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Case Report

Acute Posterior Multifocal Placoid Pigment Epitheliopathy Following COVID-19 Vaccination

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Abstract

A 41 year-old patient presented with left paracentral scotoma with initial visual acuity of 6/6 in the affected eye. Review in the clinic few days later showed deterioration of left eye vision to 6/36 and examination revealed deep retinal lesions in the posterior pole with foveal center involvement and serous retinal detachments. A diagnosis of acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was made. Detailed medical history unveiled COVID-19 vaccination 6 days prior to the visual symptoms.

Keywords: Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE); COVID-19 Vaccination

Introduction

APMPPE following certain vaccines is described in the literature. In this case report, APMPPE was thought to be caused by COVID-19 vaccination.

Case Presentation

A 41 year-old Caucasian male with no previous significant medical history, presented out of hours to the urgent care eye clinic with the main complaint of left eye visual blurring and paracentral scotomas. He denied eye pain, redness, photophobia or photopsias. At presentation his vision was right eye 6/5 and left eye 6/6 with no relative afferent pupillary defect (RAPD). Examination showed no signs of anterior chamber or vitreous inflammation in either eye. Fundal examination showed no abnormality of the right fundus but on the left there were multifocal ill-defined deep creamy yellow outer retinal placoid lesions in the posterior pole, mainly within the arcades and few surrounding the optic disc and outside the arcades. The ocular coherence tomography of the macula on Topcon Triton (SS-OCT, Topcon, Tokyo, Japan) showed swelling of the retinal layers with subtle changes at the outer retinal layer and the retinal pigment epithelium (RPE) (Figure 1). A referral to the specialized uveitis clinic was done where he was assessed three days later. The patient admitted worsening of his left eye central vision since his first visit. The visual acuity was found to be stable in the right eye 6/5 while a significant reduction of the left eye vision to 6/36 was illustrated. The Patient had left relative afferent pupillary defect (RAPD) and his color vision on Ishihara was normal in the right eye 15/15 while it was reduced in the left eye to 3/15. Static perimetry on Humphrey 120 point to screen his visual field was normal on the right side, whereas the left side showed central scotomas. The assessment on that visit showed no inflammatory cells in the aqueous or the vitreous in either eye. The right fundal examination remained normal, however the left fundal examination showed multifocal same age ill-defined deep creamy yellow outer retinal placoid lesions in the posterior pole, mainly within the arcades and few surrounding the optic disc and outside the arcades with no chorioretinal scars or healed lesions (Figure 2). These lesions looked confluent in some areas with surrounding smaller lesions. As a sign of progression, newly found localized small serous retinal detachments in the peripapillary and the fove-

al areas were evident on examination. It is also worth mentioning the absence of retinal vasculitis and optic disc swelling clinically. Repeat ocular coherence tomography on Topcon Triton (SS-OCT, Topcon, Tokyo, Japan) showed outer retinal and retinal pigment epithelial (RPE) disruption with small pockets of subretinal fluid in at least 3 areas in the left posterior pole (Figure 3A-3C). A detailed past ocular history revealed no previous ocular problems including trauma or surgery. Family history was also negative for any hereditary or inflammatory eye disease. Past medical history was unremarkable and review of systems was of no significance except for a recent history of COVID-19 mRNA BNT162b2 (Pfeizer/BioNTech) vaccination which was given 6 days prior to the onset of visual symptoms (i.e. 9 days before presentation to the urgent care eye clinic). The patient experienced mild flu-like symptoms following the vaccine which lasted around 24 hours and then resolved spontaneously. To be more specific, the patient did not experience any headaches, meningism or neurological symptoms. Fundus Fluorescein Angiogram (FFA) and Indocyanine Green Angiogram (ICGA) (HRA2, Heidleberg, Germany) were performed urgently. FFA showed unilateral left early hypofluorescent and late staining multiple lesions (Figure 4A and 4B) and on ICGA there were unilateral posterior pole initial hypocyaniscence in the macula and peripapillary area which represented precapillary choroidal hypoperfusion and persistent multiple hypocyaniscent dark lesions (Figure 5A and 5B). The clinical picture was very suggestive of unilateral APMPPE and the differential diagnosis of serpiginous or ampiginous choroiditis were also considered due to the peripapillary involvement and the confluent configuration of some of the lesions although the latter two differentials were believed to be less convincing compared to APMPPE. Also in the differential diagnosis come the variants of relentless placoid chorioretinitis and persistent placoid choroidopathy but these can only be concluded after a reasonably long period of follow up which would allow recognition of the pattern of progression and recurrence. The patient was counselled regarding the diagnosis of APMPPE and given the foveal involvement and the dramatic deterioration of his vision, the patient was offered a moderate dose oral Prednisolone (0.75 mg/kg) which was started immediately. Blood investigations, which were sent before the treatment was started, included full blood count, erythrocyte sedimentation rate, renal functions, bone profile, liver functions, C-reactive protein, serum angiotensin converting enzyme, immunoglobulin and serum electrophoresis, complement level, syphilis screen, hepatitis B and C serology as well as chest X-ray were all unremarkable. More importantly, QuantiFERON-TB Gold test was sent to exclude tuberculosis as a cause for this clinical picture and the test came back reassuringly negative. The other important causes of posterior uveitis that can mimic this presentation such as sarcoidosis and syphilis have been excluded by means of normal investigations or negative review of systems.

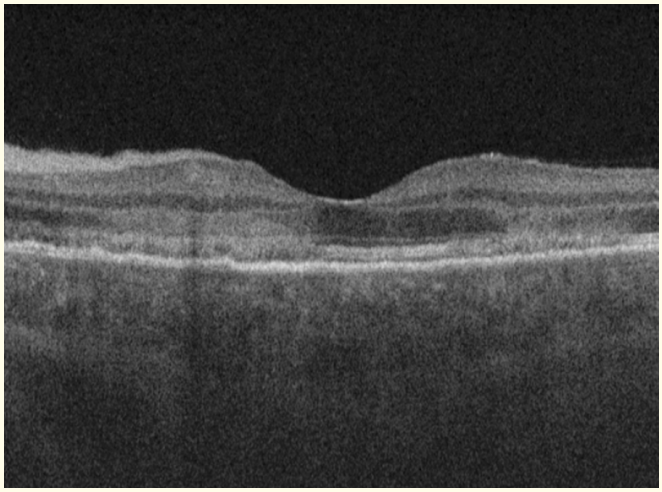

Figure 1: OCT of the right macula showing abnormal thickening of the retinal layers with changes at the outer retina and the RPE.

Figure 2: Fundal photos showing normal right posterior pole and left multifocal ill-defined creamy yellow deep retinal lesions in the posterior pole with foveal involvement.

Figure 3: OCT showing the subretinal fluid at the A) fovea, B) Extrafoveal and C) peripapillary zone.

Figure 4: FFA A) early phase at 19 seconds showing blocked fluorescence secondary to placoid lesions and hypoperfusion of the precapillary choroid and B) late phase at 5 minutes which shows staining but no leakage of the lesions.

Figure 5: ICGA A) early phase at 35 seconds showing hypocyaniscence in the posterior pole likely due to choroidal hypoperfusion and B) late stage at 5 mins showing persistent hypocyaniscence pertaining to the placoid lesions masking-effect.

The patient was reviewed 4 days after the initiation of systemic corticosteroid treatment and his left eye vision improved to 6/9 again with complete resolution of the small serous retinal detachments and remarkable improvement of the outer retinal swelling and RPE changes. The patient experienced no symptoms in the healthy-looking right eye which preserved normal vision of 6/5. The patient was reviewed again after gradual taper of oral Prednisolone over 6 weeks and the good effect was sustained. The final visual acuity was 6/5 and 6/6 in the right and left eye; respectively. There were still subtle residual outer retinal and RPE changes which are expected to be permanent (Figure 6).

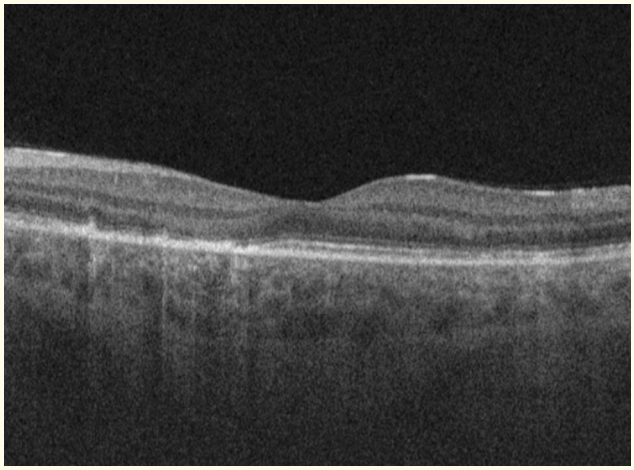

Figure 6: OCT of the left macula showing resolution of subretinal fluid and retinal swelling with more defined outer retinal structures.

Discussion and Conclusion

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is a rare form of posterior uveitis that was first described by Gass [1] in 1968. This condition is typically bilateral, either simultaneously or sequentially; however unilateral disease has also been described [2]. APMPPE is considered a condition under the umbrella term of white dots syndrome with no sex predilection, despite previous belief that it affects women more than men. Usually occurs between the 2nd and 4th decades. The etiopathology of this entity remains mysterious with more evidence in favor of the theory which assumes that the primary insult occurs at the inner

choroid level leading to the outer retinal changes, specifically choriocapillary perfusion abnormality [3-6]. The disease is thought to have self-limiting course with the lesions spontaneously resolving in weeks to months [7]. Patients with foveal involvement bear worse prognosis with a visual acuity of 6/15 or worse [8].

In this case, the presentation was primarily unilateral and since prompt treatment with systemic corticosteroids was given, it will be difficult to speculate whether this would have eventually evolved to involve the unaffected right eye.

A prodromal phase has been typically described in most cases of APMPPE and this may include viral or flu-like syndrome (fever, cough, lymphadenopathy, myalgia, malaise and nausea), headache or rarely neurological symptoms [9].

Although association with neurological involvement, namely cerebral vasculitis, was rarely described in other cases, in our case there was no indication to investigate this further given the absence of any neurological symptoms and the rarity of such complications.

There are anecdotal reports of APMPPE cases which happened following vaccinations. This association mechanism was attributed to the possible molecular mimicry and sequence similarities between the introduced antigen and the RPE which may trigger an autoimmune reaction by the host. Examples of vaccinations which were thought to be associated with APMPPE are, but not restricted to, human flu vaccine [10], swine flu vaccine [11], hepatitis B vaccine [12], meningococcal C conjugate vaccine [13] and varicella vaccine [14].

It has been presumed that this case is caused by the COVID-19 vaccination given the short interval between the vaccination and the appearance of the symptoms in addition to the unremarkable results of the investigations in the absence of any other system involvement. This presumption is difficult to prove given that there is no objective test to confirm this causation and that a coincidental occurrence cannot be ruled out completely.

To the best of our knowledge, until the time of writing this article, this is the first report of APMPPE presumed to be secondary to the novel COVID-19 vaccine. This is thought to be of a paramount importance given the mass vaccination campaign which is taking place globally aiming to stop the burden of the COVID-19 pandemic, hence the risk of more patients presenting with similar findings. This also highlights the importance of obtaining an accurate vaccination history by the ophthalmologist during this unprecedented pandemic and not to take this for granted. This case also stresses the need for urgent referrals by the non-ophthalmologists to the ophthalmology service more often if visual symptoms are experienced following COVID-19 vaccination in order to avoid wrong labelling of such presentation and reduce the chance of unnecessary investigations and inappropriate treatments to be given.

Ethics Statement

No authorization was required from the author's institution for the submission of this article for publication. This Study adheres to the Declaration of Helsinki and an informed consent was obtained from the patient.

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Conflict of Interest

There is no competing financial interest in relation to this article.

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Author Contribution Statement

MZK is the only individual involved in the writing of this article.

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