CASE REPORT

Paracentral acute middle maculopathy: precursor to macular thinning in sickle cell retinopathy

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SUMMARY

We present a case of paracentral acute middle maculopathy (PAMM) in a patient with sickle cell disease (SCD). Though gradual capillary loss may contribute to pathophysiology of sickle cell retinopathy as well, our case suggests that PAMM may be the precursor lesion to macular thinning commonly observed in patients with SCD. In addition, fluorescein angiography may be unable to detect these acute vascular events occurring at the deep capillary plexus.

BACKGROUND

Atrophic ischaemic changes in the macula of patients with sickle cell retinopathy (SCR) has been described on optical coherence tomography (OCT) as macular splaying and macular thinning.¹ It is unknown whether these changes occur as a result of gradual capillary loss or by intermittent acute events. We recently witnessed a case of paracentral acute middle maculopathy (PAMM) in a patient with sickle cell disease that suggests that atrophic ischaemic changes in SCR follow acute vascular events occurring in the deep capillary plexus of the retina.

CASE PRESENTATION

A 14-year-old girl with known history of non-proliferative SCR presented with a 1-week history of central scotoma in her left eye. Visual acuity was 20/20 in the right eye and 20/25 in the left eye with eccentric fixation. Goldmann tonometry was within normal limit for both eyes. There was no relative afferent pupillary defect. Fundus exam showed a new white retinal wedge inferonasal to the fovea in the left eye (figure 1A). The rest of the exam was unremarkable.

INVESTIGATIONS

Fluorescein angiography did not show an obvious vessel occlusion (figure 1B). Macular OCT showed hyper-reflectivity of the inner retinal layers consistent with PAMM in the left eye (figure 1A). OCT angiography was not available to measure the retinal capillary plexus in this patient.

OUTCOME AND FOLLOW-UP

At follow-up visits, her central scotoma in the left eye persisted, and OCTs showed macular splaying and progressive thinning of the inner retinal layers corresponding to the previous PAMM lesion (figure 1C and D).

DISCUSSION

Brasileiro *et al*² proposed microvasculature occlusion as the common mechanism underlying both peripheral proliferative SCR and thinning of the macular inner retinal layers. It is unclear however whether these changes occur gradually or after acute events. Recent evidence suggests that acute vascular events at the deep capillary plexus may be responsible for macular thinning. Using OCT angiography, Han *et al*³ showed that inner retinal thinning in eyes with SCR is associated with abnormalities in the deep capillary plexus. We know that acute lesions, such as PAMM, can occur in patients with SCR due to ischaemia within the deep capillary plexus.⁴ It is suggested that the deep capillary plexus is especially susceptible to hypoxia given its higher oxygen demand, greater tissue to blood

Figure 1 (A) Fundus photo of the left eye shows an area of retinal whitening infero-nasal to the foveola corresponding to patient's scotoma. SDOCT thickness map shows retinal thickening in this area and pre-existing temporal retinal thinning. Red line represents SDOCT scan location in figure D (top) (B) Fluorescein angiogram at 22 s shows no obvious occlusion of the retinal vessels in the area of whitening. (C) Photo 2 months later in the left eye shows resolution of retinal whitening. Red line represents SDOCT scan location in figure D (bottom). (D) Top: SDOCT shows inner retinal thickening and hyper-reflectivity in the area of retinal whitening, consistent with PAMM lesion. Bottom: SDOCT in the area previously depicted now shows thinning of the inner retina, causing a blunting of the foveal contour (previously described as 'splaying'), with previously noted temporal retinal thinning. PAMM, paracentral acute middle maculopathy; SDOCT, spectral domain optical coherence tomography.

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ratio and a watershed location between the inner and outer retinal vascular supply.^{4 5} This case of PAMM in a patient with sickle cell disease suggests that the atrophic changes of macular splaying and thinning in SCR may occur as a result of acute ischaemic events.

Learning points

- ▶ With the advent of optical coherence tomography (OCT), macular thinning can be detected in patients with sickle cell disease (SCD).
- ▶ Acute vascular events occurring at the deep capillary plexus, such as paracentral acute middle maculopathy, contribute to macular thinning in SCD.
- ▶ Fluorescein angiography has low sensitivity to detect such events, whereas OCT angiography may prove to be a more useful tool in this clinical setting.

Contributors All authors made substantial contribution to the conception, design and revision of the draft. KS has followed the patient for a long time in her clinic and was responsible for obtaining the OCT images, FA as well as the consent form. PC and SAH reviewed all the clinical data, compiled it as a case report and reviewed the literature.

Competing interests None declared.

Patient consent We greatly value our patients' privacy. Though a consent could not be obtained, we have not included any personal health information or identifiable data in our report that can be traced to the patient. We ensured that all data were anonymised.

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