A Case of Bilateral Acute Depigmentation of the Iris in One of Two Identical Twins

Spencer Langevin
Columbia University Medical Center https://orcid.org/0000-0002-1492-8016
Alexandra Gershkovich
Columbia University Medical Center
Brian P Marr (bpm2133@cumc.columbia.edu)
Columbia University Medical Center

Research Article

Keywords: Iris; depigmentation; Sarapin, iris transillumination

Posted Date: February 18th, 2019

DOI: https://doi.org/10.21203/rs.2.342/v1

Version of Record: A version of this preprint was published on January 6th, 2020. See the published version at https://doi.org/10.1186/s12886-019-1282-z.

Abstract

Background: Bilateral Acute Depigmentation of the Iris (BADI) is a condition which was first described in a case series from Turkey by Tugal-Tutkin and Urgancioglu in 2006.1 The condition is characterized by bilateral acute depigmentation and discoloration of the iris stroma, pigment dispersion, and deposition of pigment in the angle. In our case we report one of two identical twin sisters who developed BADI after receiving sarapin injections for chronic migraine, while the other has normal iris architecture and pigmentation and never received any like invasive procedure.

Case Presentation: Patient is a 41 year old female with history of Sarapin injections to her face for chronic migraine who later developed bilateral depigmentation of the iris. She did not have any signs of anterior segment uveitis or iridocyclitis. She has a twin sister who maintained normal iris pigmentation during the entire course.

Discussions and Conclusion: Bilateral Acute depigmentation of the iris is a recently discovered condition described in the literature in Turkish patients1,2. This condition affects mainly young females and is characterized by acute bilateral stromal depigmentation without other pathologic ocular findings. These patients usually maintain normal vision and do not develop significant glaucoma from pigment collecting in the anterior chamber angle. This condition can be mistaken for Fuch's heterochromic iridocyclitis, pigment dispersion syndrome, pseudoexfoliation syndrome, viral iridocyclitis, VZV, HSV, and CMV. This is the first reported case in North America and is important for differentiation of the above pathologies. Our patient had a history of Sarapin injections to the face but it is unsure if this is associated with our patient's development of BADI. As awareness of this condition progresses a possible etiology may be elucidated.

Keywords: Iris; depigmentation; Sarapin, iris transillumination.

Background

Bilateral Acute Depigmentation of the Iris (BADI) is a condition which was first described in a case series from Turkey by Tugal-Tutkin and Urgancioglu in 2006.[1] The condition is characterized by bilateral acute depigmentation and discoloration of the iris stroma, pigment dispersion, and deposition of pigment in the angle. This condition is of an unknown etiology and few cases have been reported in the literature. The initial cases of BADI were reported in Turkey[1,2], however recently a case has been reported in both Brazil[3] and Egypt[4]. This condition is commonly assumed to be a result from an iridocyclitis however does not share any clinical features with this diagnosis. As the etiopathogenesis is yet unknown, it has been speculated that either genetic predisposition or more likely an acute event such as infection, inflammatory, or autoimmune reaction. In our case we report one of two identical twin sisters who developed BADI after receiving sarapin injections for chronic migraine, while the other has normal iris architecture and pigmentation and never received any like invasive procedure.

Case Presentation

Patient is a 41 year old white female with past medical history significant for psoriasis, temporomandibular joint pain, hiatal hernia, and migraine for which she received Sarapin injections at multiple sites in her face and head ,prior to presentation, every month for approximately 12 months. She was in her usual state of health when her identical twin sister noticed that her irises had changed color. She presented to our clinic with a best corrected vision of 20/25 in the right eye and 20/20. Intraocular pressures were 20mmHg in both eyes. Anterior segment exam shows normal eyelids, eyelashes, and adnexa. Conjunctiva and cornea were clear in both eyes as well. There was no anterior chamber cell or flare in either eye and she was without posterior synechiae. Bilateral, symmetric, depigmentation of the peripheral iris stroma was present without transillumination defect (See Figures 1 and 2). She had pigmentary sparing along the pupillary margin in both eyes with no pigment changes in the several overlying iris nevi and freckles. There was noted to be clump-like dusting of pigment throughout iris stroma of both eyes. Anterior exam revealed no signs of stellate keratic precipiates, posterior synechiae. Gonioscopy revealed open angles in both eyes without evidence of deep pigmentation, peripheral anterior synechea or segmental pigment deposition. Dilated fundus examination revealed normal pigmentation, normal optic nerves, and peripheral retina was without tears, holes, or detachments. Patient has been followed for approximately 5 years and her irises have remained depigmented. Her identical twin sister still has no signs of depigmentation of her iris and maintains brown irides at last exam one month ago (See Figure 3).

Discussion And Conclusions

Bilateral Acute depigmentation of the iris is a recently discovered condition described in the literature in Turkish patients[1,2]. This condition affects mainly young females and is characterized by acute bilateral stromal depigmentation without other pathologic ocular findings. These patients usually maintain normal vision and do not develop significant glaucoma from pigment collecting in the anterior chamber angle. This condition can be mistaken for Fuch's heterochromic iridocyclitis, pigment dispersion syndrome, pseudoexfoliation syndrome, viral iridocyclitis, Varicella Zoster Virus, Herpes Simplex Virus, and Cytomegalovirus. Fuch's Heterochromic iridocyclitis is characterized by unilateral presentation in all but a few cases, and is characterized by white-stellate keratic precipitates in the and low grade anterior chamber inflammation[5], which our patient did not have. Pigment dispersion syndrome as well as pseudoexfoliation are conditions characterized by loss of pigment from the posterior iris pigmented epithelium with transillumination defects and accumulation of pigment in the anterior chamber angle as well as along the zonules and anterior lens capsule[6], all of which were absent in our patient as well. Herpetic iridocyclitis is almost always unilateral, and accompanied by eye pain, redness, photophobia, mild to severe anterior chamber inflammation, possible hyphema, keratic preciptates, posterior synechiae, decreased corneal sensation, iris atrophy, irregular pupil, transillumination defects, and elevated intraocular pressure[7]. Again our patient had a complete absence of these findings during her course.

Bilateral Acute Iris Transillumination (BAIT) is another recent entity described in the literature as a condition with acute onset of bilateral iris transillumination defects with loss of associated iris pigment epithelium after using fluoroquinolones and other antibiotics.[8] In our case, our patient did not have transillumination defects and did not take antibiotics before onset of her depigmentation.

BADI is a condition that is characterized by stromal loss of pigmentation with a lack of transillumination. Review of the cases of BADI including ours, we noted one important clinical feature which is a maintenance of iris pigmentation within 1-2mm of the pupil margin. The majority of cases presented in the literature had this common clinical feature[1-4]. BADI is also easily mistaken for iridocyclitis but given the absence of uveitis symptoms and a different pattern of depigmentation, it is a condition which should be readily made clinically. The etiology for such a condition is not well known as only 38.5% of patients in the review by Tugal-Tutkin had a viral prodrome before developing the condition[2]. In our patient she denied any viral prodrome but she reported monthly injections of Sarapin® injections in her face for chronic migraine monthly for a total of 12 months. Sarapin® which is a toxin obtained from pitcher plants, has been extensively studied for its analgesic effects. It acts upon the C fibers of nerves and contains an unidentified toxin that potentiates action of ammonium ions[10], resulting in decreased pain. This injection has never been reported to cause any complications. It is unknown the effect of Sarapin on iris pigmentation. The timing of the treatment to the change in iris color suggests a relationship though coincidence may be the ultimate explanation.

Reports have shown that patients with this condition can have spontaneous re-pigmentation as well, however this has not yet been noted in our patient who we have been following for several years.

Our case is significant as it is the first reported in the Western Hemisphere, as well as the first case amongst identical twins with the unaffected twin sister serving as a “control”. As awareness of this condition increases, an etiogenesis may be discovered as more cases are studied.

Declarations

Abbreviations: Bilateral Acute Depigmentation of the Iris (BADI), Bilateral Acute Iris Transillumination (BATI)

Ethics approval and consent for participate: Not Applicable

Consent for Publication: Consent was obtained for publication of this article from both the patient and her twin sister serving as the “control”, and was obtained in both written and verbal form.

Availability of Data and Materials: Not Applicable

Competing Interests: The Authors declare that they have no competing interests in this article

Funding: No funding was obtained for the preparation of this article

Author Contributions: SL was involved in drafting of this manuscript and revising critically for important intellectual content. AG was involved in drafting of this manuscript and revising it critically for important intellectual content. BPM was involved in drafting of this manuscript and revising it critically for important intellectual content. All authors gave final approval for the publication of this manuscript and have participated sufficiently in the work to take public responsibility for appropriate portions of the content agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Acknowlegements: Not applicable

References

1. Tugal-Tutkun I, Urgancioglu M. Bilateral Acute Depigmentation of the Iris. Graefes Arch Clin Exp Ophthalmol. 2006 Jun;244(6):742-6. Epub 2005 Oct 5.
2. Tugal-Tutkun I, Araz B, Taskapili M, et. Al. Bilateral Acute Depigmentation of the Iris: Report of 26 New Cases and Four-Year Follow-Up of Two Cases. Ophthalmology. 2009 Aug;116(8):1552-7, 1557
3. Maestrini HA, Maestrini AA, Machado Dde O, et. Al. Bilateral Acute Depigmentation of the Iris: The First Reported Case in Brazil.Arq Bras Oftalmol. 2013 Jan-Feb;76(1):42-4.
4. Amin R, Nabih A, Khater N. Bilateral Acute Depigmentation of the Iris in Two Sibling Simultaenously. Am J Ophthalmol Case Rep. 2018 Mar 16;10:257-260.
5. Accorinti M, Spinucci G, Pirraglia MP, et. Al. Fuchs' Heterochromic Iridocyclitis in an Italian Tertiary Referral Centre: Epidemiology, Clinical Features, and Prognosis. J Ophthalmol. 2016;2016:1458624. Epub 2016 Oct 3.
6. Niyadurupola N, Broadway DC. Pigment dispersion syndrome and pigmentary glaucoma- a major review. Clin Exp Ophthalmol. 2008 Dec;36(9):868-82.
7. Siverio Júnior CD, Imai Y, Cunningham ET Jr. Diagnosis and Management of Herpetic Anterior Uveitis. Int Ophthalmol Clin. 2002 Winter;42(1):43-8.
8. Knape RM, Sayyad FE, Davis JL. Moxifloxacin and Bilateral Acute Iris Transillumination. J Ophthalmic Inflamm Infect. 2013 Jan 14;3(1):10.
9. Reisner L. Biologic Poisons for Pain. Curr Pain Headache Rep. 2004 Dec;8(6):427-34.
10. Manchikanti L, Pampati V, Bakhit CE, et.al. Effectiveness of Lumbar Facet Joint Nerve Blocks in Chronic Low Back Pain: A Randomized Clinical Trial. Pain Physician. 2001 Jan;4(1):101-17.
11. Manchikanti KN, Pampati V, Damron KS, et.al. A Double-Blind, controlled evaluation of the value of sarapin in neural blockade. Pain Physician. 2004 Jan;7(1):59-62.

Figures

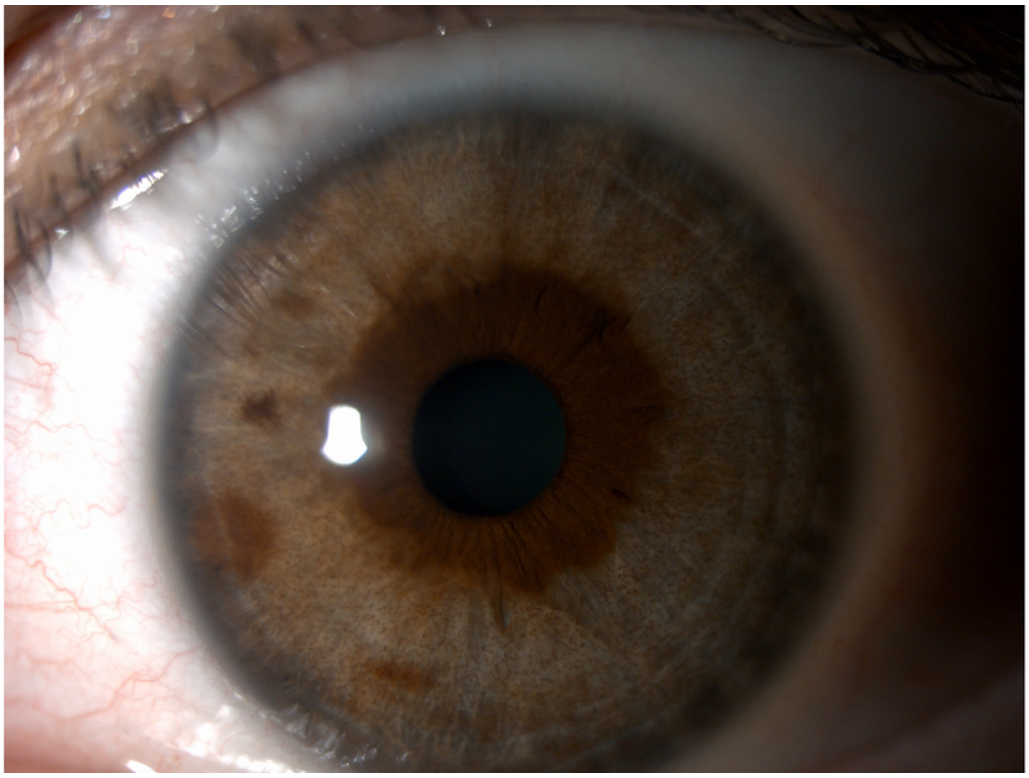

Figure 1

Depigmentation of the iris stroma of the right eye with preservation of stromal pigment within 2mm of the pupillary margin.

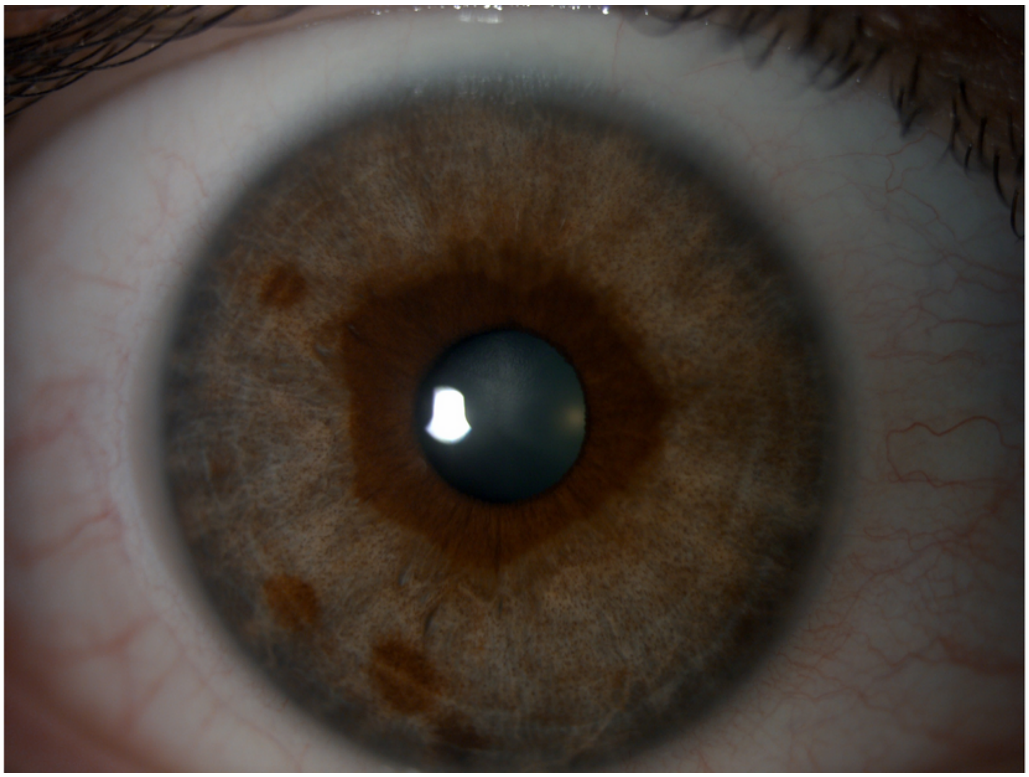

Figure 2

Depigmentation of the iris stroma of the left eye with preservation of stromal pigment within 2mm of the pupillary margin.

Figure 3

Depigmentation of the iris of both eyes in one twin sister (on the right) compared to normal pigmentation of the other twin sister (on the left)

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- supplement1.jpg