Int Ophthalmol (2013) 33:387–389
DOI 10.1007/s10792-013-9722-x

PHOTO ESSAY

Atypical findings in delayed presentation of unilateral acute idiopathic maculopathy

Soo Khai Ng · Andreas Ebneter · Jagjit S. Gilhotra

Received: 13 November 2012 / Accepted: 9 January 2013 / Published online: 23 January 2013

Abstract Unilateral acute idiopathic maculopathy is a rare disease affecting young healthy patients resulting in moderate to severe unilateral visual loss preceded by prodromal flu-like illness. We present a case of delayed presentation of unilateral acute idiopathic maculopathy associated with hand, foot and mouth disease.

Keywords Unilateral acute idiopathic maculopathy · Hand, foot and mouth disease · Delayed presentation

Introduction

Unilateral acute idiopathic maculopathy (UAIM) is a rare disease affecting young healthy patients resulting in moderate to severe unilateral visual loss preceded by prodromal flu-like illness [1–3]. Spontaneous resolution normally occurs over a period of 6 weeks with residual hyperpigmentation of the retinal pigment epithelium (RPE) resembling a bull's eye pattern [1–5].

Case

A 28-year-old male presented with a 10-day history of impaired visual acuity centrally in his right eye. Detailed questioning revealed that he developed fever, sore throat and rashes involving the palm of his hands and his feet 1 week prior to the onset of the visual symptoms. His ocular and medical history was otherwise unremarkable.

On presentation, best-corrected visual acuity was 20/80 right eye and 20/20 left eye. There was no relative afferent pupillary defect. Anterior segment and intraocular pressure were within normal limits bilaterally. Right fundoscopy showed gray-yellow wedge-shaped thickening of the RPE at the macula with slightly mottled hyperpigmentation (Fig. 1a). There was no evidence of vitreous cells, subretinal fluid, or papillitis. Fluorescein angiography of the right eye revealed mottled hyperfluorescence corresponding to the area of RPE thickening (Fig. 2a). This pattern was more pronounced in the late venous phase (Fig. 2b). Stratus optical coherence tomography (OCT), however, showed no significant abnormality (Fig. 2c).

Clinical presentations were suggestive of UAIM secondary to hand, foot and mouth disease. Compliment fixation test for Coxsackievirus A16 at week one and week three showed an 8- and 16-fold rise in the antibody titers, respectively, confirming the acute infection.

Given the self-limiting course of the disease and the stable visual acuity, the patient was managed conservatively with close follow-up. At week 6, his visual acuity normalized to 20/20 right eye, but an irregular-shaped, flat, pigmented macular scar persisted (Fig. 1b).

S. K. Ng (✉) · A. Ebneter · J. S. Gilhotra
South Australian Institute of Ophthalmology,
Royal Adelaide Hospital, Adelaide, SA, Australia
e-mail: soo.ng8098@gmail.com

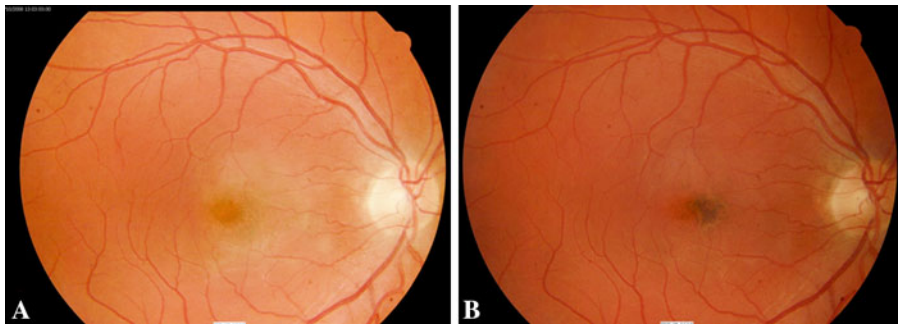

Fig. 1 **a** Right fundus photograph taken during first assessment. *Gray-yellow* wedge-shaped thickening of the RPE is noted at the macula. **b** At 6 weeks, an irregular, flat pigmented macular scar is seen. (*Note* scattered dots represent dust in the optical system)

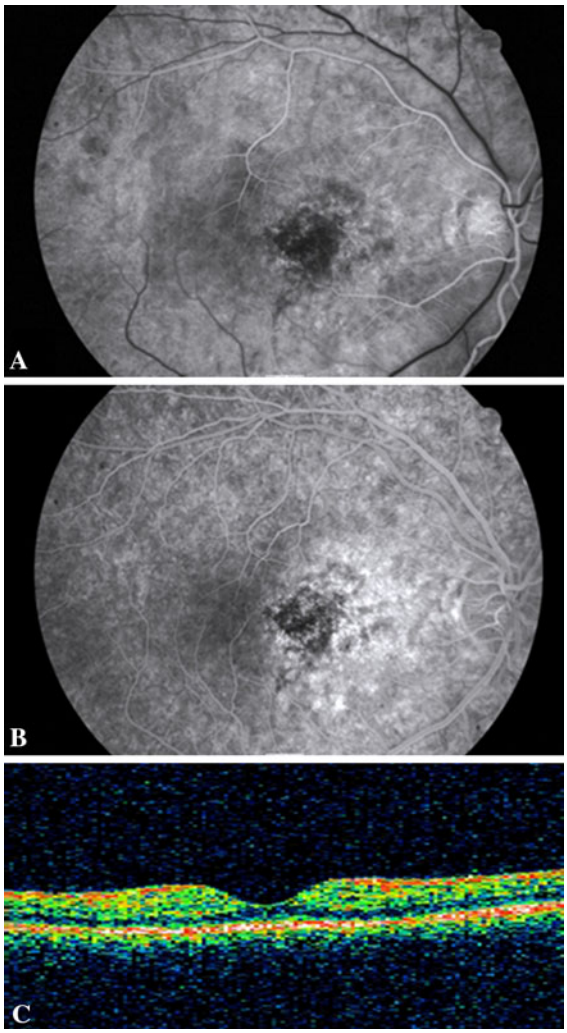

Fig. 2 **a** Fluorescein angiogram of the right eye in the arterial phase (18 s) reveals mottled hyperfluorescence corresponding to the area of RPE thickening with some areas of minimal subretinal hypofluorescence. **b** The late-stage image demonstrates more pronounced hyperfluorescence with some persistent blockage centrally. **c** Normal macular structure of the right eye on OCT (Zeiss Stratus)

Comment

Characteristically, UAIM is associated with neurosensory detachment of the macula and subfoveal fluid [1, 2]. We suspect that these findings had already resolved in our patient at the time of assessment, 10 days after symptoms onset, hence not observed. This matches the appearance of mottled hyperpigmentation in the macula which develops in conjunction with resolution of neurosensory detachment [1].

High-resolution OCT is a valuable tool to aid the diagnosis of UAIM in most cases as it can clearly portray the neurosensory detachment and the morphological changes of the RPE and outer retina [5, 6]. Aggio et al. [6] reported abnormal hyper-reflectivity and thickening at the level of RPE and outer retina in the acute phase and residual subfoveal hyper-reflectivity after resolution. These findings, however, were not observed in our patient. It may be possible that the OCT changes had already normalized as well.

This case highlights that clinical appearance of the posterior retina in UAIM can evolve quickly, and rapid spontaneous resolution is not uncommon. Some of the characteristic features of UAIM may not be seen if presentation is delayed.

Conflict of interest No conflicting relationship exists for any author.

References

1. Yannuzzi LA, Jampol LM, Rabb MF, Rabb MF, Sorenson JA, Beyrer C, Wilcox LM Jr (1991) Unilateral acute idiopathic maculopathy. Arch Ophthalmol 109:1411–1416
2. Freund KB, Yannuzzi LA, Barile GR, Spaide RF, Milewski SA, Guyer DR (1996) The expanding clinical spectrum of unilateral acute idiopathic maculopathy. Arch Ophthalmol 114:555–559

3. Haruta H, Sawa M, Saishin Y, Ohguro N, Tano Y (2010) Clinical findings in unilateral acute idiopathic maculopathy: new findings in acute idiopathic maculopathy. Int Ophthalmol 30(2):199–202
4. Beck AP, Jampol LM, Glaser DA, Pollack JS (2004) Is Coxsackievirus the cause of unilateral acute idiopathic maculopathy? Arch Ophthalmol 122:121–123
5. Jung CS, Payne JF, Bergstrom CS, Cribbs BE, Yan J, Hubbard GB 3rd, Olsen TW, Yeh S (2012) Multimodality diagnostic imaging in unilateral acute idiopathic maculopathy. Arch Ophthalmol 130(1):50–56
6. Aggio FB, Farah ME, Meirelles RL, de Souza EC (2006) Stratus OCT and multifocal ERG in unilateral acute idiopathic maculopathy. Graefes Arch Clin Exp Ophthalmol 244:510–516