

Ocular toxoplasmosis with surprisingly good retinal function

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Abstract

Ocular toxoplasmosis is an infection in the eye caused by the parasite *Toxoplasma Gondii*. A common retinal finding in its inactive stages is pigmented retinochoroidal scarring. The retinal function in the affected area assumingly reflects the amount of retinal involvement. In this manuscript, we report the case of a 48-year-old woman who has a long-standing large retinochoroidal scar in the temporal posterior pole of her left eye. She had not experienced any visual symptoms, and no recurrent infections had occurred as far as she knew. She had a scotoma in her nasal visual field that her optometrist detected by a coincidence when she was in her twenties. The corresponding visual field defect is smaller and less deep than what may be expected from the structural appearance of the scar. The reported case demonstrates that the visual function may be preserved in the visual field corresponding to a retinochoroidal scarred area due to toxoplasmosis, in spite of loss of structures in the outer retinal layers as seen with optical coherence tomography (OCT).

Sammendrag

Okulær toxoplasmose er en infeksjon i øyet som er forårsaket av parasitten *Toxoplasma Gondii*. Et vanlig retinalt funn i inaktivt stadium er pigmentert retinokoroidal arrdannelse. Retinal funksjon i det affiserte området reflekterer omfanget av retinal involvering. Denne kasusrapporten omhandler en 48 år gammel kvinne som i mange år har hatt et stort retinokoroidalt arr lokalisert inferiotemporalt i venstre øyets bakre pol. Hun hadde ikke opplevd noen visuelle symptomer, og heller ikke hatt gjentatte infeksjoner så vidt hun husket. Hun hadde et skotom i nasalt synsfelt som ble oppdaget ved en tilfældighet av hennes optiker da hun var i 20-årene. Synsfeltdefekten som korresponderer med beliggenhet av arret, er ikke så dyp som man kunne forvente ut fra det strukturelle utseendet av arret. Dette demonstrerer at det fortsatt kan være synsfunksjon til stede i et område med retinokoroidalt arr, til tross for tap av de ytre retinale lagene som vist i dette tilfellet målt med optisk koherens tomografi (OCT).

Introduction

Ocular toxoplasmosis (OT) is caused by the protozoan parasite *Toxoplasma Gondii*. The parasite is one of the world's most common reasons for retinochoroiditis and affects primarily children and young adults (Commodaro et al., 2009). The incidence of active symptomatic OT in the general population varies worldwide, but was reported to be 0.8 per 100 000 per year in United Kingdom (Gilbert et al., 1999), and is higher in warmer climates (Furtado, Winthrop, Butler, & Smith, 2013).

The macular area or the posterior pole are often the primary location for the parasites, where they are trapped inside the retinal capillaries (Padhi et al., 2017; Sutton & Torbit, 2001). The optic nerve head may also be involved, but this is less common (Padhi et al., 2017; Park & Nam, 2013). Since the retina is the

primary location for the parasite, choroidal lesions do not occur without a retinal infection (Commodaro et al., 2009; Padhi et al., 2017). Post infection, there is necrosis of the retina in the affected area which may form a retinochoroidal scar. In inactive OT, the retinochoroidal scars are distinctive with a focal necrotized retina surrounded by retinal pigments. Depending on the location and extent of the damage, the visual function may be impaired (Commodaro et al., 2009).

With optical coherence tomography (OCT) it is possible to visualize the different layers of the retina. The scar formation is reported to have varying degree of involvement and varying degree of thinning of the retinal layers (Goldenberg, Goldstein, Loewenstein, & Habet-Wilner, 2013). There are many studies that document active stages of OT, and also some case reports that present findings with OCT in inactive OT, but there is a lack of studies on the structure- and function relationship in retinas in inactive stage of OT.

Case report

The patient was a 48-year-old female wearing progressive power spectacles lenses. The patient did not report ocular symptoms like flashes or floaters, headache, blurred vision, diplopia or photophobia. Neither had she experienced episodes of visual loss. Her health was good and with no known systemic diseases.

The patient had a known visual field (VF) defect due to an OT retinochoroidal scar in her left temporal retina. An optometrist detected this during an eye examination when she was in her twenties. She had no memories of ocular infections or experiences of vision loss. Serology tests during her pregnancies were positive for toxoplasmosis.

Examinations

The patient participated in a research project at the University of South-Eastern Norway. She underwent a standardized optometric examination. This included refraction, visual acuity (Bailey-Lovie), slit lamp examination, dilated fundus examination, and retinal imaging (Topcon TRC-NW6S, Topcon, and Optomap P200C, Optos Inc.). In addition, OCT-scan was performed with the Heidelberg Spectralis SD-OCT (Heidelberg Engineering GmbH, Germany). The VF in her left eye was tested with standard automated perimetry (SAP), Central 30-2 and Peripheral 60-4 (SITA-strategy), obtained with the Humphrey VFA (Carl Zeiss Meditec AG, Jena, Germany) using an appropriate near correction. In order to plot the extent of the scotoma with greater spatial resolution, a customized VF test was used (Humphrey VFA). This consisted of supra-threshold stimuli using threshold-related three-zone strategy (6 dB and 0 dB) with 6 degree spacing in a grid performed at 9 different locations to cover a total test area of 38 × 20 degrees. There were no signs of OT in her right eye, thus this eye's results will not be presented or discussed.

Optometric examination

The refraction was +1.75/ – 0.25 × 20 (OS) for distance with addition +1.75D for near. The visual acuity was –0.1 (logMAR) at both distance and near. Pupillary reflexes were normal. The anterior chamber angles were open, and her intraocular pressure was 17 mmHg OU (ICare TA01i, ICare, Finland). The anterior segment and the cornea appeared healthy. There were no signs of uveitis or corneal precipitates. The crystalline lens was clear and considered normal.

Fundus examination

A retinochoroidal scar was visible four disc diameters (DD) from the disc (1.5 DD temporally to the fovea) and below the

horizontal midline (see Figure 1). The size of the scar was approximately four DD in width and two DD in height and had well demarked borders. It was seen as a whitish lesion with visible choroidal blood vessels and with dense irregular pigmentation in the temporal part of the scar. No other scars or active lesions were found in this eye, nor in the contralateral eye. Parapapillary atrophy was found at the temporal disc edge (3-4 o'clock), with a corresponding localized nerve fibre layer (NFL) defect (see Figure 2). However, there was no notching of the papillary rim in the corresponding segment of the optic nerve head.

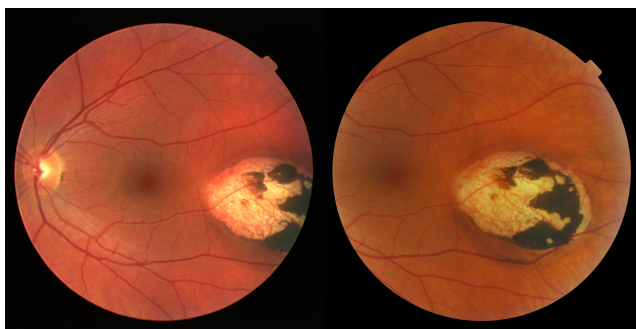


Figure 1: These images captured with a 45-degree view (Topcon, TRC-NW6S, Topcon Corporation, Japan), show the posterior pole including the optic nerve head and the extent of the retinochoroidal scar in the temporal posterior pole in the left eye.

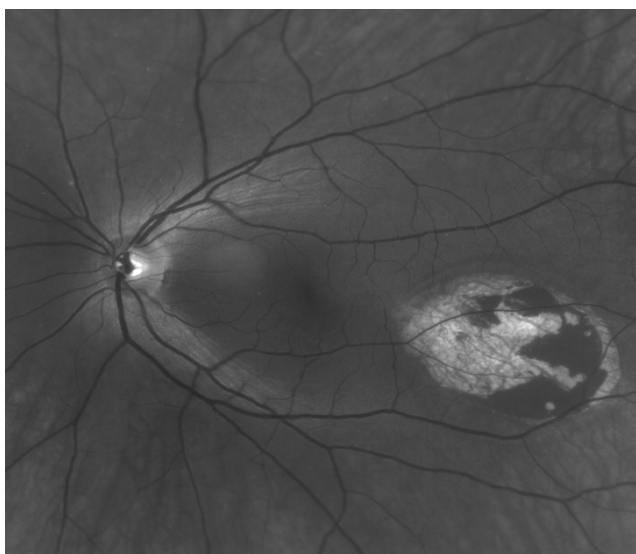


Figure 2: This figure shows the green separation view of an image captured with Optomap (P200C, Optos Inc.). The image shows the retinochoroidal scar located inferior temporal to the fovea in the left eye. The area inferior temporal to the disk (initiating 3-4 o'clock at the optic nerve head) is slightly darker than elsewhere, indicating a localized NFL defect.

A volume OCT-scan of the area with the scar (49 sections, 20° × 15°) was performed to evaluate the retinal layers (see Figure 3). The foveal pit was normal. There was an obvious split between Bruch's membrane and the external limiting membrane (ELM), and the photoreceptor layers were missing. The choroid and the underlying structures appeared to be bulging outwards, most pronounced centrally in the scar, resembling a staphyloma. The retinal pigment epithelium (RPE) was missing, but there were patches of higher reflectivity temporally in the area with the scar. In the OCT-image this is causing a shadowing effect of the underlying tissue. In the area with the scar, the lumen of the large vessels of the choroid lie closer to the RPE than elsewhere, which indicates that the choriocapillary layer is thinner in this area.

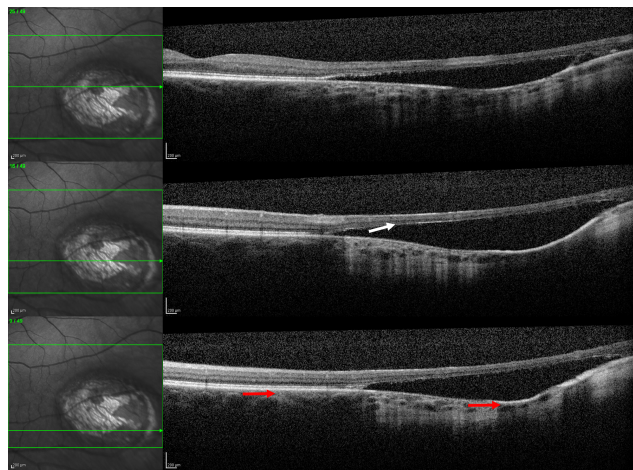


Figure 3: This figure shows an example of three OCT-sections through the scar. The left retinal image is an IR-image from the Spectralis OCT. The green square shows the scanned area of the retina. The central horizontal green line in each of the images on the left is the location for the OCT-section shown in each of the images on the right. The OCT-sections show an obvious split between the inner retinal layers and a bulging of the underlying Bruch's membrane, choroid and sclera. The edge of the scar was defined as the beginning of the visible split. The white arrow in the middle section points to the ELM. The red arrows in the lowest section point to the distance between the lumen of the large choroidal vessels and the RPE within and outside the retinochoroidal scar.

Examination of the visual field

The patient was considered to be an experienced observer. Results from the Peripheral 60-2 test showed that the peripheral VF was normal. Results for the Central 30-2 test showed a moderate to deep paracentral scotoma in the superior nasal field (see Figure 4).

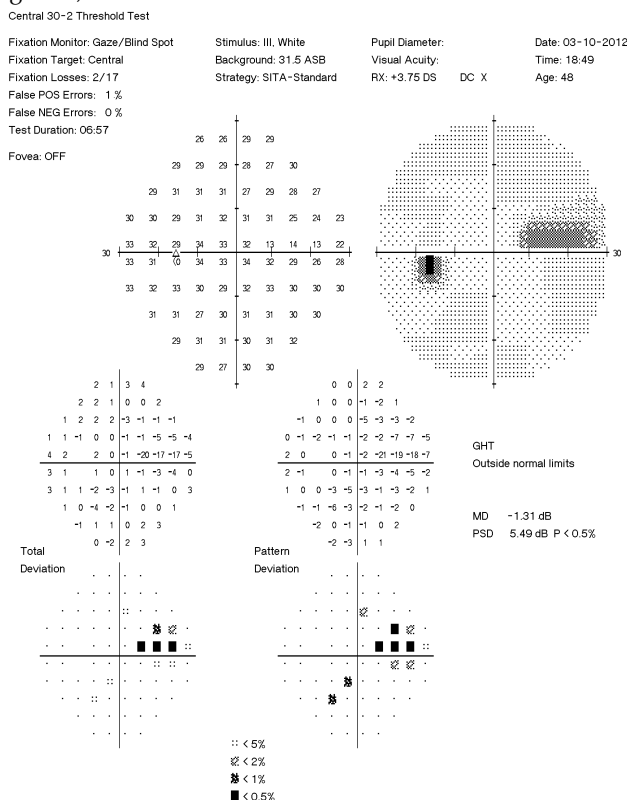


Figure 4: This figure shows the results from the VF test Central 30-2, which is a threshold test with SITA- standard strategy, tested with the Humphrey VF Analyzer.

Figure 5 displays test results from the custom-made test. The horizontal and vertical extent of the found relative defect was 14 degrees and 12 degrees, respectively. Only one test point below the nasal horizontal midline showed a defect. All test locations were seen when tested with maximum intensity (0dB).

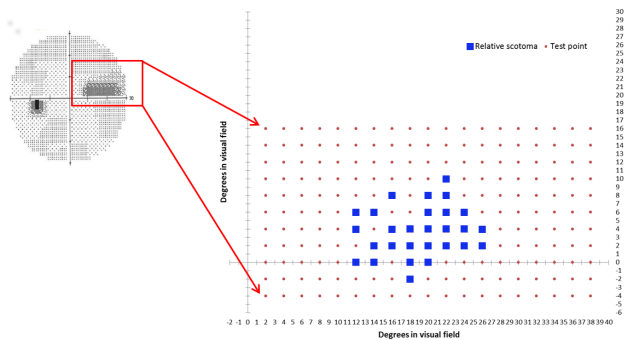


Figure 5: The left figure indicates the SITA standard 30-2 test plot of the subject's VF in her left eye. The scotoma due to the retinochoroidal scar is visible in the superior nasal field, and the red square indicates the area tested further with the custom-made tests (threshold related three-zone strategy, 209 locations spaced 2° x 2°). The graph to the right shows the results from the custom-made tests where the red dots are all locations tested which were seen, and the blue squares indicate location of relative scotoma. The x- and y-axis are degrees in the VF. None of the locations showed an absolute scotoma.

Figure 6 visualizes the spatial relationship between the fundus appearance and the visual function tested with SAP and the customized test. The location and the extent of the visual defect is in concordance with the extent of the visible scar defined by OCT for 6 out of 8 defect locations from the 30-2 VF test. The two locations that are immediately above the scar, are the same that do not respect the horizontal midline in the VF result. All relative defects (blue squares) found with the custom-made test were within the area of the scar.

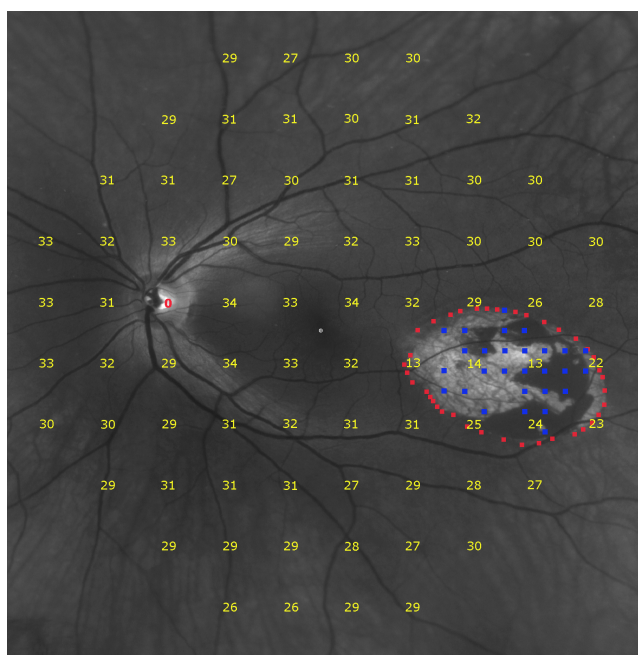


Figure 6: This figure shows the SITA 30-2 VF with dB-values (yellow numbers) and the relative scotoma from the custom-made test (blue squares) superimposed on the Optomap image to visualize the visual function inverted to compare with the structure. The test point distance is scaled to the image by using the approximate distance between fovea and the optic disc, and between the blind spot and fixation point. The red squares indicate the border of the retinochoroidal scar which has been defined by the splitting retina detected by OCT.

Discussion

The retinochoroidal scar that was found in this patient is located close to the macular area, which is typical for OT (Padhi et al., 2017). The appearance of a scar with white sclera, heavily pigmented border and a clear overlying vitreous indicates an inactive and longstanding OT (Commodaro et al., 2009). Since the patient had no memory of infections or vision loss, it is likely that the infection has occurred early in life.

OCT has become a useful and important tool in diagnosing ocular conditions. Goldenberg et al. (2013) reported four different types of retinochoroidal scars (atrophic, elevated, deep and combined scars) in inactive OT, when identified with OCT, based on the appearance of the neurosensory retina, RPE and the choroid. The retinochoroidal scar presented in this case report matches the description of both an atrophic and deep scar with atrophy of the RPE and the ellipsoid zone (inner- and outer segment junction of the photoreceptor) and thinning of the choroid according to Goldenberg's classification. However, it is not in conformity with the missing of the ELM, which is visible in the reported case, and a deformed profile of the inner retina, which is preserved for this patient. The inner retinal layers appear to be thinner but intact, with ELM as the most posterior structure seen, and are bridging the atrophic area making it look like a staphyloma (see Figure 3). In order to investigate staphylomas and to measure choroidal thickness with OCT-imaging it is necessary to use equipment that was unavailable to us.

Studies have shown that damage or disruption of photoreceptor layers associated with reduced retinal sensitivity (Hildebrand & Fielder, 2011; Rensch & Jonas, 2008) can be visualized with OCT as loss of integrity of the ELM, ellipsoid zone and interdigitation zone (Cassels, Wild, Margrain, Chong, & Acton, 2018). Therefore, the lack of the ellipsoid zone and interdigitation zone in the outer retinal layers of the reported case indicates an area where visual input is expected to be lost or severely affected. Because remnants of the outer retina were visible on the OCT-scan close to the border of the retinochoroidal scar, the extent of the assumed non-functional area was expected to be smaller than the extent of the visible scar (see Figure 6). Surprisingly, findings from SAP and the custom-made VF test, demonstrated no absolute VF defects. Instead, results from both tests indicated a residual visual function for test points within the scarred area. Test points in the close vicinity to this area were within normal range of sensitivity when tested with SAP (see Figure 4). We speculate that the residual visual function within this area may be explained by large receptive fields. Stanford, Tomlin, Comyn, Holland, and Pavesio (2005) investigated the association between VF loss with size and location of retinochoroidal scar and they found no association with size. However, a location close to the optic disc was associated with an absolute VF loss whereas a location further apart from the disc was associated with relative VF loss. Nevertheless, the authors hypothesized that an absolute defect was caused by the structural damage to the nerve fibres in scars close to the disc, and that the relative defects found in the more peripheral scars could be explained by larger receptive fields. This reasoning is in concordance with our assumption concerning the patient presented in this case study. Despite our results being consistent with the preserved visual function predicted by Stanford et al., 2005, past studies have shown that test-retest variability of retinal sensitivity is high in locations with damage (Artes, Iwase, Ohno, Kitazawa, & Chauhan, 2002) and due to unstable fixation and micro-saccades (Henson, Evans, Chauhan, & Lane, 1996).

Conclusion

The reported case demonstrates that the visual function may be preserved in the VF area corresponding to a retinochoroidal scar due to toxoplasmosis, in spite of loss of structures in the outer retinal layers as seen with OCT. Therefore, systematic investigations on the relationship between retinal structure and visual function in OT are warranted.

Disclosure

The authors report no conflicts of interest in this work.

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