Iridoschisis

Subjects: Ophthalmology

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Iridoschisis is a rare condition defined as a separation of the anterior iris stroma from the posterior stroma and muscle layers. A variety of ocular pathologies can coexist with iridoschisis, but in most cases the causal relationship, if any, is unclear. Glaucoma, primarily angle-closure glaucoma, is the most often described condition concomitant to iridoschisis. Other ocular abnormalities found relatively often in iridoschisis patients include cataract, lens subluxation and corneal abnormalities. Iridoschisis is plausibly a multifactorial disease. A patient diagnosed with iridoschisis should be screened for potential glaucoma, corneal and lens abnormalities. Iridoschisis may pose a challenge for both an ophthalmologist in an outpatient setting and an ophthalmic surgeon.

iridoschisis | iris degeneration | free-floating iris tissue | rare ocular disease | angle-closure glaucoma

1. Introduction

Iridoschisis is a rare condition defined as a separation of the anterior iris stroma from the posterior stroma and muscle layers. In iridoschisis patients, the iris strands float in the aqueous humor and create a “shredded wheat” appearance [1]. The term iridoschisis (iris splitting) was first introduced in 1945 by Lowenstein and Foster, who reported a deep, parallel split between the anterior and posterior stromal layers of the iris [2]. However, the condition was first described as early as in 1922 by Schmitt who reported on the detachment of the anterior iris layer [3]

2. Epidemiology, Inheritance and Pathophysiology

Only about 150 cases of iridoschisis have been reported to date, with a slight predominance of female patients over males [4]. Some authors speculated that iridoschisis might be inherited in an autosomal dominant manner, but these reports are scarce [4][5]. However, some researchers suggested that iridoschisis is not inherited but occurs sporadically, secondary to trauma, glaucoma or syphilis [2][6][7][8][9]. Other authors considered iridoschisis age-related atrophy, given that the condition is found predominantly in persons between 60 and 70 years of age [10] However, it needs to be stressed that iridoschisis has also been reported in youths [11][12]. A number of theories exist regarding the pathogenesis of iridoschisis, but the exact underlying mechanism of this condition is yet to be explained. For example senile changes, trauma and ischemia were suspected as predisposing factors [2][6][13][14][15][16]

3. Clinical Characteristics and Diagnostic Imaging

While iridoschisis may be unilateral at its early stages, it occurs bilaterally in most cases [11] and may have a progressive character [17]. The condition is most often found in the inferior irideal quadrants, but also other parts of the iris may be affected, and sometimes the pathological process is spread across the whole iris [1][4][18][19]. The anterior iris stroma splits from the posterior stroma and muscle layers, and the loose ends wave in the aqueous humor of the anterior chamber, giving the iris a “shredded” appearance (Figure 1).

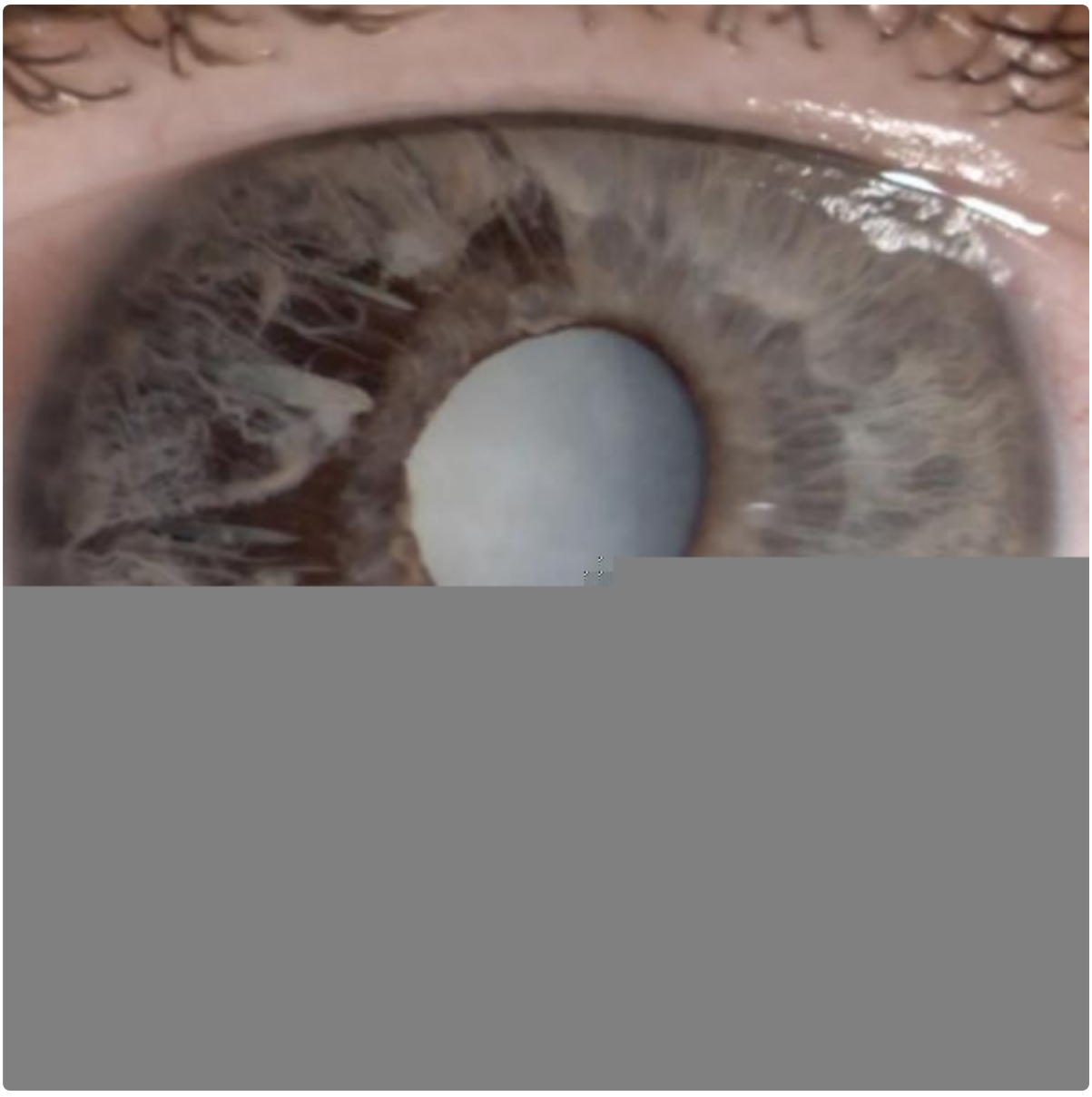

Figure 1. “Shredded” appearance of the iris: superotemporal iridoschisis and mature cataract. Adapted from “Iris-claw lens implantation in a patient with iridoschisis” by Pieklarz B, Grochowski E, Dmuchowska DA, Saeed E, Sidorczuk P, Mariak Z. Am J Case Rep, 2020; 21: e925234.

The posterior layer of the iris usually remains intact with the retained function of the sphincter and dilator fibers [13] Visual deterioration may be caused by glaucoma, cataract or corneal decompensation secondary to iridocorneal

touch [20]Anterior segment optical coherence tomography (AS-OCT), ultrasound biomicroscopy (UBM) and Scheimpflug imaging are complementary diagnostic options in patients with suspected iridoschisis (Figure 2) [10][21][22]

Figure 2. Anterior segment optical coherence tomography (AS-OCT): disorganization of the iris stroma corresponding to iridoschisis.

4. Associated Ocular Pathologies

Iridoschisis may coexist with an array of other ocular pathologies as reviewed by Pieklarz et al. [23], and in most of the cases, it is unclear whether it occurs as a cause or effect or just by coincidence. Glaucoma, primarily angle-closure glaucoma, is found in more than two-thirds of patients with iridoschisis [1]. A coexistence of iridoschisis with chronic open-angle glaucoma or angle recession glaucoma has been reported as well [9][19][24]. Some published evidence suggests that iridoschisis may coexist with presenile cataract and mature cataract [1][12][25][26][27]. While rarely, iridoschisis may also be found concomitantly to lens subluxation [7][19][25][28][29][30]. Corneal changes are uncommon and, if present, the degenerated corneal endothelial cells are mostly localized above the area of iridoschisis [31][32][33][34][35]. Total endothelial decompensation has been reported as well [36]. A rare combination of iridoschisis with keratoconus has been also reported [37][38].

5. Summary

The prevalence of iridoschisis is difficult to estimate. It is probably underreported as no dedicated registry for this condition exists. Iridoschisis is plausibly a multifactorial disease that requires particular attention from ophthalmologists and ophthalmic surgeons. A patient diagnosed with iridoschisis should also be screened for potential glaucoma, corneal and lens abnormalities. If not yet present, one of these conditions may subsequently develop, and hence iridoschisis patients should be followed-up on a regular basis.

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