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CASE REPORT

Juvenile Iridoschisis and Dense Cataract in Severe Atopic Eczema: A Rare Case Report and Literature Review

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ABSTRAK

Iridoskisis juvenil ialah gangguan mata yang jarang berlaku, dicirikan oleh pemisahan stroma iris yang biasanya diperhatikan dalam kalangan warga emas tetapi amat jarang berlaku dalam kes pediatrik. Penyebabnya masih belum jelas, walaupun pelbagai jenis hipotesis telah dikemukakan termasuk kecenderungan genetik, trauma mekanikal dan mekanisme keradangan. Walaupun iridoskisis dikaitkan dengan penyakit seperti katarak, glaukoma penutupan sudut dan subluksasi kanta, hubungannya dengan ekzema atopik masih kurang difahami. Kami melaporkan satu kes luar biasa iridoskisis juvenil dalam pesakit pediatrik yang mengalami ekzema atopik yang teruk. Kajian sistematik terhadap laporan-laporan yang diterbit terdahulu telah dijalankan untuk menganalisis corak klinikal, penemuan okular yang berkaitan dan mekanisme penyebab. Selain itu, iridosiklitis yang berlaku dalam pesakit ini menimbulkan persoalan tentang penglibatan keradangan dalam proses iridoskisis, satu ciri yang jarang dilaporkan dalam penyakit ini. Analisis proteomik terkini mencadangkan peningkatan kebolehtelapan saluran darah iris dan kebocoran protein dalam iridoskisis yang mungkin memberikan pemahaman baharu mengenai patogenesisnya. Selain itu, tabiat menggosok mata secara berulang yang lazim berlaku dalam kalangan pesakit ekzema atopik, telah dicadangkan sebagai pencetus mekanikal; namun, peranannya masih belum dapat disahkan. Kekurangan rawatan susulan jangka panjang dalam kes-kes yang dilaporkan menimbulkan jurang yang ketara dalam pemahaman mengenai perkembangan penyakit, risiko kejadian glaukoma dan komplikasi jangka panjang yang berpotensi berlaku. Kajian ini menekankan keperluan untuk penyelidikan lanjut mengenai hubungan antara atopi, biomekanik okular dan disregulasi imuniti dalam iridoskisis juvenil, serta kepentingan strategi pengurusan pembedahan dan pascaoperasi untuk pesakit yang terjejas.

Kata kunci: *Ekzema atopik; iridoskisis juvenil; katarak*

ABSTRACT

Juvenile iridoschisis is a rare ophthalmic disorder characterised by the splitting of the iris stroma, typically observed in elderly individuals but exceptionally uncommon in paediatric cases. The aetiology remains unclear, though various hypotheses, including genetic predisposition, mechanical trauma and inflammatory mechanisms have been proposed. While iridoschisis has been associated with conditions such as cataract, angle-closure glaucoma and lens subluxation, its link to atopic eczema is poorly understood. This report highlights a rare case of juvenile iridoschisis in a paediatric patient with severe atopic eczema, adding to the limited body of literature on this condition. A systematic review of

previously published cases was conducted to analyse clinical patterns, associated ocular findings and potential aetiopathogenic mechanisms. Notably, the presence of concurrent iridocyclitis in this patient raises questions about inflammatory involvement in the disease process, a feature rarely reported in juvenile iridoschisis. Recent proteomic analyses suggest increased vascular permeability and protein leakage in iridoschisis, which may provide new insights into its pathogenesis. Additionally, habitual eye rubbing, a common feature of atopic eczema has been proposed as a mechanical trigger; however, its role remains inconclusive. The lack of long-term follow-up in reported cases presents a significant gap in understanding disease progression, the risk of glaucoma and potential long-term complications. This study underscores the need for further research into the interplay between atopy, ocular biomechanics and immune dysregulation in juvenile iridoschisis, as well as the importance of tailored surgical and postoperative management strategies in affected patients.
Keywords: Atopic eczema; cataract; juvenile iridoschisis

INTRODUCTION

Iridoschisis is a rare condition predominantly observed in elderly patients, with its occurrence in paediatric cases being exceptionally uncommon (Pieklarz et al. 2020). We presented a rare case of juvenile iridoschisis associated with a white cataract, low grade iridocyclitis and atopic eczema. We had also compiled a list of previously published cases of juvenile iridoschisis, detailing their characteristics and associated ocular conditions.

CASE REPORT

A 12-year-old Malay boy with severe atopic eczema requiring oral steroids, cyclosporine and azathioprine for disease control, presented with progressive, generalised blurring of vision in his left eye over one year. His mother noticed unilateral leukocoria, prompting an ophthalmology consultation. He denied any eye redness, pain, discharge, habitual eye rubbing or trauma prior to the blurring of vision. Besides of eczema, he also had underlying allergic rhinitis which was not under proper follow up. There was no family history of atopy, blindness, glaucoma or pre-senile cataract.

On examination, visual acuity was hand movements in the left eye, with no relative afferent pupillary defect (RAPD). The cornea was clear but prominent corneal nerves were seen. The anterior chamber was shallow peripherally (Van Herick grade 2) but moderate centrally, with 1+ mixed pigmented and non-pigmented cells without flare. Intraocular pressure (IOP) was 18 mmHg. Sectoral iridoschisis was noted from 7 to 10 o'clock at the mid-iris (Figure 1), with floating iris strands contacting the corneal endothelium but no iris atrophy (Figure 2). Iris transillumination was negative. The left eye had a mature, intumescent white cataract without phacodonesis. There were no keratic precipitate, iris nodule or posterior synechiae suggestive of prior uveitis. To prevent pupillary block, pharmacologic dilation of the left eye was avoided. B-scan ultrasound showed a clear vitreous and flat retina. Anterior segment optical coherence tomography (ASOCT) showed

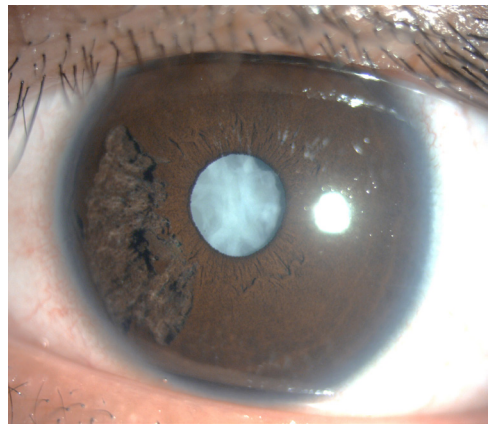

FIGURE 1: Photo of left eye showing sectoral iridoschisis and a dense white cataract

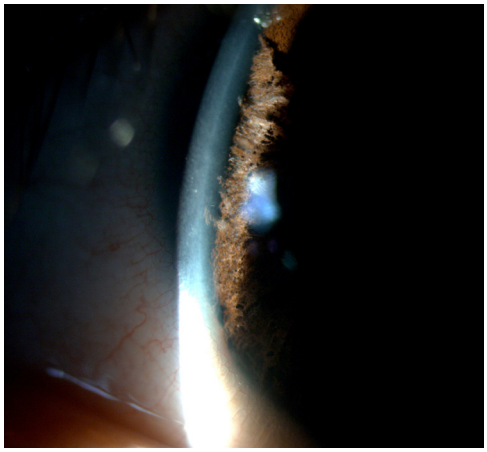

FIGURE 2: Photo showing shallow anterior chamber with free floating iris strands in contact with cornea endothelium

a "shredded" appearance of anterior surface of iris (Figure 3).

The right eye examination revealed 6/6 vision, normal anterior chamber depth and fundus. There was no evidence of iridoschisis or cataract. Occasional pigmented anterior chamber cells were observed. Systemic examination revealed Hertoghe's sign and diffuse skin excoriation over face. Extensive skin xerosis, lichenification and excoriations were also noted on body and extremities. Otherwise there were no signs suggestive of other systemic or syndromic diseases such as Marfan syndrome or congenital syphilis.

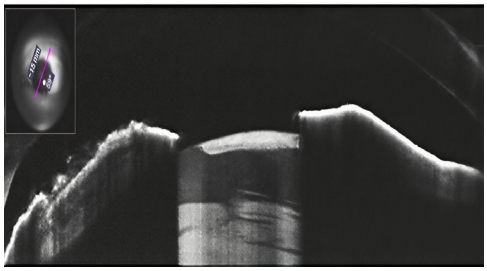

FIGURE 3: Anterior segment optical coherence tomography cutting across the schitic iris showing "shredded" appearance of anterior surface in contrast to the opposite iris which was normal

The patient was prescribed with topical dexamethasone 0.1% for both eyes and scheduled for left eye cataract extraction with intraocular lens (IOL) implantation. Anterior chamber activity decreased with topical dexamethasone treatment and achieved quiescence on follow-up. Two weeks prior to surgery, the dosage of topical dexamethasone was increased to every four hours and maintained until the day of operation. Lens aspiration with implantation of a single-piece acrylic monofocal IOL was performed under general anaesthesia. Intraoperatively, the pupil was well dilated, revealing a white cataract with a dense anterior subcapsular plaque (Figure 4). The surgery proceeded uneventfully as the iridoschisis was located opposite the aspiration port and did not interfere with the procedure. IOL was successfully implanted in the capsule. Post operatively, the patient was prescribed topical corticosteroids with a longer tapering regimen of two months. At two months postoperatively, the patient recovered well and achieved a best-corrected visual acuity of 6/7.5 without any evidence of rebound intraocular inflammation or raised IOP. Postoperative retroillumination testing revealed no transillumination defects (Figure 5), supporting the diagnosis of iridoschisis rather than iris atrophy. However, at 3 months postoperatively, there was a reduction in visual acuity to 6/12, with diffuse iris pigment noted on

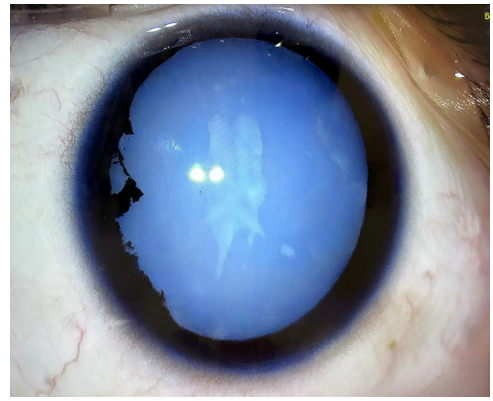

FIGURE 4: Intraoperative photo showing a well dilated eye, white cataract and central dense anterior subcapsular cataract

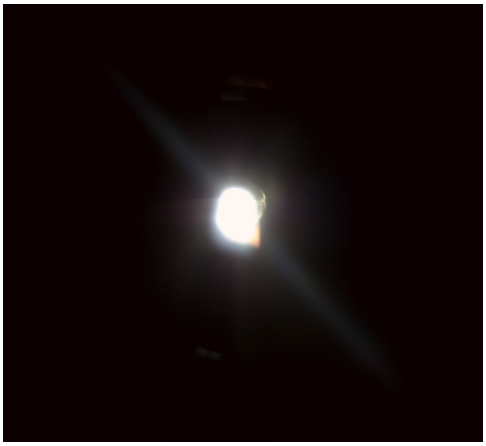

FIGURE 5: Postoperative retroillumination photography showed no iris transillumination defects, suggesting the absence of iris atrophy

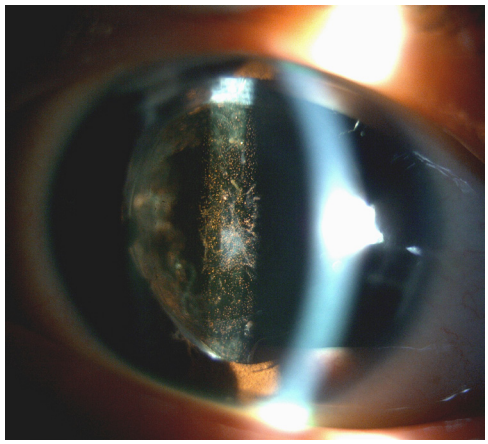

FIGURE 6: Post operative photo of dilated pupil showing diffuse iris pigment on anterior lens

the anterior surface of the IOL (Figure 6). There were no signs of anterior chamber inflammation or posterior capsular opacity. We opted for observation, monitoring for any worsening of pigment deposition or a rise in IOP.

DISCUSSION

Iridoschisis is characterised by the splitting of the anterior and posterior iris stroma, with disintegration of the anterior iris layer into floating fibrils. The exact pathogenesis remains unclear, though several theories have been proposed, including trauma, senile changes, degenerative processes and possible hereditary factors (Pieklarz et al. 2020). Iridoschisis has also been reported in association with various ocular conditions, including angle-closure glaucoma, plateau iris, cataract, lens subluxation, syphilitic eye disease, corneal decompensation and keratoconus (Gogaki et al. 2011; Pieklarz et al. 2020).

While iridoschisis primarily affects elderly patients, only a handful of juvenile cases have been documented in the literature as summarised in Table 1 (Aaberg Jr & Nelson 2017; Chapman & Demetriades 2015; Johnson & Bachynski 1986, Krohn & Garrett 1954; Summers et al. 1985; Yusuf & Salmon 2016). Juvenile iridoschisis has been reported in a wide age range (5-24 years), with no clear gender predilection. The condition appears to be more commonly unilateral (5 cases) than bilateral (2 cases). The location of iridoschisis varies, but commonly involves inferior or sectoral portions of the iris. While the exact mechanism of juvenile iridoschisis remains elusive, its frequent association with concurrent ocular pathology suggests a dependent rather than isolated occurrence. The absence of reported perinatal cases of iridoschisis suggests that the condition is unlikely to be congenital. Salmon & Murray (1992) proposed that iridoschisis may be a manifestation of elevated IOP in primary angle-closure glaucoma. However, all reported juvenile cases have shown no evidence of angle closure. Only one reported case has documented significantly elevated IOP.

To date, only one case of concurrent iridoschisis and cataract in a paediatric patient with eczema has been reported in the literature (Aaberg Jr & Nelson 2017). Cataracts are known to associate with atopic eczema, typically presenting as bilateral, symmetrical opacities affecting the subcapsular regions (Nagaki et al. 1999; Pietruszyńska et al. 2020). However, literature linking atopic eczema to iridoschisis is scarce. Kusano et al. (2025) reported three cases of adult patients with atopic dermatitis who also

TABLE 1: Characteristics of patients in previous case reports of juvenile iridoschisis

Case report	Age	Gender	Systemic comorbidity	Laterality	Visual acuity	Iridoscisis location	Lens	Other ocular comorbidity	IOP (mmHg)	Gonioscopy	Endothelial cell count (cell/mm^3)	Investigation
Krohn & Garrett (1954)	20	Male	Nil	OU	OD: 6/21 OS: 6/9	Diffuse	OD: white cataract OS: mild cataract	OU Keratoconus	16	Open angle	-	Spinal fluid analysis: normal
Summers et al. (1985)	8	Female	Nil	OD	6/7.5	2 - 10 o'clock	Normal	Intermittent right esotropia, microphthalmos	22	Open angle	2880	Chromosomal analysis: Normal 46 XX karyotype
Johnson & Bachynski (1986)	5	Male	Nil	OD	6/30	3 - 6 o'clock	Normal	Microphthalmos, sectoral corneal opacification involving visual axis	18	Open angle	-	-
Chapman & Demetriades (2015)	22	Male	Bilateral sensorineural hearing loss, Ex-premature baby 35 week	OU	OU: 6/6	OD: Diffuse OS: 2-7 o'clock	Normal	Incomplete plateau iris configuration, macular RPE mottling	Normal	Open angle	OD: 2104 OS: 2331	Syphilis serology: negative
Yusuf et al. (2016)	24	Male	Periocular atopic dermatitis	OU	OD: 6/7.5 OS: 6/9	Inferior asymmetry	Normal	Allergic conjunctivitis, keratoconus	15	Open angle	-	-
Aaberg Jr & Nelson (2017)	11	Female	Severe facial and periocular eczema	OD	HM	3 - 10 o'clock	White cataract	Nil	19	Open angle	-	Syphilis serology: negative
Pegu et al. (2020)	23	Male	-	OU	-	OD: 12 – 3 o'clock OS: 6 - 11 o'clock	Normal	Juvenile open angle glaucoma	OD: 29 OS: 26	Open angle	-	-

HM: Hand movement; IOP: Intraocular pressure; OU: Both eyes; OD: Right eye; OS: Left eye; RPE: Retina pigment epithelium

had iridochisis. Patients with atopic eczema are often prone to habitual chronic eye rubbing, which can result in mechanical trauma to the iris and elevated IOP, potentially contributing to the development of iridoschisis (Yusuf & Salmon 2016). However, our patient denied any history of eye itchiness or rubbing and developed a unilateral dense cataract and iridoschisis, which is an atypical presentation compared to previously reported cases. Our patient's presentation closely aligns with the case reported by Aaberg and Nelson (2017), raising the possibility of a shared pathophysiology or an underlying oculodermal syndrome.

One notable finding in this case was the presence of subtle anterior chamber activity, evidenced by mixed pigmented and non-pigmented cells. While it is logical to assume that the cleaved iris stroma may release iris pigment, iridocyclitis has been reported only once in the literature as an associated feature of iridoschisis (Mansour 1986). We remained skeptical about prior anterior uveitis as a causative factor of iridoschisis and cataract, given the absence of other hallmark signs of uveitis such as circumciliary injection, keratic precipitates, iris nodules or posterior synechiae. Kusano et al. (2025) found surprisingly high levels of total protein in the anterior chamber by proteomic analysis of the aqueous humor obtaining during keratoplasties in iridochisis patients, suggesting increased vascular permeability and seepage of proteins presumably from the iridoschitic sites. Fluoroiridographic investigations conducted by Carnevalini et al. (1988) in three patients with iridoschisis demonstrated normal iris vessel perfusion and stromal diffusion. However, one patient exhibited a slight pupillary dye leak. These findings may explain the presence of mixed pigmented and non-pigmented cells in our patient's anterior chamber and their responsiveness to topical steroid treatment. We initiated an intensified regimen of topical corticosteroids priming two weeks prior to surgery to mitigate the heightened risk of postoperative intraocular inflammation in paediatric patients undergoing cataract surgery. Careful consideration must be given to balancing this approach against the potential risks of IOP elevation and exacerbation of intumescent cataract. Postoperatively, there was an unusually heavy pigment deposition on the IOL despite a quiescent anterior chamber. To date, there is no published literature describing IOL pigment deposition following cataract surgery in patients with iridoschisis. We postulate that pigment dispersed from the iridoschitic iris may have adhered to the IOL surface. IOL exchange may be the only viable option if the pigment deposition progresses and vision continue to deteriorate. However, additional intraocular manipulation carries the risk of exacerbating pigment dispersion, potentially worsening the condition. This highlights a previously unrecognised, difficult-to-manage and potentially sight-threatening complication associated with iridoschisis.

One major limitation in understanding juvenile iridoschisis is the lack of long-term follow-up data in reported cases. None of the published reports provide insights into disease progression, potential complications, or long-term visual outcomes. Adler & Weinberg (2004) reported a compelling case demonstrating the progression of iridoschisis over time. By comparing a recent anterior segment photograph with an image taken 25 years earlier for iridology purposes, they provided rare longitudinal evidence of the condition's evolution. This finding suggests that iridoschisis may be a progressive condition. However, the risk of glaucomatous progression notably angle closure, corneal endothelial compromise or recurrent inflammation in these patients remain unknown.

CONCLUSION

This case contributes to the growing but still limited understanding of juvenile iridoschisis. The concurrent findings of iridoschisis, cataract, low-grade iridocyclitis and atopic eczema suggest a possible interplay of mechanical, inflammatory and genetic factors rather than a single causative mechanism. Further studies are needed to elucidate the role of atopy, ocular

biomechanics and immune dysregulation in the development of iridoschisis in younger patients. The presence of dense pigment deposition on the IOL may represent a rare, under-recognised complication of iridoschisis with potential impact on vision and management. There is also a need for longitudinal studies to determine whether juvenile iridoschisis requires lifelong monitoring, prophylactic intervention or specific management strategies to prevent late-onset complications.

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