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Strabismus revealing Axenfeld's anomaly with ocular hypertension

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Abstract:
Introduction*: Axenfeld's anomaly is a rare congenital ocular disorder characterized by an anteriorly displaced, prominent Schwalbe line and peripheral iridocorneal synechiae. The key implication of this anomaly is a 50% risk of glaucoma. The aim of our work is to report a case of Axenfeld's anomaly with ocular hypertension revealed by strabismus.*
Materials and Methods*: An eight-year-old child was referred to our consultation for strabismus. The slit lamp examination noted a gray-white circular line on the posterior surface of the cornea near the limbus with a wide anterior iridocorneal synechiae in the temporal area of the right eye. Measurements of intraocular pressure were 18 mmHg in the right eye and 19 mmHg in the left eye. Explorations showed no optic disc or extra ocular anomalies. The diagnosis of Axenfeld's anomaly with ocular hypertension was retained.*
Conclusion: *Axenfeld's anomaly is a rare congenital ocular defect that can be associated with a range of extraocular malformations. Glaucoma represents the main complication of this disorder. Diagnostic must be early in order to prevent blindness at young age.*
Key Word*: Axenfeld's anomaly; Glaucoma; Axenfeld Rieger syndrome*

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I. Introduction

Described for the first time in 1920, Axenfeld's anomaly is a rare congenital ocular disorder characterized by a prominent and anteriorly displaced Schwalbe line called posterior embryotoxon and peripheral iridocorneal synechiae [1,2]. Axenfeld's anomaly can occur isolated or part of a syndrome. It can be associated with extraocular malformations, especially of the teeth, facial bones, and periumbilical skin and is called Axenfeld-Rieger syndrome [1]. The key implication of this anomaly is a 50% risk of glaucoma [3]. A medical treatment is usually required to control the intra-ocular pressure but a surgery is often necessary in these cases. We report a case of Axenfeld's anomaly with ocular hypertension revealed by strabismus.

II. Observation

An eight-year-old girl was referred to our consultation with a strabismus recently noticed by the parents. On examination, she had right esotropia. Best corrected visual acuities were 6/10 in the right eye and 9/10 in the left eye. Slit lamp revealed a gray-white circular line on the posterior surface of the cornea near the limbus (posterior embryotoxon) and a mild corectopia with a wide anterior iridocorneal synechiae in the temporal area of the right eye. (Figure 1A,2B)

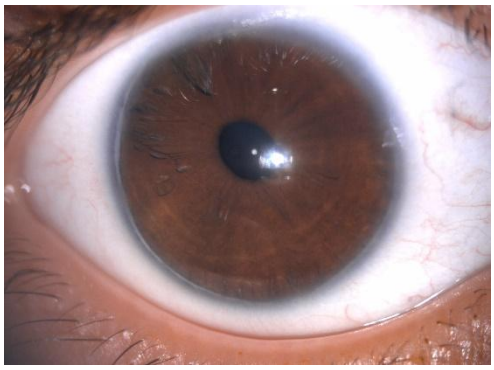

Figure 1A: Posterior embryotoxon and corectopia

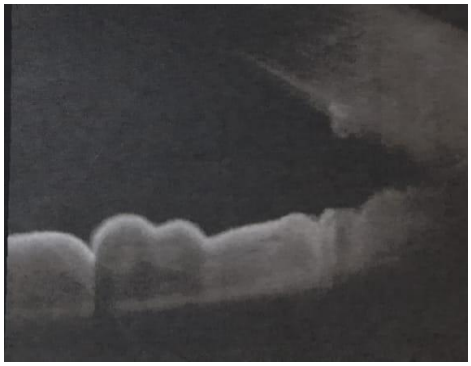

Figure 1B : OCT scan of posterior embryotoxon

Gonioscopy showed the presence of several peripheral anterior synechiae attached in the anterior chamber angle extending over more than 180° (Figure 2)

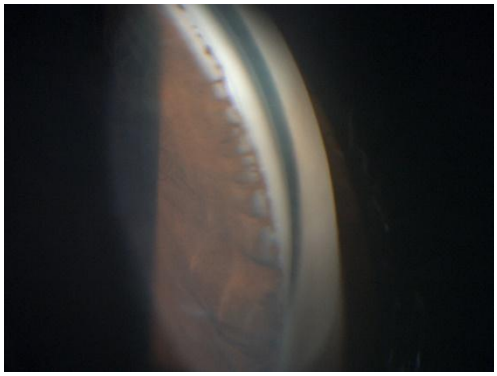

Figure 2 : peripheral anterior synechiae

The intraocular pressure (IOP) was 18 mmHg in the right eye and 19 mmHg in the left eye (Central corneal thickness was 542 μmin right and 536 μm on left). Examination of the optic disc was normal in both eyes with no sign of glaucomatous disease (Figure 3).

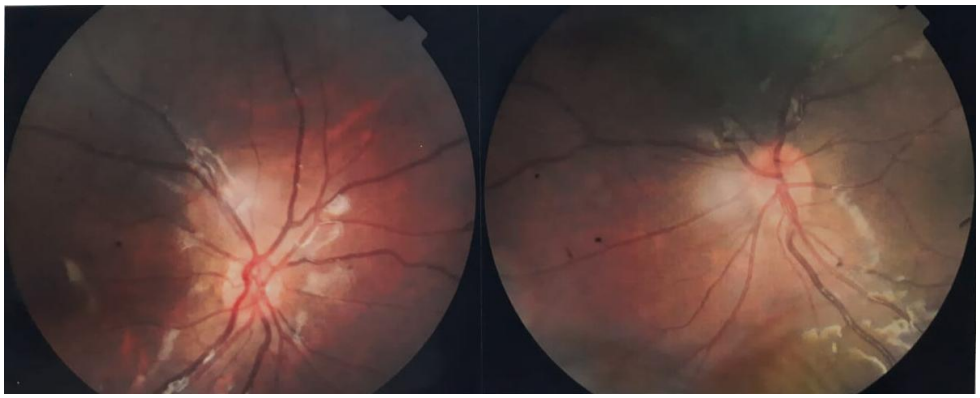

Figure 3 : Optic discs photography

Optic disc OCT showed a normal RNFL thickness in both eyes (126 µm on left and 129 µm on right) and analysis of GCL showed no abnormalities. Visual field did not allow conclusion after two attempts. Cardiac and abdominal ultra sound performed in search for extraocular malformations were normal as well as the oral examination that showed no teeth abnormalities.

Initial medical care consisted on treatment of amblyopia with occlusion of the left eye and topical preservative free latanoprost 0,005%, in both eyes. After on month follow-up, IOP measurement were 11mmHg in the left eye and 12mmHg in the right eye. Best visual corrected acuities were 8/10 right and 9/10 in the left eye. The patient has been on regular follow-up for more than 1 year now. His intraocular pressure is being maintained well within normal levels

III. Discussion

The Axenfeld's anomaly is characterized by an anteriorly displaced, prominent Schwalbe line (posterior embryotoxon), usually with attached iris strands in combination with varying degrees of iris malformation. It can be part of a large spectrum of disorders. When there are additional iris changes such as corectopia, iris atrophy, hypoplasia, iris holes, polycoria, and ectropion uveae, it is called as Rieger's anomaly. The Presence of non-ocular developmental defects defines Axenfeld-Rieger syndrome (ARS). ARS commonly regroups ocular, dental, facial, and abdominal abnormalities. Ocular findings include anterior iris stromal hypoplasia, anterior chamber synechiae, corneal opacity, microcornea, and glaucoma. Extra ocular features may include hypodontia, maxillary hypoplasia, periumbilical abnormalities and cardiac malformations.[1]

Approximately 50% of patients with Axenfeld's anomaly will develop secondary glaucoma, due to incomplete development of the trabecular meshwork and Schlemm'scanal. It usually develops during childhood or early adulthood [4]. Glaucoma is initially managed with medication, but surgery is often required on the long term. First line medical therapy is usually prostaglandin analogues and beta blockers. Surgery consists on goniotomy, trabeculotomy with mitomycin C, glaucoma drainage devices or cyclodestructive procedures for advanced cases [5]. In our case, we adopted preservative free latanoprost considering its protective effect on the ocular surface and the trabecular meshwork.

Considering that such patients are at an increased risk of developing glaucoma in adolescence or early adulthood, they need regular follow-up for early detection of glaucoma. The ocular hypertension secondary to Axenfeld's anomaly in our patient was diagnosed early and treated with medication before glaucomatous optic nerve head changes appeared. A regular monitoring with appropriate timely interventions will prevent irreversible blindness due to glaucoma.

IV. Conclusion

Axenfeld's anomaly is a rare congenital ocular defect that can be associated with a range of extraocular malformations. Glaucoma represents the main complication of this disorder. Diagnostic must be early in order to prevent blindness at young age.

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