Multimodal imaging in non-paraneoplastic autoimmune retinopathy

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Key words: Autoimmune retinopathy, anti-retinal antibodies, non-paraneoplastic autoimmune retinopathy, optical coherence tomography angiography

Autoimmune retinopathy (AIR) is a rare inflammatory disease, which remains underdiagnosed due to lack of specific diagnostic criteria.[1] Most of the patients present with bilateral, sudden progressive loss of vision,[2] visual field defects, and scotomas. The diagnostic criteria for AIR include any history of autoimmune disease in the family, clinical symptoms of progressive loss of vision, presence of circulating autoantibodies against the retinal antigens, and alterations on full-field electroretinography (ERG).[3] AIR can be classified into paraneoplastic and non-paraneoplastic AIR (npAIR). Paraneoplastic AIR is further divided into cancer-associated retinopathy and melanoma-associated retinopathy, vitelliform maculopathy, and bilateral diffuse uveal melanocytic proliferation.[4] AIR presents with diagnostic and management challenge, and the treatment of this spectrum of disease remains unclear although a proposed modified Delphi system of essential and supportive diagnostic criteria has been recently proposed.[4] So far, there has been only one case of AIR reported from India.[5] We describe multimodal imaging in a patient of serologically confirmed npAIR.

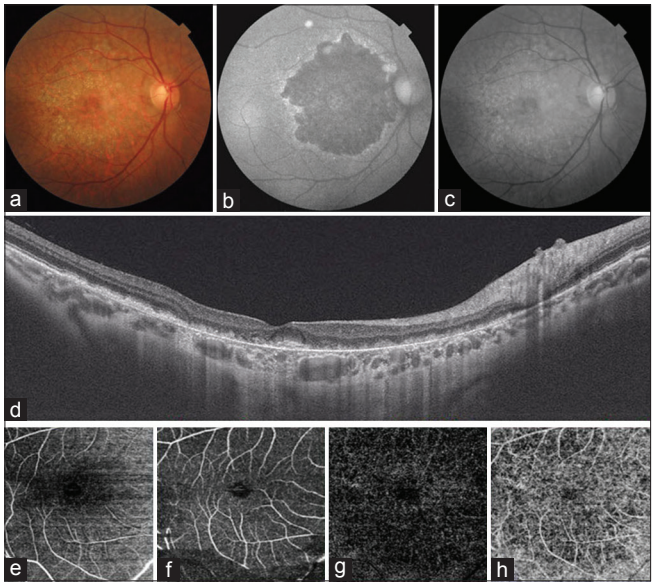

Figure 1: Color fundus photograph showed granular pigmentation and subretinal yellow-white deposits in right eye (Fig 1a). Autofluorescence showed marked hypoautofluorescence corresponding to the lesion with few areas of speckled hyperautofluoresence at the fovea (Fig. 1b). Red free photo demonstrated a giraffe-like pattern in the rest of the posterior pole (Fig. 1c). SS-OCT showed disorganization of the outer retinal layers with disruption of choriocapillaris (Fig. 1d). OCTA showed signal void areas in the choriocapillaris (Fig. 1e-h)

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A 44-year-old female presented with chief complaints of diminution of vision particularly at night, and glare. She gave a history of rapid loss of vision from 20/20 to 20/200 in both the eyes within 1 year. Fundus examination showed granular foveal pigmentation and subretinal yellow-white deposits

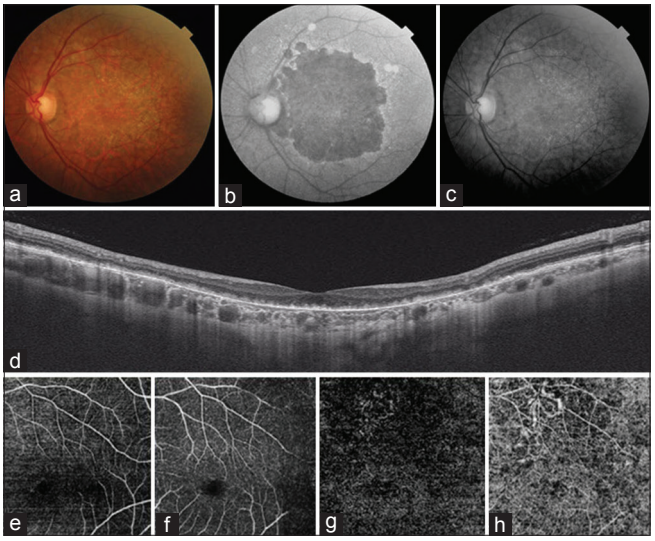

Figure 2: Color fundus photograph showed granular pigmentation and subretinal yellow-white deposits in (Fig. 2a). Autofluorescence showed marked hypoautofluorescence corresponding to the lesion with few areas of speckled hyperautofluoresence at the fovea (Fig. 2b). Red free photo demonstrated a giraffe-like pattern in the rest of the posterior pole (Fig. 2c). SS-OCT showed disorganization of the outer retinal layers with disruption of choriocapillaris (Fig. 2d). OCTA showed signal void areas in the choriocapillaris (Fig. 2e-h)

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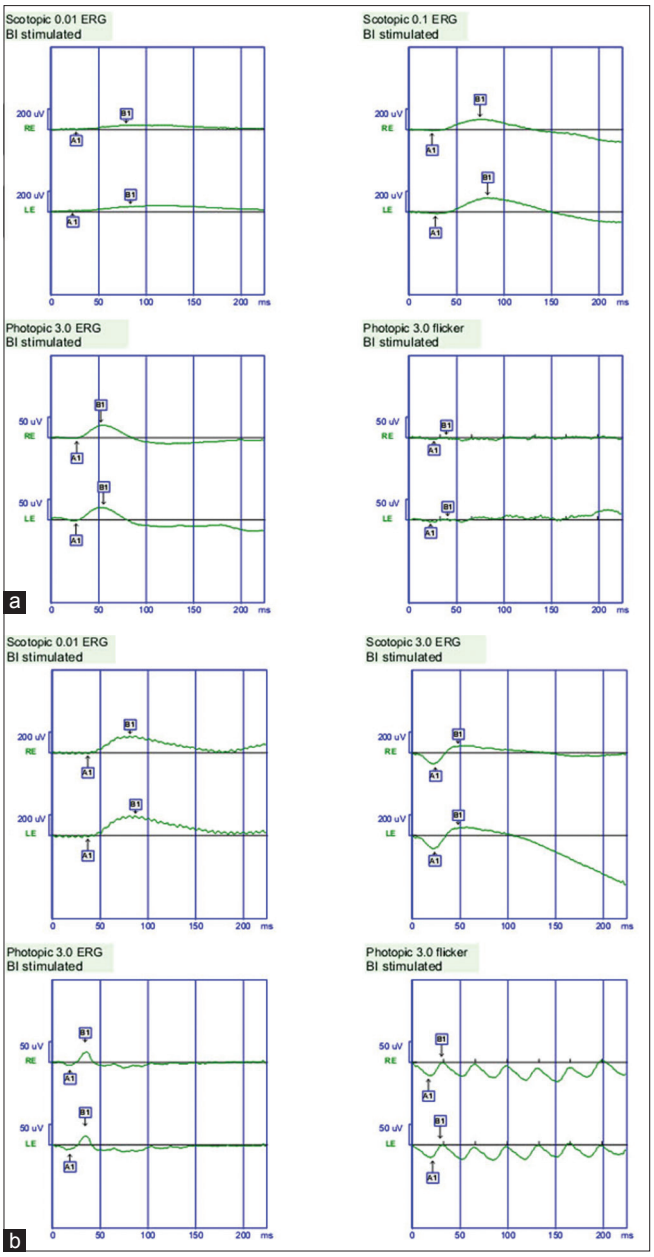

Figure 3: Electroretinography (ERG) showed a reduction in amplitude spikes on both scotopic and photopic responses (a). There was marked improvement in scotopic and photopic response on her full field ERG (b)

in both eyes [Fig. 1a and 2a]. Fundus autofluorescence (FAF) showed marked hypoautoflorescence corresponding to the lesion with few areas of speckled hyperautofloresence at the fovea [Fig. 1b and 2b]. Red-free photography demonstrated hyperpigmented spots corresponding to the subretinal deposits at the posterior pole [Fig. 1c and 2c]. Swept-source optical coherence tomography (OCT) showed significant disorganization of the outer retinal layers and inner segment and outer segment junction with disruption of choriocapillaris layer, and dilation of vessels in Sattler's and Haller's layers [Fig. 1d and 2d]. Optical coherence tomography angiography (OCTA) showed signal void areas in choroidal vasculature in the central affected area [Fig. 1e-h and 2e-h]. ERG showed a reduction in amplitude spikes

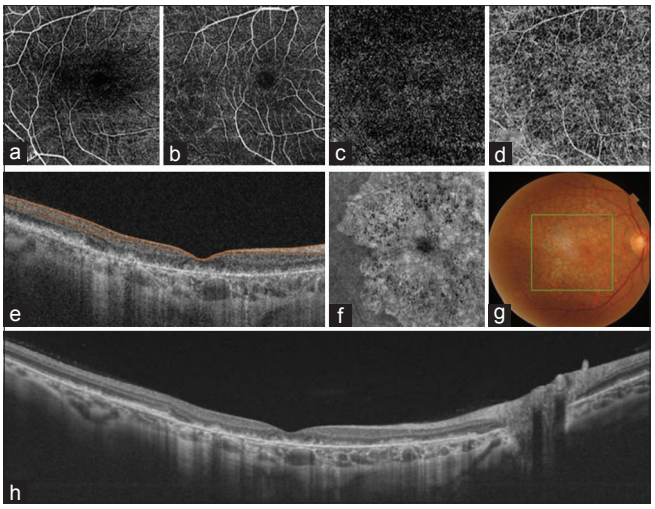

Figure 4: Right eye - Color fundus photographs, fundus autofluorescence, red-free photographs, optical coherence tomography (OCT) and OCT angiography (OCTA) showed no further changes from baseline (Figure 1)

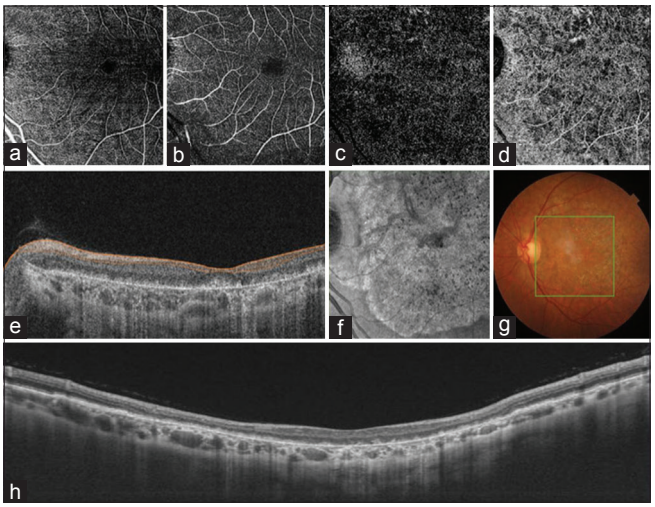

Figure 5: Left eye - Color fundus photograph, fundus autofluorescence, red-free photograph, optical coherence tomography (OCT) and OCT angiography (OCTA) showed no further changes from baseline (Figure 2)

on both scotopic and photopic responses [Fig. 3a]. The serology tests obtained on Western blot for all antiretinal antibodies against 23- (recoverin), 46- (enolase), 84-, 86-,96-, and 121-kDa proteins were positive. She was started on oral steroids. On her last visit at 12 months, the best-corrected visual acuity was 20/200; N24 with her fundus findings status quo and OCT, OCTA [Figs. 4 and 5], and FAF showed no further degeneration of outer retinal layers and choriocapillaris. However, there was marked improvement in scotopic and photopic responses on her full-field ERG [Fig. 3b].

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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