonules and homogenous moderate-to-heavy trabecular meshwork pigmentation are the most prevalent clinical findings in Chinese PDS patients. Due to their dark irides, this patient population does not often display typical spoke-like mid-peripheral iris transillumination defects (ITDs). In contrast to light iris stroma, black iris stroma has more pigment granules in the melanocytes and iris stroma. They can, therefore, obstruct an iris pigment epithelium (IPE) transillumination flaw. Despite being visible in 61.1% of Chinese PDS patients, the highly pigmented iris's dark background makes the Krukenberg spindle challenging to detect in some of them, even with slit-lamp biomicroscopy. This can sometimes result in an incorrect diagnosis or exclusion.[6]

PDS primarily affects myopic adults in their middle years; cases in children under 14 are highly uncommon.[8] The extended iridozonular contact between the anterior zonular packets and the concave peripheral iris may be the mechanism causing the pigment dispersion in PDS.[3] It typically affects myopic patients because of the anterior chamber's expanding volume and ensuing higher levels of iridolenticular contact. Deep anterior chambers and moderate myopia are conducive to these mechanisms. It has been suggested that in a deep anterior chamber, typically in myopia patients with elongated eyes, eyes with PDS may have a big iris that may bow backward and touch lens structure.[10] According to a study by Rafiq & Lokovitis[11], other iris-specific characteristics must exist to cause pigment dispersion, and iris concavity and the related iridocorneal contact must endure for a considerable time before PDS develops. These characteristics could be the primary causes of the early PDS manifestation.[11] When PG or visual symptoms appear in a patient with PDS, it is often too late to diagnose them. In the study[6], PG was present in 83.3% of patients, while elevated IOP was seen in 94.4% at PDS diagnosis.

Treatment of each patient with PDS should be customized to that individual. Not all PDS patients will develop increased IOP. The goal of the treatment is to prevent the development of PG. In this case, the patient received antiglaucoma eye drops and gave good IOP results. Treatment of PDS should begin early to reduce the high IOP and prevent the development of glaucomatous damage. The treatment can start with medical therapy before considering surgical therapy. The selection of the individual medication will depend on the stage of the disease and the presumed extent of trabecular damage. Some medications enhance uveoscleral and trabecular outflow or reduce aqueous production.[12] Pilocarpine produces a convex iris configuration, completely inhibiting pigment liberation. Laser iridotomy results in a planar configuration of the iris but may not completely prevent pigment liberation. The preventative role of laser peripheral iridotomy (LPI) is considered controversial. Laser trabeculoplasty is controversial and has not been studied as extensively as in primary open-angle glaucoma and exfoliation glaucoma. Argon laser trabeculoplasty (ALT) produces better results in younger patients because

of the location of pigment in the trabecular meshwork.[13] Compared to ALT, selective laser trabeculoplasty (SLT) employs less energy over a shorter duration. It is thought to selectively target pigmented trabecular meshwork cells, gently stimulating their metabolic state and improving aqueous outflow without collateral thermal damage.[5],[14]

The limitations of this study are the difficulty of evaluating the posterior segment in myopic patients and the short follow-up period. Further follow-up is needed to evaluate the therapy results and detect the early signs of glaucoma.

Many young patients with PDS remain undetected until the symptoms appear, while those with glaucoma are often misdiagnosed with juvenile-onset glaucoma or primary open-angle glaucoma. In the case of high IOP in young myopic patients, it is important to do a comprehensive examination in the anterior segment to find a pigment liberation in the angle and the peripheral concave iris. Especially if the patient has no Krukenberg spindle and no transillumination defects. In myopic patients, it is quite challenging to evaluate the glaucomatous progression of the optic disc. The patient should be aware of the development of PG, and they must be educated about the importance of regular eye checkups and the possible avoidance of heavy exercise, which may cause the liberation of pigment and progression of glaucoma.

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