Egg yolk in the eye: an ultrawide field evaluation

Koushik Tripathy, Rohan Chawla, Kanhaiya Mittal, Shreyas Temkar

Department of Retina and Uvea, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to

Dr Koushik Tripathy,
koushiktripathy@gmail.com

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DESCRIPTION

A 35-year-old man presented with gradual onset dimness of vision in both eyes noted for 3 years. The anterior segment, intraocular pressures and pupillary reactions were unremarkable in both the eyes. Best corrected visual acuity in both eyes was 6/18. The fundus showed a subretinal oval, smooth yellowish lesion 3×2 disc diameters in size at the macula in either eye simulating an 'egg yolk', with temporal white without pressure (WWOP) in either eye (figure 1A). Ultrawide field autofluorescence (Optos Inc, Marlborough, Massachusetts, USA) demonstrated brilliant autofluorescence corresponding to the subretinal lesion (figure 1B). Spectral domain optical coherence tomography (Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, California, USA) showed a collection of hyper-reflective material creating a mound at the macula (figure 1C). The inner-segment-outer segment junction (photoreceptor inner segment ellipsoid line)¹ was discontinuous at the fovea, possibly explaining the visual deficit in both eyes.² The ultrawide field fluorescein angiogram showed blockage of

choroidal fluorescence at the egg yolk lesion without any evidence of leakage. Window defects were evident bilaterally in the temporal periphery (figure 2A). The Goldmann visual field showed a central scotoma correlating to the egg yolk lesion (figure 2B). The Arden ratio on electrooculography (EOG) was 140% in the right eye and 133% in the left eye, confirming the diagnosis of Best vitelliform macular dystrophy.

Best disease is an autosomal dominant dystrophy. The vitelliform stage of the disease is characterised by a drastic fundus appearance of subretinal egg yolk with minimal or no visual compromise. EOG is typically abnormal. The accumulated material (lipofuscin) is brilliantly autofluorescent. Ultrawide field imaging uses a scanning laser ophthalmoscope with optics of an ellipsoid mirror to provide a single image of 200°, or 82% of the retina.³ The ultrawide field imaging also picked up peripheral WWOP and mild window defects on the fluorescein angiogram. Ultrawide field imaging often picks up peripheral retinal abnormalities in diseases that are clinically defined by macular involvement.

Figure 1 (A) Ultrawide field fundus pseudo-colour image (Optos Inc, Marlborough, Massachusetts, USA) of left eye shows subretinal oval, smooth egg yolk-like yellowish lesion with regular margins at macula, with temporal white without pressure (WWOP). (B) Ultrawide field autofluorescence (Optos Inc, Marlborough, Massachusetts, USA) reveals brilliant autofluorescence corresponding with the subretinal macular lesion. (C) Spectral domain optical coherence tomography (Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, California, USA) showed a collection of hyper-reflective material creating a mound at the macula.

Figure 2 (A) The ultrawide field fluorescein angiogram showed blockage of choroidal fluorescence at the egg yolk lesion, with no evidence of leakage, and window defects at the temporal periphery. (B) The Goldmann visual field showed a central scotoma correlating with the egg yolk lesion along with the blind spot.

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Learning points

- ▶ Vitelliform stage of best disease is characterised by an egg yolk lesion at the macula, which is brilliantly autofluorescent.
- ▶ Ultrawide field imaging captures 200° of the retina and picks up central as well as peripheral retinal disorders in a single image.

Contributors KT and RC had full access to all of the data in the study, and took responsibility for the integrity of the data and the accuracy of the data analysis. KT and KM were involved in the acquisition of data. KT and ST were involved in the analysis and interpretation of data. All the authors were involved in the drafting of

the manuscript, critical revision of the manuscript for important intellectual content and, study concept and design. RC was involved in the administrative, technical or material support: RC was involved in the study supervision.

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