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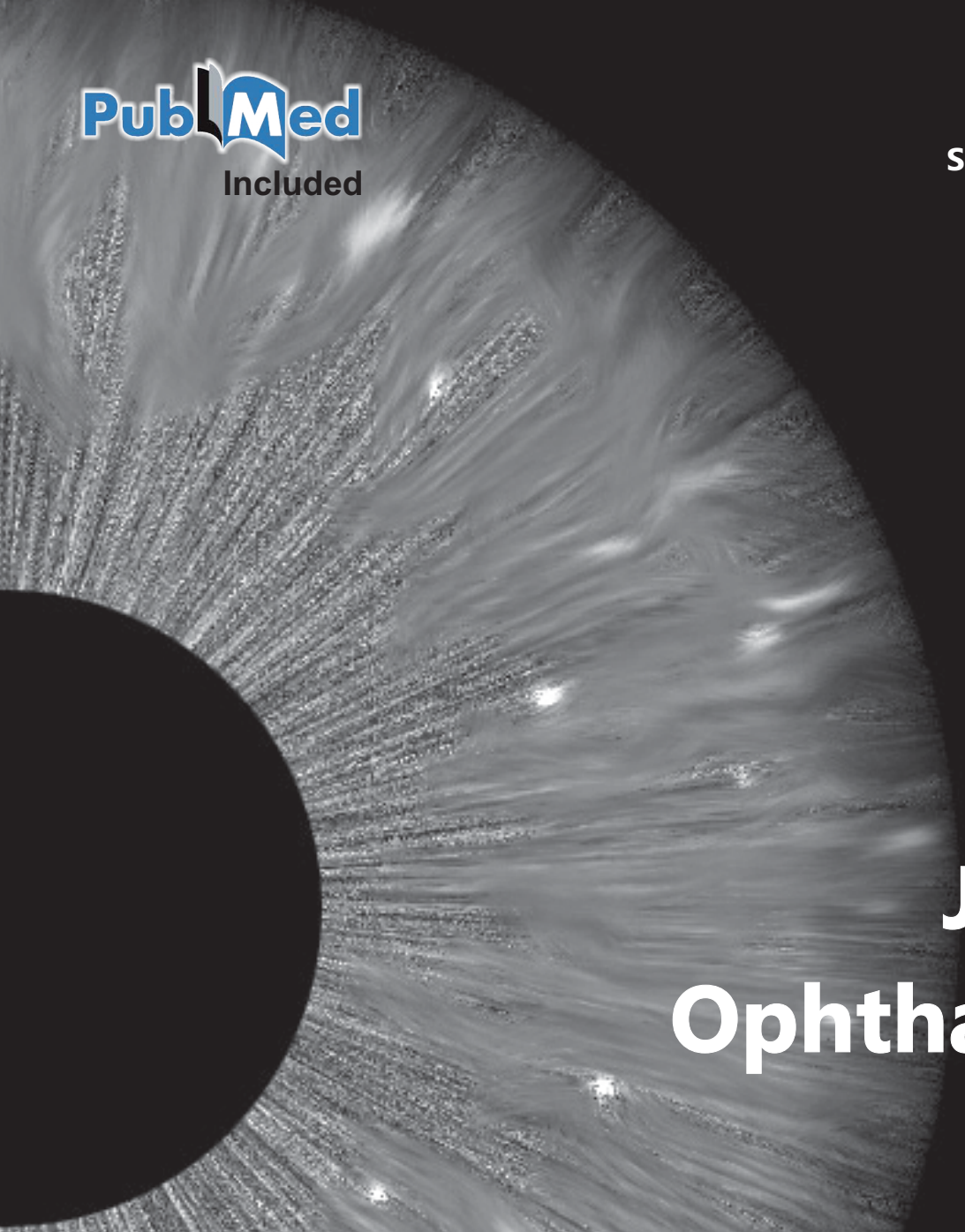

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Acute exudative polymorphous vitelliform maculopathy: To bolus or not to bolus?

Pier Luigi Grenga, Serena Fragiotta, Alessandro Cutini, Enzo Maria Vingolo

Abstract:

Acute exudative polymorphous vitelliform maculopathy (AEPVM) is a rare bilateral maculopathy characterized by chronic and long-term course. We report a case of AEPVM with an unusual presentation and management in a middle-aged man. He presented with clinical features of bilateral AEPVM accompanied by multiple intraretinal cysts, with a sudden increase of intraretinal fluid and visual function deterioration over a span of few days. Therefore, we administered empirically an intravenous bolus injection of methylprednisolone. One week after, there was a full recovery of visual acuity and cystic intraretinal spaces completely disappeared.

Keywords:

Acute exudative polymorphous vitelliform maculopathy, fluorescein angiography, intraretinal cysts, optical coherence tomography, steroid therapy

Introduction

Acute exudative polymorphous vitelliform maculopathy (AEPVM) is a rare retinal disorder first described by Gass as bilateral multiple yellow-white subretinal lesions and exudative macular detachment. Almost all patients are young men between 20 and 30 years with acute onset of headaches followed by vision loss, with incomplete resolution over a period of 1–2 years.[1] Several authors have empirically administered oral corticosteroid therapy, but its efficacy has not been proven. The agent most commonly used is oral prednisone, which is slowly tapered over weeks or months.[1,2]

The aim of this report was to describe an unusual presentation of AEPVM characterized by intraretinal cysts resolved with an intravenous bolus injection of methylprednisolone.

Case Report

A 39-year-old man presented to our department with sudden onset of blurred vision in both eyes and persistent headache for several days. He had a recent history of hypertension treated with lisinopril, and he was an inactive carrier of hepatitis B. His ocular history was unremarkable. He denied any flu-like symptoms in previous days. Written informed consent was obtain from the patient before examination.

His best-corrected visual acuity (BCVA) was 20/22 in both eyes, the anterior segment was unremarkable, and Goldmann applanation tonometry was within normal limits in both eyes. Pupils were equal, round, reactive to light with extraocular movements intact. His color vision was normal. Fundus examination revealed a serous macular detachment associated with multiple round yellowish lesions along vascular arcades within posterior pole in both eyes. The optic nerve and peripheral retina were normal, but retinal vessels were convoluted with

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some arteriovenous nicking. No signs of vitritis were noted. Fundus autofluorescence showed slight mild hyperautofluorescence of the multiple round lesions along vascular arcades. Fluorescein angiography (FA; HRA2, Heidelberg Engineering) showed focal filling defects along vascular arcades in the early phase and slight mild diffuse hyperfluorescence in the late phase without leakage [Figure 1].

Spectral domain optical coherence tomography (SD-OCT, software version 5.4.7.0; Heidelberg Engineering GmbH, Heidelberg, Germany) showed a marked alteration in retinal profile with a large neurosensory retinal detachment and multiple intraretinal cystic spaces located in outer nuclear layer, within the 3-mm ring of the ETDRS grid [Figure 2a and b].

A laboratory workup was performed including complete blood count with differential, liver function test, autoantibodies, rheumatoid factor, purified protein derivative of tuberculin (with anergy panel), antistreptolysin O titer, HIV-1/HIV-2 tests, herpes simplex virus blood test, treponema pallidum-specific tests, rubella (IgG, IgM), Epstein-Barr Virus (IgG, IgM), and cytomegalovirus (IgG, IgM). Laboratory tests and radiological chest examination were normal. To exclude the presence of melanoma, a dermatological consultation was required. A dermatological consultation was required and it ruled out the presence of melanoma. No treatment has been administered, and the patient was monitored every week.

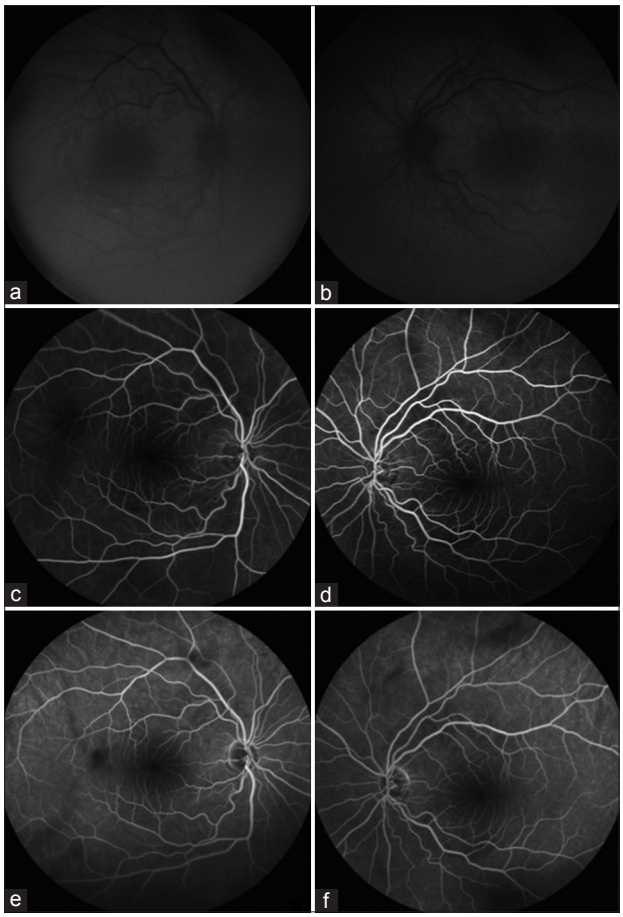

Figure 1: Autofluorescence imaging shows faint hyperautofluorescence of the vitelliform lesions along vascular arcades both in the right (a) and left (b) eyes. (c and d) Early phase of fluorescein angiogram demonstrates focal filling defects in correspondence of vitelliform lesions and no abnormal fluorescence of subfoveal lesion in both eyes; (e and f) Late-phase angiogram shows slightly diffuse late staining of vitelliform oval-shaped lesions along arcades, with no leakage and no modification in subfoveal lesion

A month later, his BCVA dropped to 20/40 in both eyes. Fundus evaluation revealed further increase in subretinal fluid and in the amounts of yellow oval-shaped deposits along vascular arcades. SD-OCT examination showed a significant increase in retinal thickness and number and extension of cystic spaces that involved both inner and outer nuclear layers, exceeding the 6-mm ring of the ETDRS grid in both eyes [Figure 2c and d]. Because of this worsening, steroid therapy was administered as 1 g of intravenous methylprednisolone per day for 3 days on an empirical basis with no tapering. One week after starting treatment, BCVA was 20/20 in both eyes. SD-OCT showed a significant improvement in retinal thickness and complete disappearance of cystic spaces in both eyes [Figure 2e and f]. After 8 months, subretinal lesion evolved to gravitational, meniscus-like, yellowish lesion in both eyes. Fundus autofluorescence showed slight mild hyperautofluorescence of both the multiple round lesions along vascular arcades and a meniscus-like appearance in the inferior part of paramacular region, just above inferior vascular arcade. FA demonstrated hypofluorescent area corresponding to the lesions in the early phase and late staining [Figure 3].

At 12-month follow-up visit, examination revealed 20/18 BCVA and no subretinal fluid bilaterally, with few residual vitelliform deposits along vascular arcades in both eyes; SD-OCT showed a normal retinal profile, persistence of the thickening of ellipsoid band, and minimum residual of subretinal fluid in both eyes [Figure 2g and h]. After 45 months, BCVA was 20/20 in both eyes, anterior chamber was deep and quiet, normal iris and pupillary reflex, crystalline lens was transparent. Fundus examination revealed an increase in subretinal fluid in the left eye and persistence of vitelliform deposits within the subretinal space along vascular arcades [Figure 4]. There was no vitreous cell and the peripheral retina was flat.

Discussion

The pathogenesis of AEPVM is still unclear, and an inflammatory or immune-mediated pathogenesis

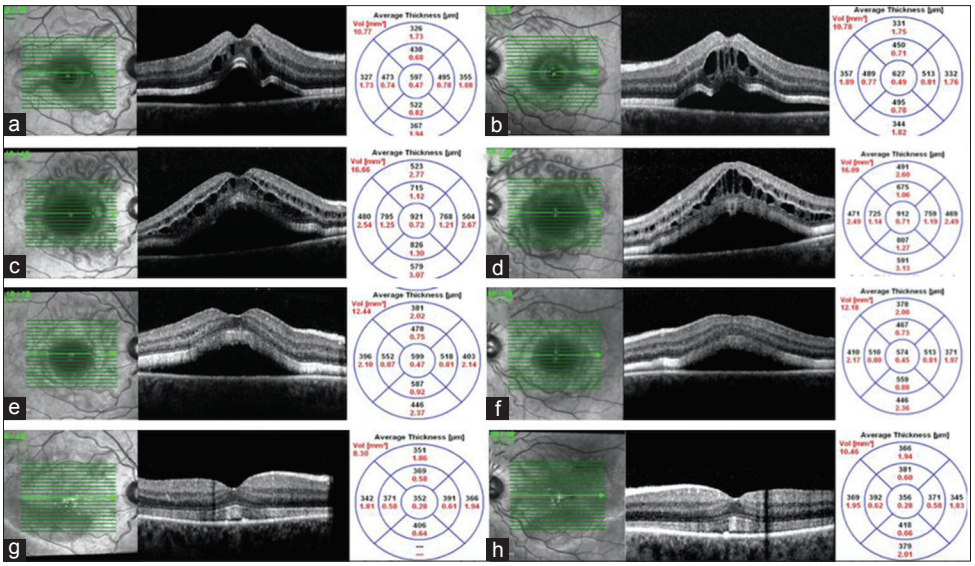

Figure 2: Spectral domain optical coherence tomography (baseline) shows large serous neurosensory detachment with confluent intraretinal cysts in outer nuclear layer in the right (a) and left (b) eyes; (c and d) At 1 month, marked increase in subretinal fluid with thickening of ellipsoid portion of the inner segments; intraretinal cysts are increased in number and shape involving both inner and outer nuclear layer. (e and f) One week after bolus, reduction in subretinal fluid and complete disappearance of intraretinal cysts. (g and h) At 12 months, normal retinal profile with a minimum residual subretinal fluid and ellipsoid band thickening

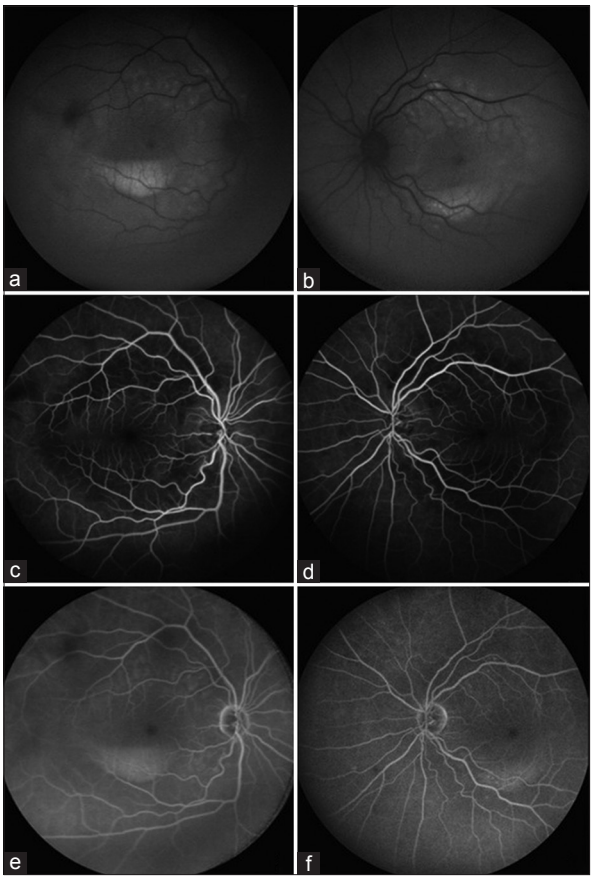

Figure 3: Autofluorescence imaging shows hyperautofluorescence of both multiple roundish lesions along vascular arcades and meniscus-like located inferiorly in paramacular region and near vascular arcade both in the right (a) and left (b) eyes. (c and d) Early phase of fluorescein angiogram demonstrates hypofluorescent area corresponding to the lesions as a blockage of the background fluorescence in both eyes. (e and f) Late-phase angiogram shows late staining of both vitelliform lesions along vascular arcades, especially in the inferior arcade and in the paramacular region

of AEPVM has been proposed. AEPVM is often described in association with malignant neoplasm, and autoantibodies were identified in serum samples suggesting an autoimmune response direct against retinal pigment epithelium cells.[1,2]

This report shows an unusual clinical presentation of AEPVM characterized by intraretinal cysts within inner and outer nuclear layers managed with steroid bolus. Although this clinical presentation is rarely reported in literature, the administration of steroid in such cases has never been reported.[3,4] In our case, the increasing in intraretinal cysts was accompanied by visual acuity decay. Therefore, we decided to treat the patient with steroid on an empirical basis. The choice of bolus as administration route was driven by the acute presentation and general status of our patient, to reduce side effect on hypertension due to long-term oral prednisone, as the patient already suffering from poorly controlled systemic hypertension.

Methylprednisolone is a potent anti-inflammatory agent (potency 1.25 times vs. prednisolone) with a low tendency to produce sodium and water retention (glucocorticoid: mineralocorticoid effect = 6:1).[5] Moreover, it has been successfully used in the treatment of some ophthalmic diseases as optic neuritis.[6]

In the present case, the disappearance of intraretinal cysts and visual acuity recovery were so fast to be reasonable related to the effect of methylprednisolone bolus. Nevertheless, the effect of the bolus did not affect the course of the disease with as demonstrated by persisting

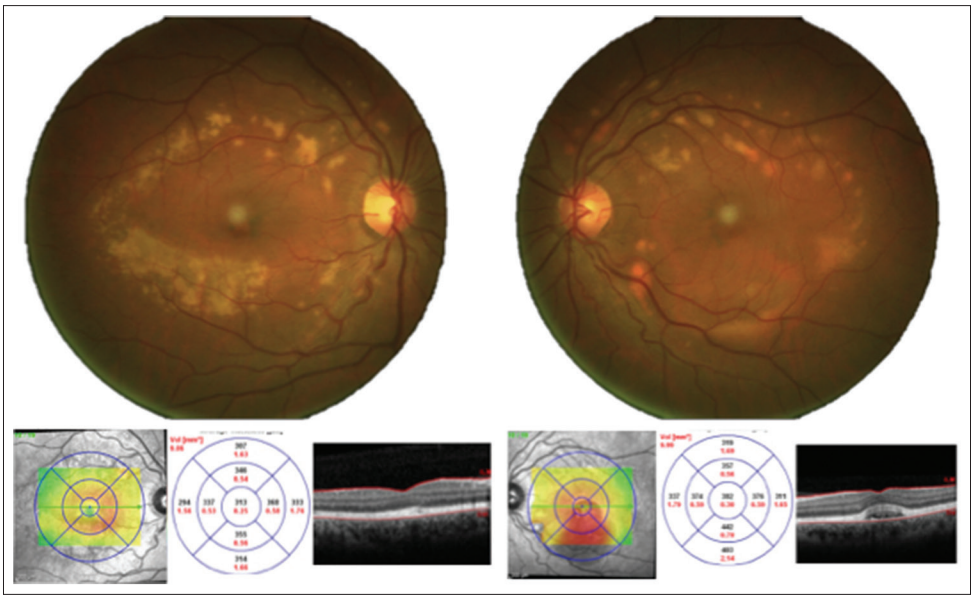

Figure 4: Retinography and spectral domain optical coherence tomography at 45 months of follow-up. Retinography shows the persistence of yellowish subretinal vitelliform-like deposits along vascular arcades both in the right (upper right) and left (upper left) eyes. Spectral domain optical coherence tomography scans show no significant changes in the right eye (bottom right) and an increase of subretinal fluid and hyperreflective material inside in the left eye (bottom left)

gravitational vitelliform-like deposition and incomplete subretinal fluid resolution till 45 months later. This may suggest that steroid effect is mostly related to the acute manifestation of the disease, justifying the variable outcomes reported with steroid use. Prolonged systemic steroidal therapy has been used on empirical basis, but its effect is still debated and it seems to do not alter the natural course of the disease.[1,2,7]

Conclusion

This is the first report that describes intraretinal cysts associated with visual decline as early presentation of AEPVM responding to methylprednisolone bolus. However, the disease course seems to be not influenced by steroidal therapy. These findings add more information about disease presentation and management. Further studies are necessary to evaluate the rationale about the use of corticosteroids in such cases. To date, no studies have reported the use of steroid in case of sudden visual decay associated with increased subretinal fluid and intraretinal cysts. Our findings add some interesting information on AEPVM complications and their management, but further studies would be desirable to evaluate the rationale of steroidal therapy in such cases.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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