Iridoschisis in Latin America: a Case Report and Literature Review

Abstract

Background: Iridoschisis is a rare condition characterized by the thinning and cleavage of the anterior stroma of the iris; usually bilateral and associated with narrow angles, ocular hypertension and glaucomatous optic neuropathy. We report the case of three Latin American patients with iridoschisis; all of them associated with narrow angles, and one with history of chronic angle closure glaucoma. The patients had no previous history of ocular trauma or inherited ocular disease. As a new finding, we present a case of unilateral hyphema as the presenting sign of this entity.

Conclusions: Chronic open-angle glaucoma, as well as intermittent angle-closure glaucoma, should be excluded in all patients with iridoschisis, and regular follow-up should be established after diagnosis. After having presented an unusual way of presentation such as unilateral hyphema (with no previous documented reference of this sign), we have widened the spectrum of clinical findings for the suspicion and diagnosis of this pathology.

Keywords: iridoschisis, angle-closure glaucoma, iris degeneration.

Introduction

Iridoschisis is a rare condition that consists of intrastromal atrophy with separation of the anterior mesodermic layer of the iris from the posterior, with subsequently extensive splitting with portions of the anterior layer floating freely in the aqueous humour. This condition was first described in 1922 by Schmitt, becoming the first person to present a case report of “Iris splitting” in 1945. Lowenstein proposed the term Iridoschisis after describing a localized cleavage of the iris’ stroma in two layers in an elderly patient with absolute glaucoma. This condition usually manifests bilaterally and localized in the inferior iris, occurring more frequently between the fifth and the seventh decade of life, with no identified pathophysiology and is frequently associated with angle closure glaucoma, idiopathic iris atrophy related to age, congenital abnormalities, congenital syphilis or previous history of trauma.

In Colombia, there have been two case reports of patients with this condition: the first published in 1996, associated with glaucoma, and the second one in 2011 without this association. In the current report, we describe 3 cases of Iridoschisis: one of them associated with angle closure glaucoma and another one emphasizing an atypical presentation form, with unilateral hyphema as an initial manifestation; which to the best of our knowledge, is the first case report of Iridoschisis with hyphema as the first clinical sign of the disease.
A 70-year-old man was referred to the Ophthalmology emergency unit of the Military Central Hospital in Bogota, Colombia, presenting with a 1-day history of visual loss from the right eye. The patient had no previous history of ocular trauma or inherited ocular disease. He was on antihypertensive treatment. His best-corrected visual acuity (VA) was 6/6 bilaterally, and the intraocular pressure at the time of admission was 28 mmHg in both eyes. At slit-lamp examination, showed sectorial splitting of the right iris stroma; inferior and nasally from the 4 to the 6 o’clock position (Figure 1A). Both anterior chambers were remarkably shallow and in the right eye, loose ends of degenerated anterior stromal leaf fibrils were observed floating freely in the aqueous. Gonioscopy showed a closed angle (Shaffer grade 0–1) for 270° in both eyes (Figure 1B). The lens showed a mild cataract and a posterior segment examination revealed a healthy optic disc and macula. Visual fields were normal. Ultrabiomicroscopy showed narrow angles and marked iris convexity. Syphilis serology was negative. Diagnoses of right idiopathic iridoschisis and narrow angles were made. The patient was scheduled for bilateral peripheral laser iridotomies, an ocular hypotensive agent was started and a 6-month follow up was scheduled for glaucoma review.

**Case 2**

A 72 years-old man presented to the Ophthalmology emergency unit of the Military Central Hospital in Bogota (Colombia), with a 6-month history of visual loss from the left eye. The patient had previous history of closed angle glaucoma currently treated with drops and iridotomy. His best-corrected visual acuity (VA) was 6/24 OD and CF (count fingers) in OS. The intraocular pressure at the time of the admission was 12 mmHg in both eyes and the slit-lamp examination showed narrow anterior chamber and localized splitting of the left iris stroma inferior-nasally (Figure 2A). The pupil was round...
and reactive to light and accommodation. Gonioscopy showed permeable iridotomy in both eyes, and angles Shaffer grade 3 in both eyes. Pseudophakic in OD and mature cataract in OS. Fundoscopy revealed evidence of glaucomatous disc damage, with bilateral vertical cup to disc ratios of 0.9 and thinning of neuroretinal rims. Ultrabiomicroscopy showed narrow angles, iridotomy in both eyes and splitting of the iris in OS. Syphilis serology was negative. A diagnosis of left iridoschisis associated with history of anterior closure glaucoma was made, the patient was scheduled for trabeculectomy combined with cataract extraction in OS, showing an increase on the splitting of the iris, and an increase in the number of fibrils floating in anterior chamber in the postoperative period (Figure 2B).

Case 3

A 59 years-old woman, presented with a 1-day history of visual loss from the right eye associated with pain. The patient had no previous history of ocular trauma or of inherited ocular disease. She was on antihypertensive treatment. The intraocular pressure at the time of the admission was 14/16 mmHg. Her best-corrected visual acuity (VA) was 6/7.5 bilaterally, and a slit-lamp examination showed OD with moderate conjunctival hyperemia, narrow anterior chamber with 5% hyphema, and localized splitting of the both iris stroma inferior nasally (Figure 3). Gonioscopy showed Shaffer grade 2 angles in both eyes and the lens showed mild cataract. A posterior segment examination revealed a healthy optic disc and macula. Syphilis serology was negative. A diagnosis of bilateral idiopathic iridoschisis, narrow angles, and right hyphema was made. The patient was scheduled for bilateral peripheral laser iridotomies and 6-month follow up for glaucoma review.

Discussion

Iridoschisis is a rare condition that consists of separation of the anterior stroma of the Iris from the posterior with subsequent splitting of the anterior layer and portions of the anterior layer floating freely in the aqueous humor. This condition usually manifests bilaterally and localized in the inferior iris, occurring more frequently between the fifth and the seventh decade of life. Iridoschisis and glaucoma are closely associated, especially angle closure glaucoma, which has been reported to occur in approximately 40% of eyes with iridoschisis. Until 1999 there have been 72 documented cases of iridoschisis from which 28 cases were associated with angle closure glaucoma and 2 cases associated with open-angle glaucoma. Since then, 5 new cases of Iridoschisis related to angle closure glaucoma have been reported.5-9

Iridoschisis not associated with glaucoma occurs in elderly patients; usually between fifth and seventh decade of life, but there have been three reports of iridoschisis without glaucoma in juveniles as young as 8 years old.1,10,11 However, it is not common the presence of corneal alterations. There are 4 recent reports of corneal endothelial decompensation presenting with bullous keratopathy,12-15, and three cases associated with keratoconus.16 Iridoschisis does not seem to be gender specific nor appears to have related involved genes.17

The pathophysiology of this entity has not been yet identified. Lowenstein and Foster suggested that there is an anatomic basis leading to an atrophic effect caused by lytic substances in the aqueous, attributed to senile changes, or that it is originated from a glaucomatous condition. Later they proposed another theory associating it with a history of trauma, assuming that a posttraumatic peak of intraocular pressure could lead to the splitting of the anterior and posterior stroma,
shearing along the dilator fibers. Albers and Klein also attributed this finding to senile changes with sclerosis of blood vessels in the anterior stroma, which could tear the tissue between the anterior and posterior stroma during the constriction and dilation of the iris. There is normal perfusion by blood vessels from the inner pupillary margin to the outer iris of affected sectors. In histopathological studies, it has been found that tissue fibrosis and atrophy with evidence of small clefts. There is also thinning of the stroma with diminished number of collagen fibrils in the affected area, demonstrated by the use of electron transmission microscopy. Corneal changes correspond to degenerated endothelial cells localized over the affected area, being rare but, if present, may be produced by contact between strands of the degenerated iris and the corneal endothelium.

As mentioned before, iridoschisis is frequently associated with angle closure glaucoma, idiopathic atrophy in aged eyes, congenital abnormalities, congenital syphilis or a history of trauma. Specifically angle closure glaucoma has been proposed as a consequence of iridoschisis. Supporting that, a study made by Salmon and Murray proposed that primary angle closure glaucoma should be excluded in patients who present iridoschisis. On the other hand, supporting the idea that iridoschisis might be part of an anterior anatomic malformation, there have been case reports of complete and incomplete plateau iris configuration associated to iridoschisis with anterior location of ciliary body. Lens subluxation, either posterior or anterior, with secondary shallowing of the anterior chamber causing angle closure glaucoma and in other cases hand rubbing against the iris as a mechanical trauma have been described as a precipitating factor of iridoschisis. Nanophthalmos and one case with proven anterior segment dysgenesis has been documented as other related condition.

Those findings seems to support the theory of a familial character, as mentioned by Danias et al, in which iridoschisis evidenced by ultrasound biomicroscopy was found in a father and his daughter. The pattern of inheritance suggested is autosomal dominant with deletion of short (p) arm of chromosome X. There has also been a number of reported cases of iridoschisis and interstitial keratitis due to syphilis.

The most important differential diagnosis includes iris stroma anomalies, Axenfeld-Rieger syndrome or iris, corneal neurocristopathy, and iridocorneal endothelial syndrome (ICE), in addition to congenital uveal ectropion syndrome and essential iris atrophy.

The diagnosis of iridoschisis is made based on the clinical characteristics, age of onset, laterality, progression or coexistence with other pathologies. Table 1 shows the clinical features found in these 3 cases, remarking that the age of presentation in all patients was between the fifth and the seventh decade of life with an inferior iris location of the defect as reported in the majority of studies published to date. In all three cases, the presence of narrow angles in gonioscopy was a common feature and the diagnosis of ACG in one of the exposed cases, corresponding to a percentage of 33%, was similar to that reported in the available literature.

The detection of iridoschisis alone is no indication for an intervention; however, when associated with high intraocular pressure, medical or surgical control with laser iridotomy and even extraction of lens is essential for controlling intraocular pressure and/or the resolution of the angular closure in the cases that require it. Some authors have postulated the management with prophylactic iridectomy and removal of the floating flaps of fibers that can play a role in preventing corneal decompensation, and even penetrating keratoplasty in some cases.

**World burden**

To date, there are almost 100 cases reported in the world literature of Iridoschisis, distributed mostly in North America, United Kingdom and Africa. As far as South America is concerned, there are only 3 reported cases of patients with iridoschisis: one in Brazil, corresponding to the first case reporting the association between iridoschisis and non-arteritic ischemic optic neuropathy, and the other two published in Colombia; the first in 1996 showing association with ACG and the second as a demonstration of the technique of cataract surgery in a patient with iridoschisis.

**Conclusion**

Chronic open-angle glaucoma, as well as intermittent angle-closure glaucoma, should be excluded in all patients with iridoschisis, and regular follow-up should be established in search of progression of the pathology. Ruling out the presence of conditions such as syphilis by means of serology should also be done. In this article, we report an unusual presentation of iridoschisis with unilateral hyphema, with no previous reference of this sign in the literature, widening the spectrum of clinical findings for the suspicion and diagnosis of this pathology, and also supporting the current literature, we found as a common characteristic an inferior iris location of the defect in all three cases, as reported in the majority of studies published to date.

The low incidence in South America lead us to ask...
ourselves, why the lower incidence of this pathology in our continent, and if it is affected by geographical factors, or perhaps the need to increase the publication of cases not reported in the literature in areas with low accessibility to health centers with access to specialists in ophthalmology trained to diagnose the disease.

References