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The visual system in infants with microcephaly related to presumed congenital Zika syndrome

Islane Verçosa, MD, Marcia Beatriz Tartarella, MD, PhD, Paula Carneiro, MD, Reno Verçosa, MD, Renata Girão, MD, Erlane M. Ribeiro, MD, PhD, Andre Pessoa, MD, Nadia G. Almeida, MD, PhD, Paloma Verçosa. MD

Introduction

The Zika virus (ZIKV) is a single-stranded RNA virus of the Flaviviridae family and genus flavivirus. It is transmitted by the mosquito Aedes Aegypti.

Zika infection during pregnancy can cause embryopathy and the newborns have severe central nervous system malformations, microcephaly, arthrogryposis and ophthalmic abnormalities. The vertical transmission to the fetus can occur at any period of gestation.

Objectives

To describe and analyze ocular features in infants with microcephaly due to presumed congenital Zika syndrome (CZS).

Methods

A clinical study was conducted at CAVIVER, a nongovernmental organization clinic and referral center dedicated to visually disabled children, at the city of Fortaleza, Brazil.

Ophthalmological evaluation included: assessment of visual acuity using Teller Acuity Cards (FIGURE 1), anterior segment evaluation, strabismus evaluation, and binocular indirect ophthalmoscopy performed with mydriasis (tropicamide 1%).

Figure 1

The patients with presumed CZS who were diagnosed with retinal changes were examined under anesthesia to measure intraocular pressure (Perkins handheld applanation tonometer) and to obtain wide-angle fundus digital images using the RetCam for documentation and detailed study and mapping of retinal lesions (FIGURE 2).

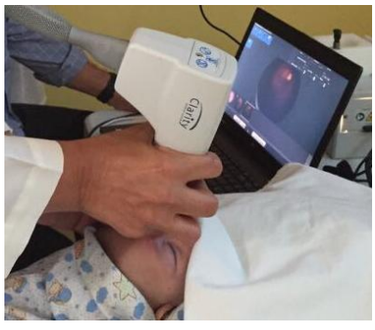

Figure 2

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Results

A total of 70 infants with microcephaly were included; of these, 25 (36%) had ocular abnormalities.

Anterior segment analysis was normal in all patients. None had cornea, iris, or lens abnormalities.

Of the 70 patients, 18 (26%) were diagnosed with intraocular abnormalities.

A total of 33 eyes were structurally affected. Macular chorioretinal atrophy was observed in 27 eyes of 15 infants (60% of the patients with ocular abnormalities), and wide-angle digital imaging was obtained.

Chorioretinal atrophic lesions were whitish, flat, circumscribed with linear borders, resembling choroidal coloboma (colobomatous-like lesions), unique or multiple, varying in size from about 1 to 10 disk diameters (FIGURE 2-3).

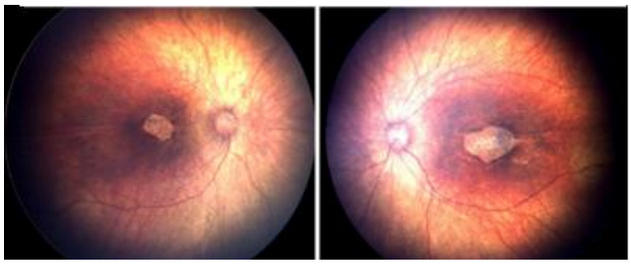

Figure 2

No retinal detachment, vitreous changes, or inflammatory signs or cells were observed in any affected eye. Mottled retinal pigment epithelium changes were observed in the posterior pole in 18 patients (72%). Optic nerve pallor was observed in 10 children (bilateral in 7 and unilateral in 3).

Strabismus was observed in 10 infants (14%): esotropia in 9 infants and exotropia in 1 infant (FIGURE 5) . Nystagmus was observed in 6 patients. Six cases of strabismus and 1 case of nystagmus were detected in children without intraocular abnormalities.

Visual acuity was obtained in 11 patients. All results of visual acuity were below normal range for age-matched children.

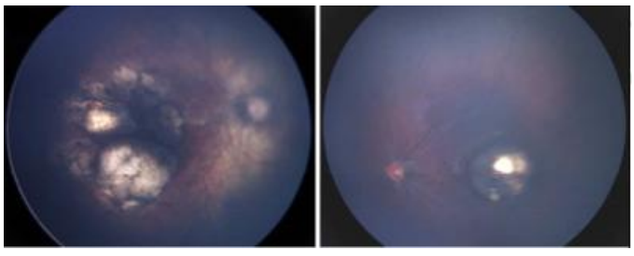

Figure 3

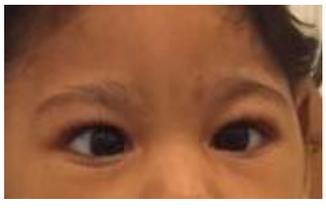

Figure 5

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Conclusions

The 5 major signs that differentiate CZS from other congenital infections are as follows: (1) severe microcephaly with craniofacial disproportion, narrow and laterally depressed frontal bone and occipital prominence with abnormal skull morphology; (2) central nervous system malformation and thin cerebral cortex with subcortical calcifications; (3) macular scarring with chorioretinal atrophy and mottled retinal pigment epithelial changes; (4) congenital contractures or arthrogryposis; and (5) early hypertonia and extrapyramidal involvement. All systemic and ocular characteristics of CZS do not necessarily occur in the same patient.

In our study, 25 of 70 infants (36%) with microcephaly due to presumed CZS showed ocular anomalies, including 18 patients with intraocular lesions (26%) and 7 (10%) with altered ocular motility (strabismus or nystagmus). The circumscribed chorioretinal atrophy and optic nerve abnormalities observed in our patients were similar to those reported previously in babies with CZS.

Optic nerve anomalies such as hypoplasia, atrophy, or pallor are a common ocular feature in CZS. Optic nerve pallor occurred in 14% of the cases in this study.

Strabismus occurred in 10 infants, at an early mean age of 4 months. Seven patients (10%) without intraocular abnormalities had strabismus (6 cases) or nystagmus (1 case). This rate of strabismus may increase, as patients get older.

It is not always possible to test visual acuity in infants with CZS because of neurological problems.

Visual acuity was obtained in 11 patients in this study. All infants tested showed functional vision deficits, with visual acuity below normal for age-matched children.

Ophthalmological screening guidelines and follow-up of the children with CZS is mandatory.

Children with CZS may have visual impairment and need early visual intervention and visual rehabilitation programs.

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