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Prognostic importance of congenital cataract morphology: A case report

Konjenital katarakt morfolojisinin prognostik önemi: Olgu sunumu

Çağrı İlhan [1]

[1] Hatay State Hospital, Antakya, Hatay, Turkey

Abstract

Congenital cataract (CC) has an important place in pediatric ocular diseases. CCs are different from senile nuclear cataracts in terms of their etiologic, clinic and morphological characteristics. CCs occur many different forms such as non-hereditary isolated cases or autosomal dominant bilateral cases. In addition, many of ocular and systemic diseases can be associated with CC and ophthalmologist should be aware of these potential risks. In this article, we queried whether the different morphological features of CC have prognostic importance by considering a case of CC.

Keywords: Congenital cataract, Cataract morphology, Polar cataract, Sutural cataract

Öz

Pediatrik göz hastalıkları içinde konjenital kataraktlar önemli bir yere sahiptir. Etiolojik, klinik ve morfolojik özellikleri göz önünde bulundurulduğunda konjenital kataraktlar erişkin nükleer kataraktlardan farklıdır. Konjenital kataraktlar ailesel geçiş göstermeyen izole vakalar şeklinde görülebileceği gibi otozomal dominant bilateral vakalar şeklinde de görülebilir. İlaveten konjenital kataraktlar, bir çok oküler ve sistemik hastalıkla ilişkili olabilir ve göz hekimleri bu potansiyel birlikteliklerin farkında olmalıdır. Biz bu yazımızda, bir konjenital katarakt olgusu üzerinden, farklı konjenital katarakt morfolojilerinin prognostik önemi olup olmadığını sorguladık.

Anahtar kelimeler: Konjenital katarakt, Katarakt morfolojisi, Polar katarakt, Sutural katarakt

Introduction

Cataract means the opacification of human natural crystalline lens. It is known as a geriatric disease because incidence of cataract increases with aging. Nevertheless, cataract can be seen in pediatric population. If cataract exists in birth, it is called as congenital cataract (CC). If cataract does not exist in birth and it occurs before 16-year-old, it is called as juvenile cataract [1]. Some books mentioned from infantile cataract which, means formation of cataract occurred in first year of life [2]. CC is responsible from 15-20 percent of pediatric blindness [3]. CCs are different from senile cataracts in terms of their etiologic, clinic and morphologic characteristics. Always, there is no visual impairment in disorder's clinic. In this article, we queried whether the different morphological features of CC have prognostic importance by considering a case of CC.

Case presentation

A 14-year-old female patient presented to our clinic for routinely ocular examination. Best corrected visual acuity (BCVA) and intraocular pressure for both of eyes were normal. In biomicroscopic examination, sutural cataract and blue dot opacities were seen in both of eyes (Figure 1). Retina, macula and optic disk were normal. When queried her medical history, she did not report having any systemic and ocular chronic disease, using any cataract-related drug or experiencing any trauma. Similar cataract morphology that did not impair vision, was also found in 46-year-old mother of patient (Figure 2). Anterior segment photographs were taken and written approval was obtained from her mother for using of photographs in academic purposes. Ocular examinations of other family members were completely normal.

Corresponding author / Sorumlu yazar:
Çağrı İlhan
Adress: Hatay Devlet Hastanesi, Antakya, Hatay, Türkiye
e-Mail: cagriilhan@yahoo.com

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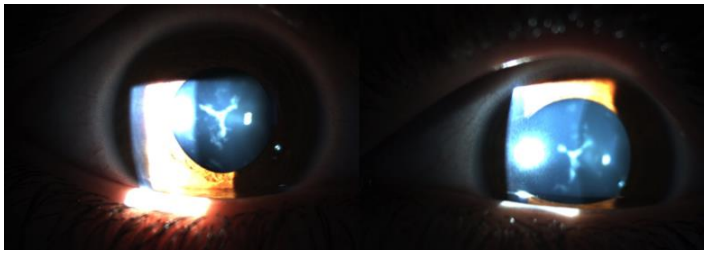

Figure 1: Fourteen-year-old patient's anterior segment photo

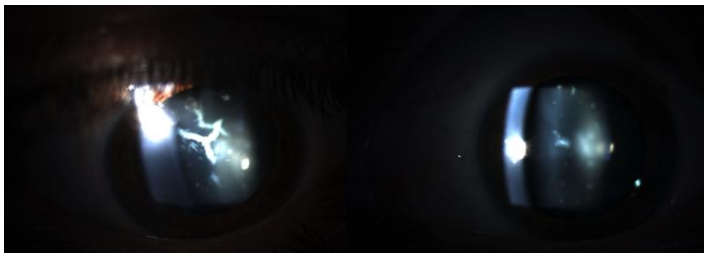

Figure 2: Forty-six-year-old mother's anterior segment photo

After the examinations of patient and her mother, findings were considered as isolated autosomal dominant congenital sutural cataract. We did not plan any surgery for clinically non-significant cataract and we recommended annual ocular examination for both of patients.

Discussion

Sixty-percent of pediatric cataracts are idiopathic while 10-25 percent of them are associated with genetic disorders [4]. CC is the most important etiologic factor in pediatric cataracts and it is seen in 40-60 of 100000 living birth [1, 5]. In generally, unilateral CCs are idiopathic and they are not associated with any systemic or genetic disorders. However, the majority of bilateral CCs result from several genetic conditions. These genetic conditions can be trisomy (13, 18, 21), deletion (5p, 18p, 18q) and disorders autosomal recessive inherited. In addition, a study reported that CC was inherited as autosomal dominant in 30 of 39 families with bilateral CC [6].

Defects in genes effecting crystalline lens development cause CC. These defective genes encode some proteins which play a role in enzyme synthesis (like galactosemia) or signal transmission or are used as structural substance (like crystalline) or transmembrane proteins (like aquaporin and connexin) [7].

Some multisystem diseases can cause CC. These diseases can affect primarily kidneys (Lowe syndrome, Alport syndrome, Hallerman-Streiff-Francois syndrome), skeletal system (Stickler syndrome, Smith-Lemli-Opitz syndrome), central nervous system (Marinesco-Sjogren syndrome, Zellweger syndrome), muscular system (myotonic dystrophia) or skin (Cockayne syndrome, incontinentia pigmenti, ichtiosis).

Intrauterine trauma, radiation or TORCH infections (the acronym consisted of Toxoplasmosis, Other, Rubella, Cytomegalovirus, Herpes infections) can be reason of CC. Microphthalmia, aniridia and retinal abnormalities commonly accompany these cataract formations.

Morphology of CC effects visual prognosis, like duration and reason of cataract. CCs are very different from senile nuclear cataracts in terms of morphology of them. They can be classified based on the place of opacity in the lens. Nuclear, lamellar, cortical, sutural, pulverulent, cerulean and colariform cataracts locate in central of the lens. Polar cataracts locate in anterior (anterior polar, anterior pyramidal, anterior subcapsular) or posterior (posterior subcapsular, posterior lenticonus, posterior fetal vasculature) layers of the lens [2]. In addition, many of CC morphologies give hint about CC associated ocular and systemic abnormalities.

Congenital nuclear cataracts locate between Y sutures of embryonic or fetal nucleus and they are present in birth. This cataract has autosomal dominant inheritance and it generally impairs patient's vision [2]. Opacification of fetal nucleus' superficial lamellas called as congenital lamellar cataract. These opacifications can be different grades of severity and formation and they are commonly occurred as bilateral and asymmetric. Congenital lamellar cataracts less decrease vision when compared with other forms of CC [8]. Y shaped opacifications are occurred in congenital sutural cataracts. This cataract morphology does not progress and patient's vision does not decrease unless cortical and nuclear cataracts. Congenital sutural cataract can be inherited as autosomal dominant or it can be occurred idiopathic. Rarely, it can be seen in Nance-Horan syndrome and cranio-lenticulo-sutural dysplasia [9, 10]. Congenital pulverulent cataract has an appearance like thin cloud of dust. In this cataract morphology, opacities take place in embryonic nucleus. In general, congenital pulverulent cataract is non-progressive and it does not decrease vision [11]. Congenital cerulean cataract is seen as blue-white dots and these opacities take place in superficial layers of fetal or adult nucleus. In general, congenital cerulean cataract is bilateral and progressive and it does not decrease vision until to adulthood [12].

Dot like opacities in anterior lens surface are called as congenital anterior polar cataract. This non-progressive CC morphology can be unilateral or bilateral. It does not decrease vision unless it takes place in central of lens. Nevertheless, if it projects from anterior lens capsule to anterior chamber, it causes blurred vision and amblyopia. At this time, it is called as congenital anterior pyramidal cataract and it is more serious cataract morphology than anterior polar cataract [13]. One clinical study revealed that more than one of four of anterior lens opacifications causes amblyopia [14].

The severity of congenital posterior polar cataract morphologies increases from Mittendorf spot to persistent fetal vasculature. In these cataract formations, persistent fetal vasculature resulting from abnormal regression of primary vitreous can decrease vision seriously [15].

Complete opacification of nucleus and cortex of lens cause total cataract. Trauma, familial cases and metabolic diseases can be responsible for this cataract morphology.

In conclusion, all of CCs are not same and they have different etiologic, clinic and morphologic features. These differential features can help to predict course of disease and to diagnose of related ocular, systemic and genetic conditions. Thus, morphology of CC may be an important factor to tune of cataract surgery time for better visual results. In summary, the different morphological features of CC may have prognostic importance and effect decision of surgery.

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