American Journal of Ophthalmology Case Reports 20 (2020) 100870

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Case report

Imaging features and functional outcome in a case of idiopathic acute exudative polymorphous vitelliform maculopathy

Michael Sommer *, Christoph Singer, Daniel Djavid, Gerald Seidel

Universitäts-Augenklinik Graz, Medical University of Graz, Auenbruggerplatz 4, 8036, Graz, Austria

ARTICLE INFO

Keywords:
AEPVM
Multimodal imaging
OCT

1. Case report

A 52-year-old man presented at a tertiary ophthalmology referral center complaining of increasingly blurry vision on both eyes for one month. His previous medical- and family history were unremarkable.

Visual acuity (VA) was 20/50 on the right and 20/40 on the left eye. Fundoscopy revealed bilateral subretinal bleb-like vitelliform lesions mainly within the vascular arcades and serous foveal detachments (Fig. 1). These lesions appeared hyperfluorescent with hypofluorescent borders in fundus autofluorescence (FAF) (Fig. 2). Fluorescein angiography was unremarkable apart from hypofluorescence of serous macular detachments (Fig. 3A). Late-phase indocyanine green angiography revealed hyperfluorescence of the subretinal deposits (Fig. 3B). Based on these findings, the diagnosis of acute exudative polymorphous vitelliform maculopathy (AEPVM) was made. A workup for uveitis and systemic malignancies was negative. No genetic testing was performed. The patient agreed to a trial of oral acetazolamide (250mg bid) and improved continuously gaining 20/16 vision on both eyes within 8 months as the subretinal fluid resolved with minor vitelliform residues (Fig. 4).

2. Discussion

AEPVM is a rare condition leading to an acute onset of blurry vision due to vitelliform lesions and a serous foveolar detachment.[1] The diagnosis is based on the characteristic fundoscopic features. Multimodal imaging including OCT and FAF can be helpful to monitor the clinical course. Because AEPVM cases have been associated with malignancies, a workup to rule out systemic malignancies is recommended.[2] Although AEPVM shares similarities with Best's disease and pattern dystrophies, BEST1 and peripherin/RDS genes are normal.[3] The prognosis is excellent with most patients regaining excellent VA as serous detachments resolve over a course of several months.[3] Minor vitelliform residues might persist for over a year (Figs. 2 and 4).

3. Conclusion

AEPVM is a self-limiting condition and patients usually regain excellent vision. The resolution of subretinal fluid may take several months and vitelliform material might persist for a longer period. A workup for systemic malignancies should be performed. We cannot claim the effectiveness of acetazolamide from this single case.

Acknowledgement and disclosures

The patient consented to publication of the case in writing.
Funding: No funding or grant support.
The authors have no financial disclosures.
All authors attest that they meet current ICMJE criteria for authorship.

Acknowledgements

none.

* Corresponding author.
E-mail address: michael.sommer@medunigraz.at (M. Sommer).

https://doi.org/10.1016/j.ajoc.2020.100870
Received 22 April 2020; Received in revised form 15 July 2020; Accepted 9 August 2020
Available online 13 August 2020

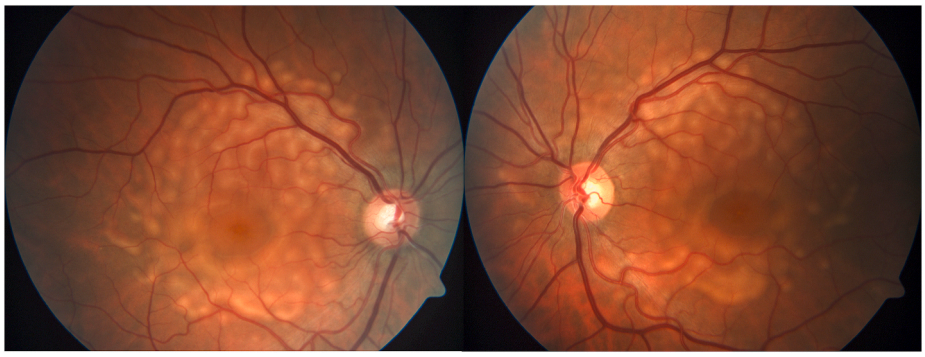

Fig. 1. Fundus photograph.
Fundus photograph one month after symptom onset.

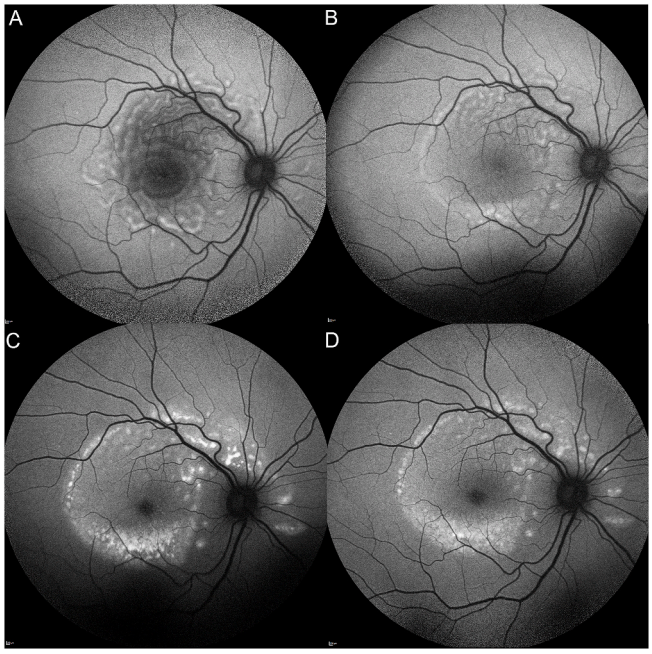

Fig. 2. Autofluorescence.
Autofluorescence at baseline (A) 4 months (B) 9 months (C) and 16 months (D).

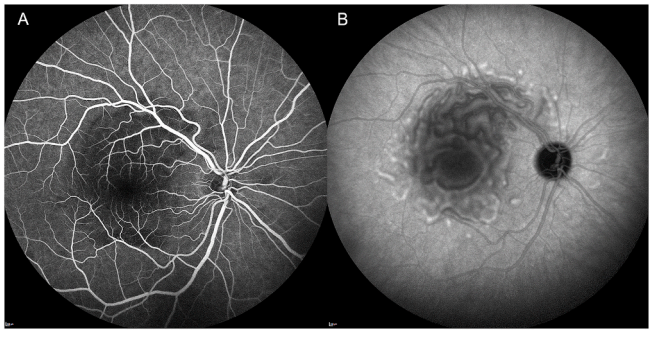

Fig. 3. Angiography.
Fluorescein- (A) and indocyanine green angiography (B) one month after symptom onset.

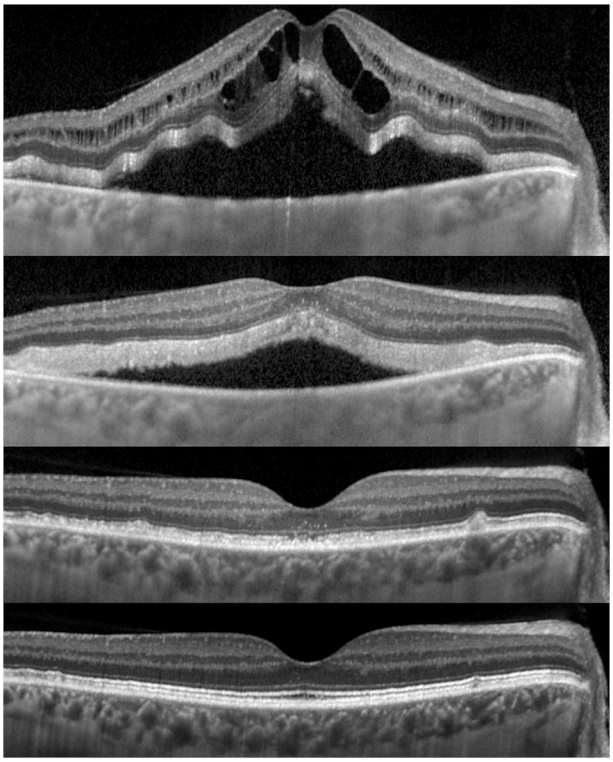

Fig. 4. OCT.
OCT at baseline, 3 months, 6 months and 24 months.

References

1. Gass JD, Chuang EL, Granek H. Acute exudative polymorphous vitelliform maculopathy. *Trans Am Ophthalmol Soc*. 1988;86:354–366.
2. Koreen L, He SX, Johnson MW, Hackel RE, Khan NW, Heckenlively JR. Anti-retinal pigment epithelium antibodies in acute exudative polymorphous vitelliform maculopathy: a new hypothesis about disease pathogenesis. *Arch Ophthalmol*. 2011; 129:23–29.
3. Barbazetto I, Dansingani KK, Dolz-Marco R, et al. Idiopathic acute exudative polymorphous vitelliform maculopathy: clinical spectrum and multimodal imaging characteristics. *Ophthalmology*. 2018;125:75–88.