CLINICAL SNAPSHOT

Headache and Eye Pain in a Patient With Unusual Facial Morphology

A 21-year-old man presented to the ophthalmological emergency room complaining of left eye pain and headache of two days' duration that had not responded to oral analgesics (acetaminophen 250 mg bid). Eye examination revealed bilateral dysgenesis of the anterior ocular segment due to Axenfeld-Rieger syndrome (ARS). This is an autosomal dominant disorder with a prevalence of one in 200 000, high penetrance, and variable ocular and systemic expression of malformations due to disturbed differentiation of neuronal stem cells. This patient's symptoms were due to elevated intraocular pressure in the left eye. The more common ocular manifestations of ARS are posterior embryotoxon, iridial atrophy, corectopia, and ectropium uveae; 50% of patients develop refractory glaucoma. Eye pressure compensation could be achieved by the topical application of a combined beta blocker and carbonic anhydrase inhibitor, but adequate lowering often requires extensive glaucoma surgery.

Figure: Anterior ocular segment in advanced Axenfeld-Rieger syndrome.

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