Purtscher-like retinopathy in anti-MDA5 dermatomyositis: a window to underlying microvasculopathy

Sirs,

We report a case of a 36-year-old man presenting with acute painless visual impairment following a diagnosis of anti-melanoma differentiation-associated protein 5 dermatomyositis (anti-MDA5 DM). The patient presented with marked dyspnoea and classic cutaneous findings of dermatomyositis. Serological testing for Anti-MDA5 antibodies confirmed the diagnosis. Initial treatment comprised of pulse methylprednisolone and rituximab, with oral prednisolone on discharge. Three weeks after treatment commencement, painless blurred vision developed in his left eye. Best-corrected visual acuity (BCVA) was 20/20 (right) and 20/80 (left) respectively. Anterior segment examination was normal. Fundoscopy revealed peripapillary cotton-wool spots (CWS) around the posterior poles bilaterally (Fig. 1-AB). There was no abnormality on fluorescein angiogram or optic disc swelling. Optical coherence tomography (OCT) demonstrated bilateral macular oedema (Fig. 1-CD). A clinical diagnosis of Purtscher-like retinopathy (PLR) was made.

Further deterioration in visual acuity (BCVA 20/30 on right and 20/200 on left) prompted another course of pulse methylprednisolone, and commencement of low-dose aspirin. Fundoscopy demonstrated increasing size and number of CWS in the left eye, with new involvement of the papillomacular bundle. Repeat OCT showed improving bilateral macular oedema. Fluorescein angiography and OCT angiography again demonstrated no evidence of macular ischaemia or retinal vasculitis. Mycophenolate, hydroxychloroquine, and monthly intravenous immunoglobulin were commenced in addition to oral prednisolone. Six weeks after initial visual loss, BCVA improved to 20/40 (left) and remained stable at 20/30 (right). By three months, vision improved to 20/20 bilaterally.

Anti-MDA5 DM is a subset of dermatomyositis most notable for its amyopathic presentation and rapidly progressive interstitial lung disease. A range of inflammatory ocular processes have been described in DM (1-3). Retinal involvement in DM is not uncommon, and the degree of vision loss will depend on the target vessels involved and subsequent extent of ischaemia (2, 4).

Purtscher-like retinopathy (PLR) is a complement-mediated occlusive microangiopathy with characteristic fundoscopic signs of CWS, retinal haemorrhages, and confluent areas of ischaemic retinal whitening termed 'Purtscher flecken' (5). The pathology is at the pre-capillary arteriolar level where occlusion leads to inner retinal oncosis, manifesting as polygonal areas of retinal whitening. 'Purtscher flecken' are pathognomonic for PLR, but are only seen in 50% of patients (6). PLR has been rarely reported in DM, but not specifically in anti-MDA5 DM (7, 8).

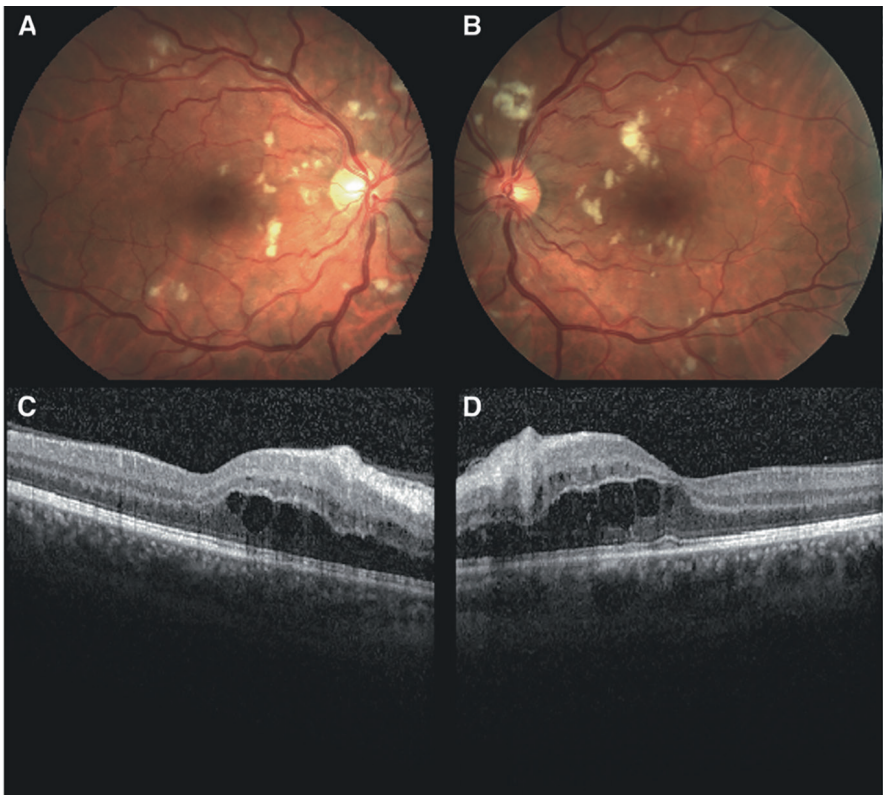

Fig. 1. Fundus photography and optical coherence tomography (OCT).
A and **B:** Photographs of right and left fundi, respectively showing multiple peripapillary and posterior pole cotton wool spots.
C and **D**: OCT of the right and left maculae respectively, showing bilateral macular oedema.

Like 'Purtscher flecken', CWS are makers of retinal arteriolar ischaemia. However, histologically they reflect accumulation of debris in the innermost nerve fibre layer due to interruption of axoplasmic flow (9). CWS tend to persist for up to 6 weeks, whereas whitening of 'Purtscher flecken' retinal oncosis can fade after 7-14 days (9). Strictly speaking, PLR and retinal vasculitis are different retinal manifestations. In retinal vasculitis, pathology can occur at the level of the arterioles or venules. It is associated with fundoscopic evidence of perivascular sheathing or cuffing, best identified with fluorescein angiography. In earlier reports of retinal involvement in DM, the distinction between retinal vasculitis and other forms of retinopathy was not well-made.

It is relevant to consider the shared mechanism of injury underlying the cutaneous and retinal manifestations in anti-MDA5 DM. Vasculopathy is a prominent feature of the disease (10), and histopathology of cutaneous lesions reflects microvascular endothelial injury (11). Aberrant complement activation and presence of endotheliitis have also been associated with cutaneous ulcers (12). Increased complement C5b-9 deposition on endothelial cells is observed in thrombotic microangiopathies, also seen in DM (13). An upregulated Type 1 interferon gene signature can drive a number of these pathological processes (14).

This is the first described case of PLR in anti-MDA5 DM, demonstrating the prominent microvasculopathy associated with the disease. Retinal involvement may reflect the extent of systemic pathology, and early recognition is important. Understanding potential pathogenic pathways may shed light on therapeutic options.

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