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CASE REPORT

Torpedo maculopathy: a morphofunctional evaluation

Elisabetta Pilotto · Maria Elisabetta Zannin · Enrica Convento · Marta Cortese · Edoardo Midena

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Abstract To describe the optical coherence tomography (OCT), the standard short-wavelength fundus autofluorescence (SW-FAF) and near-infrared fundus autofluorescence (NIR-FAF), and the microperimetric findings in a child with a unique unilateral lesion of the temporal macula previously called torpedo maculopathy. A 4-year-old female with torpedo maculopathy was evaluated with spectral-domain OCT (SD-OCT), standard SW-FAF (excitation 488 nm, emission >500 nm) and NIR-FAF (excitation 787 nm, emission >800 nm). Microperimetry was performed to assess retinal sensitivity changes correlated to the macular lesion. SD-OCT showed an abnormally thin retinal pigment epithelium signal and an increased signal transmission in the choroid corresponding to the torpedo lesion with no neuroretinal changes. SW-FAF resulted in normal fluorescence of the lesion except for a small hyperfluorescent area at the tail level. NIR-FAF showed hypofluorescence corresponding to the lesion. Macular microperimetry showed reduced retinal sensitivity along the pigmented margins of the lesion with normal values over the lesion. The patient was re-evaluated 12 months later and no change was documented with all diagnostic techniques. This case supports a congenital defect of retinal pigment epithelium. The absence of both functional changes at lesion level and neuroretinal changes at OCT may depend on the very early detection of this lesion.

Keywords Torpedo maculopathy · Microperimetry · Spectral-domain optical coherence tomography · Standard short-wavelength fundus autofluorescence · Near-infrared fundus autofluorescence

Introduction

Torpedo maculopathy is a unique, apparently congenital asymptomatic flat macular lesion, which always localizes to the temporal fovea, with a pointed, torpedo-like tip directed toward the foveola [1]. It is often discovered on routine ophthalmological examination of asymptomatic patients. We report an unusual case of torpedo maculopathy in a child.

Case description

During routine eye examination, a 4-year-old girl was discovered to have a macular lesion in the right eye.

The study was performed at the Department of Ophthalmology, University of Padova, Padova, Italy.

E. Pilotto (✉) · E. Convento · M. Cortese · E. Midena
Department of Ophthalmology, University of Padova, via Giustiniani 2, 35128 Padua, Italy
e-mail: elisabetta.pilotto@unipd.it

M. E. Zannin
Department of Pediatrics, University of Padova, Padua, Italy

E. Midena
Fondazione G.B. Bietti per l'Oftalmologia, IRCCS, Rome, Italy

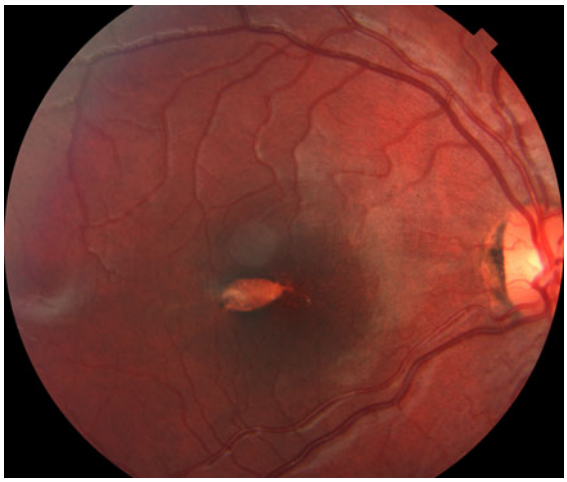

Fig. 1 Non-pigmented macular lesion with a tip toward the foveola and a hyperpigmented tail toward the ora serrata

The fellow eye was unremarkable. The right eye lesion was flat, non-pigmented, with a pointed-oval border directed toward the foveola, and hyperpigmented 'frayed tail' appearance toward the ora serrata. It measured 1.4 mm horizontally and 0.6 mm vertically and was located 3.5 mm temporal to the optic disc (Fig. 1). Ocular and medical history was unremarkable. The uncorrected visual acuity was 20/20 in both eyes. Extraocular motility, color vision and Amsler grid testing were negative. Optical coherence tomography (OCT) scans of the lesion (using both Cirrus HD-OCT; Carl Zeiss Meditec, Inc, Dublin, CA, USA

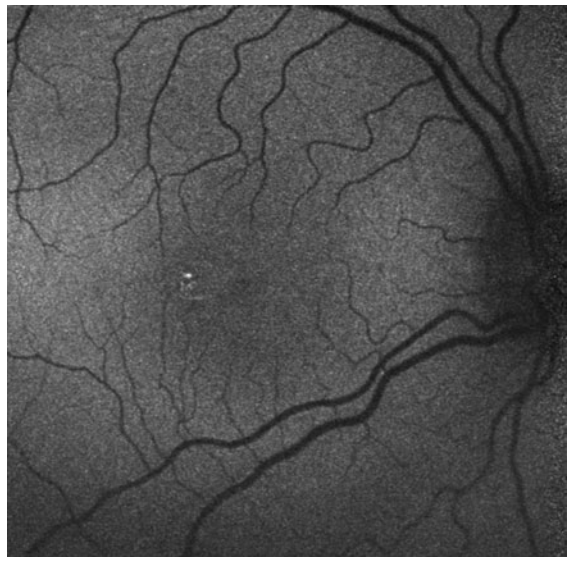

Fig. 3 Standard SW-FAF resulting in normal fluorescence of the lesion except for a small hyperfluorescent area at the tail level

and Heidelberg Spectralis HRA + OCT; Heidelberg Engineering, Heidelberg, Germany) showed an abnormally thin retinal pigment epithelium (RPE) signal and an increased signal transmission in the choroid corresponding to the torpedo lesion. The overlying outer and inner retina were normal (Figs. 2, 5). Standard short-wavelength fundus autofluorescence (SW-FAF) (excitation 488 nm, emission >500 nm) resulted in normal fluorescence of the lesion except for

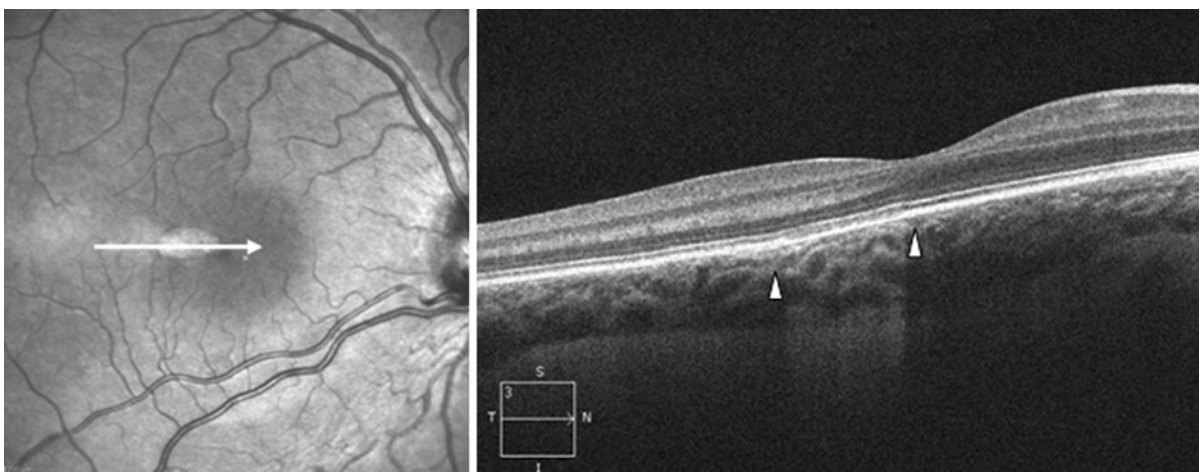

Fig. 2 Infrared fundus image (HRA2) and horizontal OCT (Cirrus HD-OCT). The arrow indicates the OCT scan site. The OCT scan shows an abnormally thin RPE signal and an increased signal transmission in the choroid corresponding to the torpedo lesion (*arrowheads*)

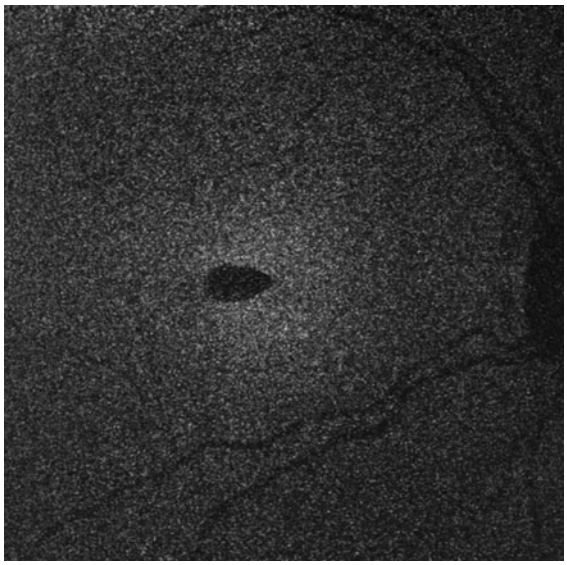

Fig. 4 NIR-FAF showing hypofluorescence corresponding to the area of the lesion

a small hyperfluorescent area at the tail level (Fig. 3). Otherwise, near-infrared FAF (NIR-FAF) (excitation 787 nm, emission >800 nm) showed hypofluorescence corresponding to the area of the lesion (Fig. 4)). Macular microperimetry (MP1 Microperimeter; Nidek, Gamagori, Japan) showed reduced retinal sensitivity along the pigmented margins of the lesion with normal values over the lesion (Fig. 5). The patient was re-evaluated 12 months later and no change was documented with all diagnostic techniques.

Comment

This case differs from previous reports. On standard SW-FAF, hypofluorescence corresponding to the lesion, reported by some authors, was not found [2, 3]. Standard SW-FAF allows topographic mapping of lipofuscin distribution in the outer segment of photoreceptors and in the RPE in vivo [4]. Standard SW-FAF has been shown to increase significantly with age [5]. When RPE is absent, a severely reduced signal is obtained in standard SW-FAF [6]. NIR-FAF visualizes the distribution of melanin, a fluorophore present in the RPE cells and choroid [7]. When melanin is absent, a severely reduced signal is obtained in NIR-FAF [8]. Therefore, we may speculate that in our case normal autofluorescence at standard SW-FAF may be due to the overall weak autofluorescence signal because of limited lipofuscin accumulation in such a young girl.

As retinal sensitivity is concerned, microperimetry, which allows an exact topographic relationship between retinal changes and retinal threshold, was

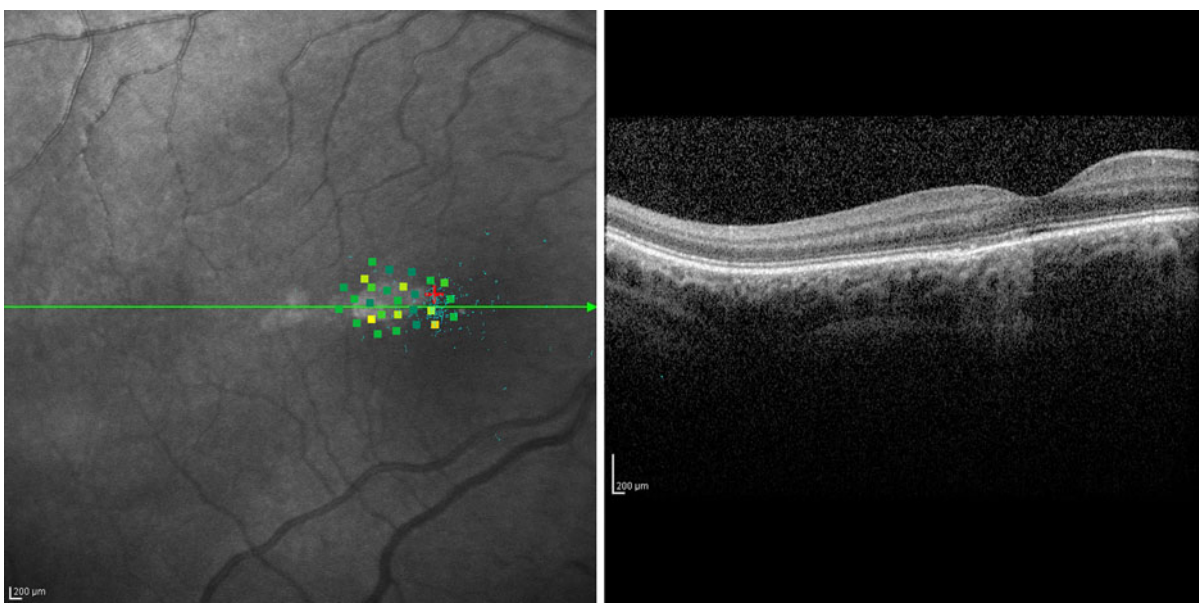

Fig. 5 Macular microperimetry (MP1 microperimeter) superimposed to the infrared image of the Heidelberg Spectralis (HRA + OCT) showing reduced retinal sensitivity along the pigmented margins of the lesion with normal values over the lesion

never previously reported. Retinal pigment epithelial atrophy causes secondary choriocapillaris loss and photoreceptor degeneration with secondary functional correlate [9]. Except for reduced threshold bordering the lesion, retinal sensitivity was normal with no scotoma being detected; this differs from previous reports [2, 3]. Therefore, we may speculate that in this case the RPE cells are amelanotic and thinner than normal but still functionally present as their overlying photoreceptors.

Histopathological studies of this particular lesion have never been reported, and to date its etiology remains speculative. Roseman and Gass [10] described a torpedo macular lesion they originally called a solitary 'hypopigmented nevus of the RPE'. Its uniform location and shape support a congenital defect of choroid and RPE due to the proximity to the emissary canal of the long temporal posterior ciliary artery and nerve [2]. Otherwise, a persistent defect in the development of RPE in the fetal temporal bulge has also been proposed [11]. This case supports a congenital defect of RPE. The absence of both functional changes at lesion level and neuroretinal changes at OCT may depend on the very early detection of this lesion. A longer morphofunctional follow-up of our case will be useful to assess if this is a different form of torpedo-like lesion without neuroretinal and visual field defects.

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