CONGENITAL ANIRIDIA WITH ASSOCIATED ABNORMALITIES - A CASE REPORT

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Abstract

Aniridia is a condition where iris is present in an extremely rudimentary state remaining hidden behind the corneo-scleral margin and thus making it invisible except on gonioscopy. A number of associated anomalies have been described with this condition. Here we report a case of young female with aniridia who had no associated skeletal, systemic or mental anomaly. On ophthalmoscopic examination the fundi were normal. Glaucoma, degenerative changes in cornea and subluxated cataractous lenses were present in both eyes. No influence of heridity could be traced. Bilateral partial ptosis had been a feature in the present case which is lacking in literatures on aniridia.

Key words : Clinical aniridia, Associated anomalies, Inheritance, Fundus findings, Partial ptosis.

Introduction

Aniridia, a condition where iris is extremely rudimentary is relatively rare, its occurance was estimated to be about 1 in 1,00,000 of population (6). The term aniridia is correct only in clinical sense as rudimentary iris is always present. Its apparent absence on clinical examination is due to the fact that the short stump is hidden behind the corneoscleral margin and is invisible except on gonioscopy. The anomaly is nearly always bilateral, but unilateral cases have also been described. Evidences refer to the primary fault of the failure in development of retinal ectoderm and an aberrant development of vascular mesoderm (2).

With aniridia, additional ocular deformities are extremely common. The cornea may be small, conical shaped are frequently showing opacities and degeneration. Blue sclerae have been noted. Glaucoma occurs in childhood in most aniridic patients (8). The periphery of the hypoplastic iris observed by gonioscopy is pulled over the trabecular meshwork and the iris is often bound to the cornea with mesodermal tissue so that the angle of anterior chamber is obliterated and it leads to glaucoma (4). The lens may be ectopic, colobomatous and in almost every case cataractous. Aplasia or hypoplasia of the ciliary

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