

PLATEAU IRIS SYNDROME – CASE SERIES

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Abstract

Plateau iris is characterized by closing the anterior chamber angle due to a large ciliary body or due to its anterior insertion that alters the position of iris periphery in respect to the trabecular meshwork. There are two aspects that need to be differentiated: plateau iris configuration and plateau iris syndrome. The first describes a situation when the iris root is flat and the anterior chamber is not shallow, the latter refers to a post laser iridotomy condition in which a patent iridotomy has removed the relative pupillary block, but gonioscopically confirmed angle closure recurs without central shallowing of the anterior chamber. Isolated plateau iris syndrome is rare compared to plateau iris configuration. We hereby present two case reports of plateau iris syndrome in young patients who came to an ophthalmologic consult by chance.

Key words: plateau iris configuration, plateau iris syndrome, glaucoma

Introduction

Plateau iris represents one of the most frequent causes of primary angle closure in young patients. It is diagnosed frequently under the age of 50, in patients with narrow anterior chamber angle, after a correct and patent laser peripheral iridotomy was made. There are two aspects that need to be differentiated: plateau iris configuration and plateau iris syndrome [1]. Iris plateau configuration exhibits a flat iris plane with an anterior chamber that is not shallow axially. Plateau iris syndrome refers to a post laser iridotomy condition in which a patent iridotomy has removed the relative pupillary block, but gonioscopically confirmed angle closure recurs without central shallowing of the anterior chamber. Isolated plateau iris syndrome is rare

compared to plateau iris configuration. It usually occurs in a younger age group than pupillary block angle closure [2]. The treatment is laser iridoplasty or long term use of pilocarpine postoperatively. This condition must be considered in the differential diagnosis when the intraocular pressure rises unexpectedly following an adequate peripheral iridotomy/iridectomy procedure for angle closure glaucoma. [3,4]

Case 1

We present the case of a late diagnosed closed angle glaucoma in a young patient who came to an ophthalmologic consult by chance. Mother (43 years old) brought her daughter to our department in April 2015, where the child was diagnosed with retrobulbar optical neuritis.

At the child's regular visit at one month after the acute episode of optic neuritis, mother requests a consult for herself as vision became blurry for some time in OD.

Personal history: AO – moderate hyperopia
 AOVD= 0.5 cc (sph +3.75)
 AVOS= 1 cc (sph +4.5)

Visual field (Humphrey Visual Field Analyzer II -120°, 3 zones)

OD – visual field constriction up to the central 3° area

OS – normal visual field (VF)

HFA II, 30-2 central, Full Threshold Strategy, size III stimulus showed in OD reduced retinal sensitivity (MD = -30.88 dB) and central/paracentral scotoma (PSD=6.88 dB), see (Fig. 1-2).

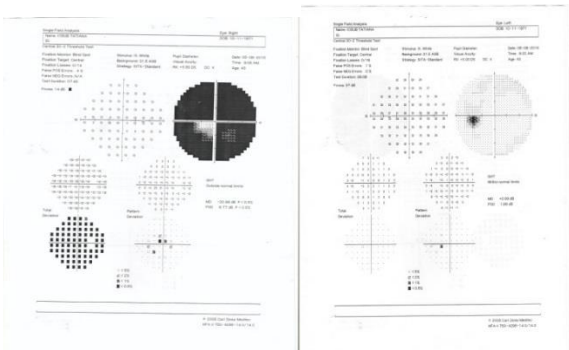
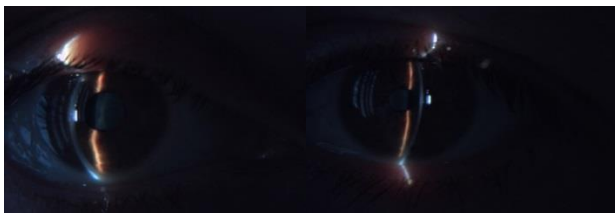


Fig. 1-2 HFA II central 30-2 visual field

Anterior segment: AO shallow anterior chamber in periphery, but normal depth in the center see (Fig. 3-4)



OD – anterior chamber OD – anterior chamber

Fig. 3-4 anterior chamber shallow in periphery, normal depth centrally

IOP-OD=70 mmHg
 IOP-OS=36 mmHg

When fundus exam was performed, it revealed almost total excavation in right eye (C/D=1), whereas C/D ratio in OS was 0.5/0.4.



FOOD – C/D=1(v)/ 0.9(h) FOOS – C/D=0.5/0.4

Fig. 5 FOAO- C/D ratio

Gonioscopy showed in AO that iris assumed a steep approach at its insertion before flattening centrally (iris drapes over the ciliary body producing the double “hump” aspect on indentation – see Fig. 6).

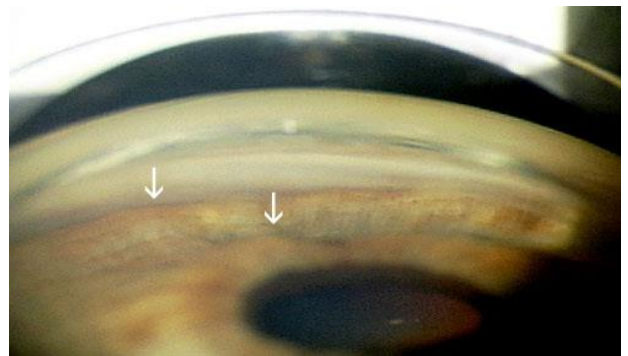


Fig. 6 gonioscopy in plateau iris “double hump” sign (indentation)

We diagnosed the patient with plateau iris syndrome; thus deciding to systemically prescribe manitol 20% (iv) and acetazolamide plus Pilocarpine 2% topically.

When anamnesis was rediscussed, the patient revealed multiple ophthalmology visits where she complained of headaches and blurred vision that were always interpreted as poor optic correction, but no fundus examination was ever thoroughly done. When the IOP decreased to a satisfactory level, a peripheral laser iridotomy (IP) was done along with an ALPI procedure

(argon laser peripheral iridoplasty) (see **Fig. 7-8**)

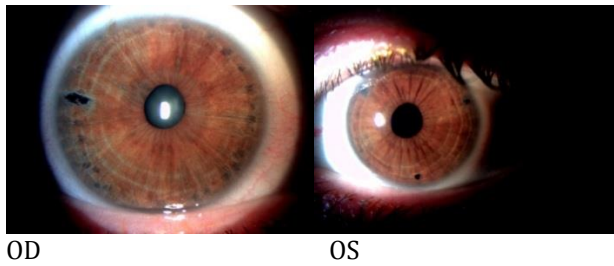


Fig. 7-8 Status post IP+ALPI

IOP after laser procedures was 21 mmHg in OD, thus beta blocker and carbonic anhydrase inhibitor was prescribed to lower supplementary the IOP; in OS the IOP was 15 mmHg without any adjuvant medication. Pilocarpine was prescribed in both eyes.

At 3 months check-up Pilocarpine 2% was withdrawn, and the patient remained under supervision for IOP and gonioscopy each 3 months; last control showed open angle/ 360° (Schaffer II or III) and IOP in both eyes was 15 mmHg.

Case 2

B.E, female, 29 years old was referred to our Ophthalmology Clinic for a diagnosis of OU Juvenile decompensated glaucoma. Major complaints were ocular pain and bilateral decreased vision, right eye more than the left eye.

Last ophthalmological visit stated: AVOD = 0.1 sc; AVOS = 0.7sc, IOP-OD=38 mmHg, IOP-OS=40 mmHg; fundus examination: OU - retinal degenerescence, C/D ratio OD=0.5, OS=0.6. Treatment was first initiated with topical beta blockers, then due to the lack of response, prostaglandin analogues (PGA) and carbonic anhydrase inhibitor (CAI) were added topically; Brimonidine was needed topically only in OS. Under this treatment IOP was OD=19 mmHg, OS=28mmHg.

A presumed diagnosis of autosomal recessive bestrophinopathy (ARB) is suspected

by the retina specialist. An electrophysiology testing is mandatory for diagnosis confirmation.

Our current examination found AVOD=0.2 sc; AVOS=0.7 cc (sph +2)

IOP-OD=10 mmHg (PGA+BB+CAI), IOP-OS=19 mmHg (PGA+BB+CAI+Brimonidine)

Corneal central thickness: OD = 469µm, OS=474 µm

Slit lamp examination shows shallow anterior chamber in periphery, but normal axial depth (see **Fig. 9-10**).

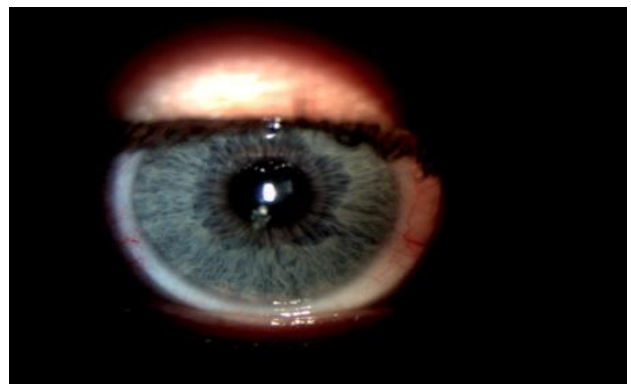


Fig. 9 anterior segment OD



Fig. 10 Shallow peripheral AC (OD)

Gonioscopy reveals convex iris, closed angle/ 270° and open inferiorly (Schaffer I); by indentation the angle opened Schaffer IV/ 360°, "double hump" sign positive (see **Fig. 11-12**).



Fig. 11 closed angle



Fig. 12 "double hump" sign

Visual field (HFA II - central 24-2 SITA Standard) found in OD decreased general retinal sensitivity and paracentral inferior/ central with fixation spot involvement scotoma, MD=-3.69 dB, PSD=5.15 db. In OS there was also a decreased general retinal sensitivity and a superior incomplete arcuate scotoma, with MD=-5.65 dB, PSD=6.26 dB (see **Fig. 13-14**).

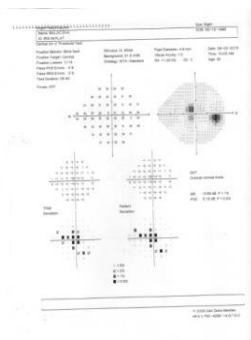


Fig. 13 VF - HFA II c24-2 (OD)

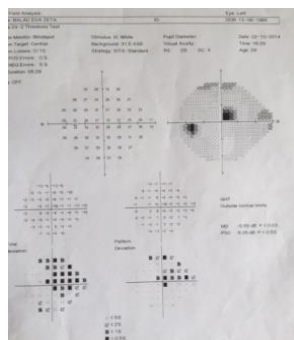


Fig. 14 VF - HFA II c24-2 (OS)

Fundus examination showed asymmetric excavation C/D OD= 0.2, C/D OS=0.5 in small optic disks and in both eyes, diffuse irregularity of retinal pigment epithelium, dispersed punctate flecks and chronic accumulation of fluid beneath the neurosensory retina in the macular region (**Fig. 17-18**).



Fig. 15 FOOD presumed autosomal recessive bestrophinopathy

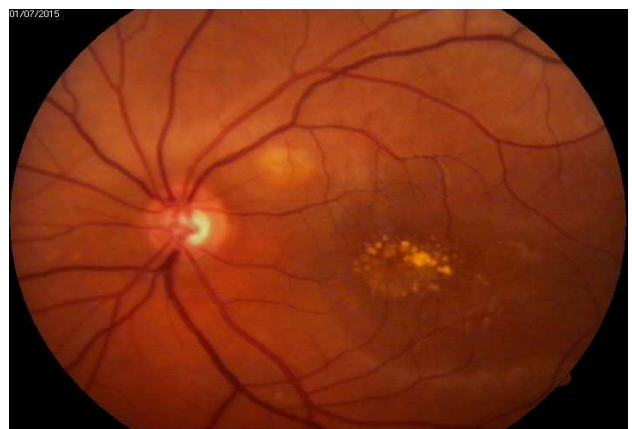


Fig. 16 FOOS presumed autosomal recessive bestrophinopathy

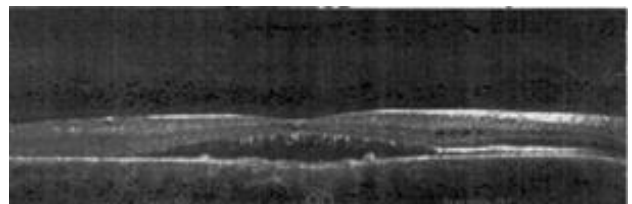


Fig. 17 OD- SD OCT

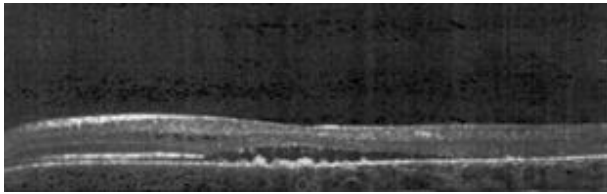


Fig. 18 OS- SD OCT

This patient had initially laser peripheral iridotomies. Her angle depth improved in each eye but there was concern that the angle in the left eye was still occludable. She underwent a bilateral ALPI procedure without complication and repeated gonioscopy that proved improvement in the angle depth OU (see Fig. 19-20).



Fig. 19 OD-post laser treatment

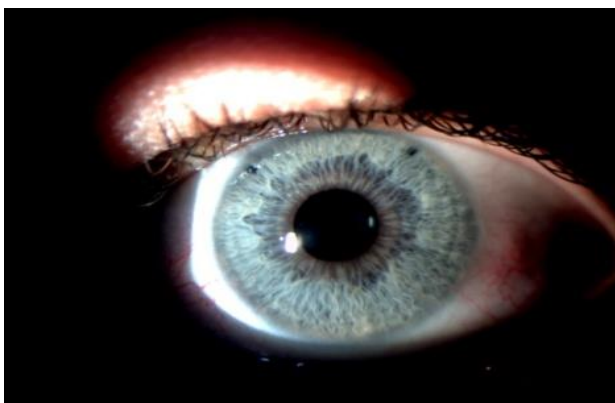


Fig. 20 OS post laser treatment

At the last visit, the angle does not appear to be occludable and we are monitoring her gonioscopically every 3 months.

IOP-OD=16 mmHg; IOP-OS=16 mmHg (BB+CAI)

Discussion

Plateau iris syndrome is a relatively uncommon form of primary angle closure glaucoma that is seen more often in younger adults than pupillary block angle-closure glaucoma⁵. Patients can come at the hospital presenting angle closure, either spontaneously or after pupillary dilation [6] as one of the cases presented here (case 2), but more commonly, patients are asymptomatic and the diagnosis is made on routine examination (case 1).

Surgical management is the primary treatment modality in patients with plateau iris configuration or syndrome. A patent iridotomy may be therapeutic in reducing risks of angle closure. However, in some patients, laser iridotomy may not significantly alter the anterior chamber depth or anatomy. Even after a successful iridotomy resulting in a well-open angle, periodic gonioscopy remains crucial because these patients may have incomplete plateau iris syndrome or the angle may narrow further with age because of the enlargement of the lens.

Argon laser peripheral iridoplasty (ALPI) is the procedure of choice to effectively open an angle that remains occluded after successful laser iridotomy. It is highly effective, and the effect is maintained for years [7]. Yet, even after successful opening of the angle, regular gonioscopy is mandatory.

Our cases exhibit certain particularities: diagnosis made by chance, advanced glaucoma that passed undiagnosed until advanced stage, thorough history and gonioscopy helped getting a correct diagnosis and eliminate other causes of increased IOP in young patients (congenital glaucoma, pigmentary/ inflammatory/ post-traumatic glaucoma, so on); C/D ratio not proportionally increased with the large IOP fluctuations show the intermittent angle closure history; macular associated pathology contributed to additional visual decay.

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