American Journal of Ophthalmology Case Reports 10 (2018) 257–260

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.elsevier.com/locate/ajoc

Brief Report

Bilateral acute depigmentation of the iris in two siblings simultaneously

Rana Amin[a,b], Amena Nabih[b,*], Noha Khater[a,b]

[a] Department of Ophthalmology, Faculty of Medicine, Kasr El Aini Hospital, Cairo University, 1 Kasr El Aini Street, Al Manial, Cairo, Egypt
[b] Al Mouneer Center for Diabetic Eye Care, 92 El Tharir Street, 4th Floor, Giza, Egypt

ARTICLE INFO

Keywords:
Acute
Depigmentation
Pigment dispersion
Symmetrical
Krukenberg
Gonioscopy

ABSTRACT

Purpose: To report the first simultaneous onset of bilateral acute depigmentation of the iris (BADI) in two siblings.

Observations: Two sisters presented with bilateral ocular pain, redness and light sensitivity. Examination revealed bilateral circulating pigment in the anterior chamber with pigment dusting on backs of the corneas, patchy iris depigmentation and heavy pigment deposition in the angle. Both patients had recently suffered from upper respiratory tract infections. Bilateral visual acuities were preserved and no transillumination defects were observed. The patients were diagnosed with BADI. Both cases were successfully controlled with topical corticosteroids and anti-glaucoma drops as well as topical glanciclovir gel.

Conclusions and Importance: To date, there had been no published reports of BADI in the Middle East and Africa. This is the first observation of this entity in these regions. Moreover it is the first occurrence of BADI in two immediate siblings simultaneously. We also report the rare asymmetrical presentation with BADI in one of our patients. These observations point to the possibility of genetic factors underlying BADI as well as an infectious cause behind the etiology.

1. Introduction

Bilateral acute depigmentation of the iris (BADI) is a newly described entity characterized by acute onset of depigmentation of the iris stroma.[1] Pigment often disperses into the anterior chamber and is deposited heavily in the anterior chamber angle. Clogging of the trabecular meshwork often results in increased intraocular pressure. Patients present with red eyes, photophobia, ocular discomfort and/or pain. BADI involvement is usually bilateral, symmetrical and most commonly occurs in young females in their third and fourth decades.[1–3] Contrary to the closely related syndrome of bilateral acute iris transillumination (BAIT), BADI patients do not present with symptoms of transillumination and pupillary deformities.[1,4] Cases of BADI and BAIT often occur after a flulike illness or upper respiratory tract infection. Moreover, several cases of BAIT have been associated with recent ingestion of oral moxifloxacin.[5,6] Previous cases of BADI have been well controlled with topical corticosteroids, although in many cases corticosteroids lead to an increase in IOP.[2]

Since its initial characterization by Tukal-Tutkun et al., in 2005, fewer than 100 cases of both BADI and BAIT have been described worldwide: 51 in Turkey, 4 in the Netherlands, 4 in Belgium, 4 in Colombia, one in Spain, one in France and one in Brazil.[1–3,7–9] To our knowledge, there are no previous reports of BADI cases in the Middle East and Africa. Here, we present the first two cases of BADI in the region.

2. Case 1

A 28 year old female presented on October 1st, 2017 with acute severe bilateral ocular pain, light sensitivity and redness. Her best-corrected visual acuity (BCVA) was 1.0 in both eyes. Slit lamp examination revealed bilateral +2 ciliary injection, pigment clumps on the back of the corneas (Krukenberg spindles), +2 circulating pigment in the anterior chamber and symmetrical patchy depigmentation that was mostly close to the iris root, resulting in a more exaggerated appearance of the iris furrows. No transillumination defects were present (Fig. 1a–d). Pupils were rounded, regular and reactive. The lens was clear and posterior segment examination was unremarkable with no vitreous cells. Her intraocular pressures (IOPs) were 16 mmHg and 18 mmHg in the right and left eye, respectively. Gonioscopy revealed open angles with heavy pigmentation of the trabecular meshwork, especially in the inferior quadrant (Fig. 1e and f).

The review of systems was unremarkable apart from an upper respiratory tract infection one month prior to the onset of symptoms. It remains unknown whether the patient was treated with moxifloxacin.

The patient was immediately started on topical corticosteroids and

* Corresponding author.
E-mail address: anabih@almouneer.com (A. Nabih).

https://doi.org/10.1016/j.ajoc.2018.03.016
Received 18 January 2018; Received in revised form 28 February 2018; Accepted 15 March 2018
Available online 16 March 2018

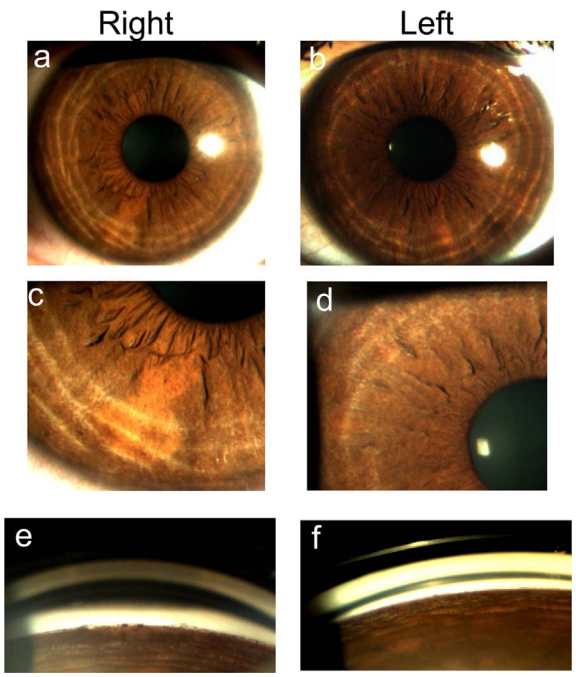

Fig. 1. Case 1. Slit-lamp photographs of Case 1, 28 year old female presenting with bilateral acute symmetrical iris depigmentation. Pictures were taken 7 weeks after initial presentation. 1a and 1b show right and left patchy peripheral geographic stromal atrophy respectively. 1c shows area of normal iris pigmentation surrounded on either side by areas of lost pigmentation in the patient's right eye. 1d shows peripheral patchy loss of pigmentation in left eye. 1e and 1f show gonioscopy of patient's heavily pigmented inferior angles due to pigment deposition.

cyclopentolate drops, which resulted in marked relief of symptoms and a decrease in circulating pigment in the anterior chamber to +1. Ten days after initial presentation, there was an increase in IOPs to 20 mmHg and 23 mmHg in the right and left eye, respectively. Any attempt to taper topical steroids early prior to the full resolution of pigment dispersion resulted in the recurrence of the initial symptoms. IOP remained persistently elevated at 23 mmHg. Topical anti-glaucoma was then prescribed to lower IOP.

Serological tests for herpetic viruses were all negative with the exception of highly positive CMV IgG count indicating a past CMV infection.

3. Case 2

A 25 year old female (the sister of Case 1) presented on October 4th, 2017 with parallel symptoms of redness, pain and photophobia. Symptoms were more severe in the right eye. Her BCVA was 1.0 in both eyes. Slit lamp examination revealed bilateral +2 ciliary injection in the right eye and +1 in the left eye, pigment clumps on back of both corneas (Krukenberg spindles) that were more pronounced in the right eye, +3 circulating pigment in the right anterior chamber and +1 in the left eye, and symmetrical patchy depigmentation of the iris, mainly close to iris root. No transillumination defects were present (Fig. 2a–d). Pupils were rounded, regular and reactive. The lens was clear and posterior segment examination was unremarkable with no vitreous cells. Her IOPs were 16 mmHg and 22 mmHg in the right and left eyes, respectively. Gonioscopy revealed open angles with heavy pigmentation of the trabecular meshwork especially in the inferior quadrant, in addition to the presence of a fibrotic band on the back of the cornea in the inferotemporal quadrant of the right eye and nasal quadrant of the left eye. (Fig. 2 e,f).

Patient history included recurrent sinusitis with a recent exacerbation after an upper respiratory tract infection, which was treated with oral moxifloxacin in June.

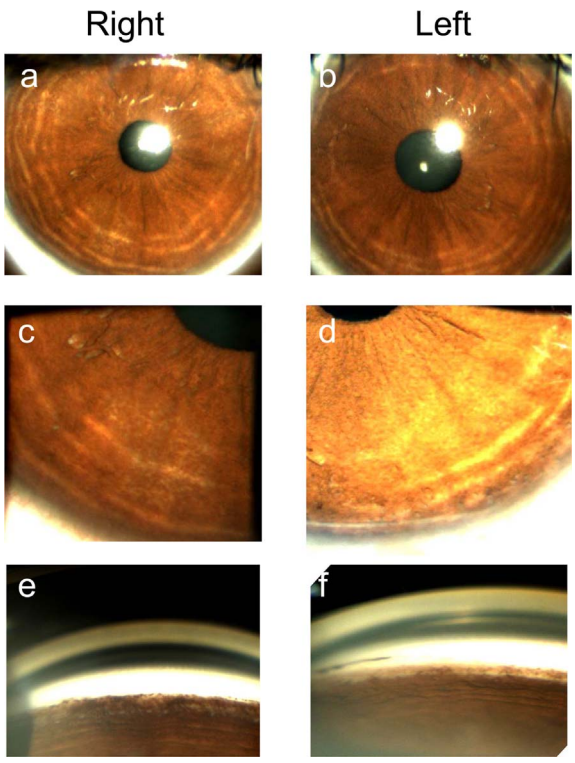

Fig. 2. Case 2. Slit-lamp photographs of Case 2, 25 year old female presenting with bilateral acute asymmetrical iris depigmentation. Pictures were taken 7 weeks after initial presentation. 2a and b show right and left patchy bands on the back of inferotemporal cornea in right eye and nasal cornea of left eye. Fig. 1c and d shows areas of normal iris pigmentation with peripheral areas of lost pigmentation in the patient's right and left eyes respectively. Fig. 1e shows gonioscopy of patient's heavily pigmented inferior angle of right eye due to pigment deposition. Fig. 1f shows nasal quadrant of left eye with pigmented line on back of peripheral cornea.

The patient was immediately started on topical corticosteroids and cyclopentolate drops, which alleviated pigment dispersion. Ten days later, the IOPs measured were 22 mmHg and 28 mmHg. Any attempt to quickly taper topical steroids to control IOP resulted in a flare with an increase in circulating pigment in anterior chamber. Topical anti-glaucoma was then prescribed to lower IOP.

Similar to Case 1, serology results were negative for herpetic viruses with the exception of highly positive CMV IgG counts. Seven days later, serology for CMV IgG count was repeated for both cases to find out if there is a rising titer for the antibodies. The results came back with the same numbers for both cases.

Both cases were prescribed the same treatment of cortical steroids and cyclopentolate drops. During the gradual tapering of topical steroids from six times daily, the amount of pigment dispersion decreased yet stabilized at +0.5 - +1.0 with no further improvement in both patients. At the stage of three times daily topical prednisolone acetate, topical ganciclovir 0.15% ophthalmic gel (trade name Ganvir, Orchidia Pharmaceuticals, Egypt) was prescribed five times daily. One week after the initial use of topical glanciclovir there was complete resolution of the pigment in the anterior chamber in both cases despite continuous topical steroid tapering. IOP returned to normal (Case 1 OD: 20, OS:18, Case 2: OD 17, OS:17) two weeks after anti-glaucoma treatments were halted.

During the course of the disease, the anterior chamber reaction and pattern of iris depigmentation were strikingly similar in the two siblings. Both patients had patchy stromal iris depigmentation, gradual rise in IOP (despite decreasing steroid drops frequency), minimal flare, and absolutely no signs of inflammatory keratic precipitates or transillumination iris defects. The only dissimilarity between the two cases was the symmetrical presentation in Case 1 and asymmetrical