Congenital ocular toxoplasmosis with torpedo maculopathy and retinopathy of prematurity in a premature baby

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1. Introduction

Retinopathy of prematurity (ROP) is a vasoproliferative, multifactorial disorder of the retina that occurs principally in newborn preterm infants and is strongly related to low gestational age, low birth weight, supplemental oxygen therapy, fluctuations in oxygenation and poor postnatal growth.1,2

Congenital Toxoplasma infection occurs via vertical transmission from the mother and may cause severe congenital malformation, fetal developmental retardation, and the infection of neural tissues. The best described clinical presentation of ocular toxoplasmosis is focal necrotizing chorioretinitis, ultimately resulting in characteristic atrophic scars.3 Torpedo maculopathy (TM) is a benign, congenital lesion of the retinal pigment epithelium (RPE) located usually temporal to the fovea with orientation towards the central macula. It is usually unilateral, asymptomatic and can affect the outer retina and the choroid.4,5 Association with other ocular conditions include numerous conditions including infections such as congenital Zika syndrome.6-9

Herein we report an unusual case of congenital torpedo maculopathy in an eye with retinopathy of prematurity (ROP) and a non-specific pigmentary retinopathy in the fellow microphthahmic eye with congenital cataract in a premature baby with positive Toxoplasma serology.

2. Case report

An extremely premature baby boy, born at 26 weeks of gestation with birth weight of 800 g, was referred to the Medical Center for repair of intestinal perforation at 31 weeks of gestation. He already had intraventricular hemorrhage and necrotizing colitis diagnosed at the referring hospital. On admission, the baby had hypotension and features of septic shock. He underwent laparotomy for repair of stomach perforation as well as perforation along the greater curvature of stomach at the site of the feeding tube.

The first ophthalmic examination was done at 32 weeks, which revealed dense tunica vasculosa lentis, dilated iris vessels and dull view of fundus showing zone 1 vascularization, stage 1 with no Plus or pre-plus disease in the right eye (OD). The left eye (OS) had less pronounced tunica vasculosa lentis, normal iris vessels and same retinal findings in addition to perimacular lesion. Close surveillance was recommended.

Subsequent screening after 1 week showed very dull fundus views due to worsening of anterior segment findings in OD. There were central lenticular changes, iris vessels encroaching on lens and poor pupillary dilation due anterior synechiae with anterior segment inflammation. Fig. 1. Limited funduscopy view showed vascularization in zone 2 stage II with no Plus or pre-plus disease and non-specific peripheral
pigmentary retinopathy OD. There were residual tunica vasculosa and stage 2, zone II with no Plus or pre-plus disease but peri-macular lesion in OS. The lesion did not show any activity such as bleeding or fluid exudation and resembled torpedo shape. Pupillary dilation could not be performed due to unstable systemic condition with respiratory distress syndrome of newborn and pulmonary interstitial emphysema. The baby was evaluated extensively including the TORCH profile, but none of the tests was positive. Prednisolone acetate 1% eyedrops was started in OD four times daily. Follow-up examination after 2 days showed further deterioration, pupillary occlusion, central cataract, shallow anterior chamber and corneal edema in OD. B-scan ultrasonography demonstrated microphthalmia (axial length was 14.25 mm and 16.54 mm in OD and OS, respectively), and flat retina. On subsequent examination within a week, OS showed pre-plus disease and straightening of vessels at the periphery, stage 3 in posterior Zone II. Avascular area in the periphery was fluffy and raised with "popcorns" forming. Darkening of macular scar was also seen in OS Fig. 2. Subsequently, panretinal photocoagulation was performed in OS, while periorbital triamcinolone acetonide was administered in OD in an effort to decrease the ocular inflammation.

Consequently, left eye responded well to laser and ROP resolved. No further treatment was required. Macular scar was stable with central darkening representing retinal pigment epithelium (RPE) and peripheral oval RPE atrophy (Fig. 3). Portable OCT was not available for imaging. Inflammation in OD also subsided after the periorbital injection of steroid. B-scan showed no tractional retinal detachment and elective cataract extraction was scheduled at 41 weeks of gestational age. Lensectomy with anterior vitrectomy due to partially calcified posterior capsule were done in OD uneventfully before the baby was discharged from the neonatal intensive care unit. Post-surgical examination revealed healthy disc with peripheral pigmentary retinopathy and RPE atrophy at its margin but no inflammation or hemorrhage in OD (Fig. 4) Outpatient cyclorefraction was performed and aphakic contact lens was given.

In the post-operative period, while out of the country, the baby was sent for additional laboratory tests including Toxoplasma antibody test which revealed increased both IgG 239 IU/ml (normal range <3.0) and IgM 5.47 (normal range <0.54) confirming the diagnosis of congenital ocular toxoplasmosis in addition to ROP. Systemic antibiotic treatment was immediately commenced.

3. Discussion

The index case describes coexistence of ROP and congenital ocular toxoplasmosis which was detected upon subsequent serum antibody testing following negative TORCH screening. To the best of our knowledge only two, very recent, publications report on coexistence of ROP and congenital ocular Toxoplasmosis. Hasanreisoglu et al. describe two premature babies with stage 2 ROP in one eye of each baby and both intracranial and ocular toxoplasmosis with bilateral traction retinal detachments in both babies in addition to mild vitritis. The cases were successfully treated with anti-toxoplasma treatment, vitrectomy surgery, endolaser and internal tamponade. In another report, Astasheva et al. describe a child with signs of toxoplasmosis in the period of regression of stage 2 ROP at 48 weeks of postmenstrual age with rapid progression to retinal detachment. Our case did not demonstrate any signs of intracranial involvement and no retinal detachment. Inflammation was limited to anterior segment. As demonstrated above, coexistence of these two conditions is extremely rare and the reason is unclear. It is well known, however, that serious congenital infections
such as chorioamnionitis, placental infections, and sepsis increase the risk for ROP in susceptible premature infants. ROP in the left eye was controlled with retinal laser photocoagulation and the TM lesion did not expand following the laser.

While TM is usually described as a hypopigmented lesion, the index case shows slightly unusual central pigmentation with surrounding hypopigmentation. The central pigmentation increased during observation and following peripheral retinal laser photocoagulation. In light of clinical presentation and laboratory confirmation it is reasonable to believe that TM is related to infectious retinochoroiditis. This report expands clinical spectrum of retinal findings in eyes with coexisting ROP and ocular toxoplasmosis.

4. Conclusions

In summary, we describe a prematurely born baby with bilateral ROP (one eye treated) and hyperpigmented torpedo maculopathy in one eye and congenital cataract with pigmentary retinopathy in the fellow eye as a result of serologically confirmed congenital toxoplasmosis. Subsequent tests for detection of toxoplasma are needed despite negative TORCH screening.

Patient consent
Moorfields Eye Hospital UAE Research and Ethic Committee waived the need for IRB review for this Case report. Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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