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Sir,

Peripapillary lesions causing blind spot enlargement in a case of multiple evanescent white dot syndrome

Multiple evanescent white dot syndrome (MEWDS) is a multifocal chorioretinal inflammatory disease that was first reported by Jampol *et al*. in 1984.[1] Their original case series described 11 young adults with a unilateral decrease in visual acuity. Ophthalmoscopic findings revealed white dots distributed at the level of deep retina and/or retinal pigment epithelial layer (RPE) – the site thought to be primarily affected in MEWDS. In addition there was macular granularity.

Since the original description many cases of MEWDS have been described[2–5] and the clinical picture better defined. Most cases are preceded by a viral illness, and symptoms of photopsia are common.[2] Vitritis, optic neuritis and enlargement of the physiological blind spot are also typical features. While the exact aetiology of MEWDS remains unknown, the cause of the enlarged blind spot has been the focus of several studies. Takeda *et al*.[6] suggested optic nerve dysfunction as the cause. Fletcher *et al*.[7] maintained that peripapillary retinal dysfunction was the mechanism after studying patients with blind spot enlargement and normal optic discs. The following case of MEWDS is unusual in that peripapillary lesions were demonstrable (and thought to be responsible for the enlarged blind spot) in addition to an undoubted optic neuritis.

Case report

A 34-year-old woman presented with a 2 week history of photopsia in the left visual field that was associated with reduced visual acuity and floaters in the left eye. Visual acuity in the right eye was 6/4 and in the left 6/36. A left relative afferent pupillary defect, red desaturation and blind spot enlargement on automated perimetry was present. Fundal examination revealed a vitritis, papillitis, and marked peripapillary nerve fibre layer swelling on the left, as well as multiple mid-peripheral white lesions at the level of the retinal pigment epithelium (RPE) (Fig. 1). Fundal fluorescein angiography (FFA) in the acute phase showed early hyperfluorescence and late staining of lesions typical of MEWDS. There was also extensive leakage of dye from the optic nerve head consistent with a papillitis, and no peripapillary lesions could be recognised (Fig. 2).

The patient was commenced on prednisolone 80 mg daily and 1 week later her visual acuity improved to 6/18. The left relative afferent defect was less obvious as were the chorioretinal lesions on fundoscopy. The

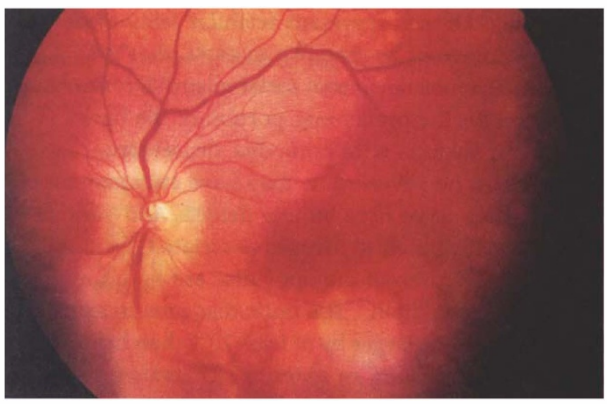

Fig. 1. *A fundus photograph of the left eye at presentation, showing swelling of the optic disc, and peripapillary nerve fibre swelling. Multiple mid-peripheral whitish lesions, best seen supero-temporally, are also present. (The circular pale area infero-temporal to the macula is artefactual.)*

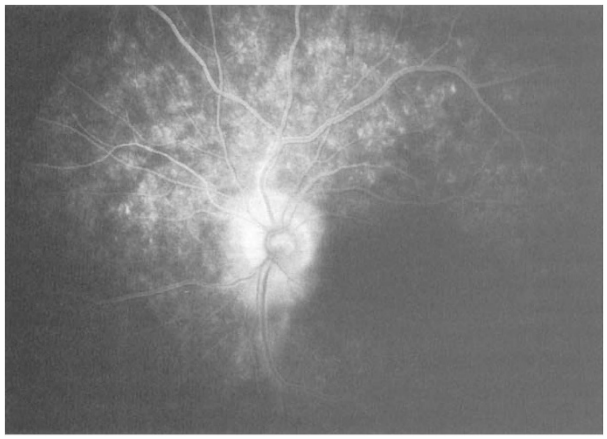

Fig. 2. *Fluorescein angiogram (at 11 min) showing extensive leakage from the optic nerve head, and widespread focal areas of hyperfluorescence.*

steroids were tailed off and 8 weeks later the visual acuity was 6/6 in the left eye with fewer subretinal white dots clinically. A repeat FFA revealed the presence of multiple lesions in a peripapillary distribution with clustering inferotemporal to the disc (Fig. 3). The enlarged blind spot was still present. Four months later all clinical evidence of MEWDS had resolved with the exception of the enlarged blind spot. The visual acuity was 6/5 and there was no evidence of optic nerve dysfunction on pattern visual evoked potential testing.

Discussion

The case discussed demonstrates clearly that MEWDS is not a disease confined to the RPE. The most marked clinical feature was in fact the papillitis and swelling of the peripapillary nerve fibre layer. Optic nerve dysfunction was present in the acute stage as verified by the relative afferent pupillary defect and colour

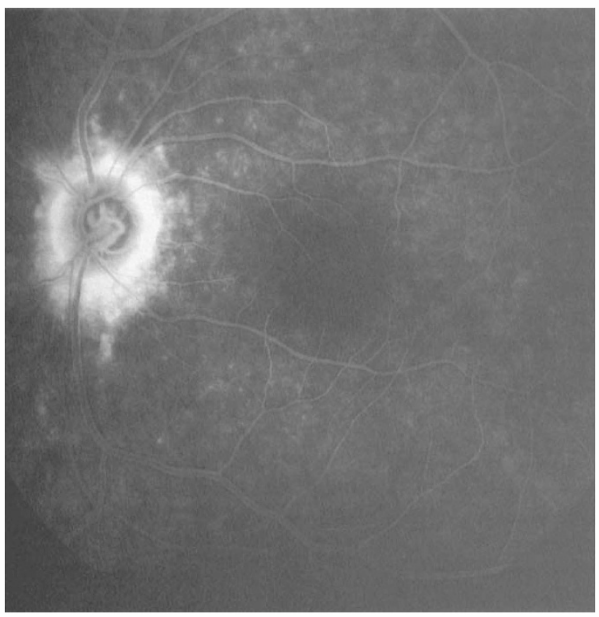

Fig. 3. *Fluorescein angiogram at 8 weeks after the acute presentation, showing multiple lesions in a peripapillary distribution, particularly infero-temporal to the optic disc.*

desaturation. However, the enlarged blind spot persisted after the optic nerve had recovered, suggesting a mechanism other than optic nerve dysfunction as the cause.

Electroretinogram (ERG) studies have confirmed RPE and photoreceptor dysfunction in MEWDS,[3] and in our patient the peripapillary lesions could adequately explain the enlarged blind spot. Jampol[4] has commented on the fact that the enlarged blind spot in MEWDS is absolute and has steep margins (as has the blind spot in our case) and this coupled with electrophysiological studies suggests that enlargement of the blind spot is due to peripapillary retinal dysfunction rather than optic nerve dysfunction.

It is not clear why the enlarged blind spot should persist when the RPE lesions have resolved and acuity has returned to normal. Impaired visual acuity in MEWDS may reflect a global dysfunction of the RPE and hence sensory retina also. Takeda *et al.*[6] found in their MEWDS case that the ERG a-wave and early receptor potential were decreased in amplitude, demonstrating the functional dependence of the sensory retina on the underlying RPE. When the RPE cells recover (partially or completely) there is no longer a generalised depression of sensory retinal function and visual acuity therefore returns to normal. However, the majority of the RPE that is affected lies mid-peripherally in MEWDS, outside the zones of high cone and neural bundle concentration, and residual focal RPE dysfunction in these regions therefore is unlikely to manifest clinically. However, if the lesions are concentrated in the peripapillary region, then one could postulate that in such a visually important area focal, mild impairment of the RPE might be enough to cause an enlarged blind spot, in the absence of reduced visual acuity or abnormal clinical findings. Normally the blind spot eventually recovers when RPE function has returned completely (70% of the cases in Reddy *et al.*'s study had follow-up visual fields and all had normalised by 6 to 47 months).[5]

Borruat *et al.*[8] have described a similar case to ours in which indocyanine green angiography (ICGA) detected an area of peripapillary hypofluorescence that they believed could explain the enlarged blind spot. They concluded that MEWDS is primarily a choroidopathy, in which only the significant lesions manifest funduscopically or on FFA by causing outer retinal involvement and hence dysfunction. An initial ICGA may have shown this peripapillary zone in our case, but the florid papillitis could have confounded the findings as a result of choroidal masking (Borruat's case only had mild disc oedema). Hahmed *et al.*[9] and Singh *et al.*[10] have also described juxtapapillary lesions in patients with MEWDS. However, in their cases optic disc oedema was not present.

Recently (1997) a MEWDS patient presented with a circumpapillary chorioretinal lesion that resolved clinically and angiographically (FFA) before the development of the classical funduscopic findings of MEWDS.[11] This case suggests a pattern of lesion evolution with MEWDS. Our case may represent lesions

found in the interval between such an initial circumpapillary plaque and the later typical MEWDS manifestations. The peripapillary retinal lesions in our case may be an extension of 'significant' peripapillary choroidal lesions or resolution of the peripapillary plaque-like lesion.

We feel two conclusions can be drawn from this case. Firstly, the peripapillary lesions are the likely cause of the enlarged blind spot. Secondly, the marked peripapillary nerve fibre swelling may be secondary to the underlying RPE/photoreceptor inflammation and subsequent retrograde spread of the inflammation to the optic nerve may cause or aggravate the optic neuritis. Choroidal inflammation could be the primary source of spread to the optic nerve. The peripapillary lesions in our case were found only after the papillitis had settled, and the unmasking of these lesions is most obvious in the late phases of the fluorescein angiogram. Perhaps more cases of this type will be discovered if repeat FFA is performed in the recovering phase of MEWDS patients with enlarged blind spots. There may also be a role for focal ERG testing to quantitate the degree of peripapillary RPE dysfunction and correlate this with the progress of the blind spot enlargement. It is clear that the morphology of these lesions depends as much on the timing as the type of angiographic investigation performed – making the term 'evanescent' most appropriate.

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