Paracentral acute middle maculopathy following SARS-CoV-2 infection: the D-dimer hypothesis

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DESCRIPTION

We herein report a case of bilateral acute-onset scotoma following SARS-CoV-2 infection in a 19-year-old girl attributed to the presence of paracentral acute middle maculopathy (PAMM). She was tested positive for SARS-CoV-2 infection 2 weeks prior by reverse transcription PCR testing and radiologically. Her medical reports revealed a raised D-dimer levels (1.23 μg/mL), for which she was started on intravenous steroids and enoxaparin by the treating physician. Her presenting visual acuity was 20/40, N6 and 20/125, N6 in the right (RE) and left eye (LE), respectively. Dilated fundus evaluation exhibited the presence of cotton wool spots along the vascular arcades along with subtle white lesions at macula bilaterally (figure 1A, B).

Optical coherence tomography (OCT) showed presence of focal hyperreflective change in the inner and outer plexiform layers (IPL, OPL) with inner nuclear layer (INL) volume loss parafoveally (figure 1C, D) features consistent with PAMM. On review after 4 weeks, her visual acuity improved to 20/20 in RE and 20/25 in LE, repeat D-dimer levels were normal. OCT showed thinning of INL with the irregularity of IPL and OPL along with the resolution of the cotton wool spots (figure 2A–D).

Sarraf et al1 first described PAMM to be a spectral-domain OCT manifestation of a thickened hyper-reflective band at the level of the OPL and INL.2 PAMM has been attributed to the acute phase of ischaemia of intermediate and deep capillary plexuses which may herald presence of underlying secondary conditions like retinal vascular diseases. D-dimer, commonly elevated in coronavirus disease 2019 (COVID-19) patients, is a fibrin-degradation product which is increased in thrombotic events, indicating fibrinolysis.3 Raised D-dimer values, lead to activation of coagulation cascade secondary to systemic inflammatory response syndrome, correlate to the disease severity and high mortality in such patients.4–6 Virgo and Mohamed7 interestingly reported two cases of PAMM and acute macular neuroretinopathy following SARS-CoV-2 infection, possibly under the umbrella of ‘paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 infection’.8–10

We hypothesise that the retinal capillary plexus ischaemia leading to PAMM could be secondary to a thrombotic milieu which is translated in the form of raised blood D-dimer levels. Routine oculovital fundus examination and ordering D-dimer assay in patients with COVID-19 presenting with PAMM is extremely crucial as prompt anticoagulation is mandated in these patients.11 12

In supposition, the relationship of D-dimer levels with PAMM in patients with COVID-19 warrants further attention before a meaningful conclusion can be drawn.

Figure 1 (A, B) Colour fundus picture of right and left eye, respectively, shows resolving perifoveal whitish lesions and cotton wool spots; (C, D) Swept source optical coherence tomography of right and left eye passing through macula shows reduced hyperreflectivity with the irregularity of inner and outer plexiform layers and thinning of inner nuclear layer parafoveally.

Figure 2 First month follow-up (A, B) Colour fundus picture of right and left eye, respectively, shows resolving perifoveal whitish lesions and cotton wool spots; (C, D) Swept source optical coherence tomography of right and left eye passing through macula shows reduced hyperreflectivity with the irregularity of inner and outer plexiform layers and thinning of inner nuclear layer parafoveally.

Patient’s perspective

I am thankful to my treating ophthalmologist that my disease could be diagnosed at an early stage and appropriate intervention was done for prevention of progression of the same.
Learning points

- Paracentral acute middle maculopathy (PAMM) and acute macular neuroretinopathy can occur following SARS-CoV-2 infection.
- Retinal capillary plexus ischaemia leading to PAMM could be secondary to a thrombotic milieu which is translated in the form of raised blood D-dimer levels.
- Routine ocular fundus examination should be done in these patients as prompt anticoagulation is mandated in patients presenting with PAMM.

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