CASE REPORT

Systemic Lupus Erythematosus Presenting as Orbital Myositis

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ABSTRACT

The authors present a case of diplopia and eye pain due to orbital myositis in a patient with a de novo diagnosis of systemic lupus erythematosus. Systemic lupus erythematosus is a rare cause of orbital myositis and should be considered when other, more common, conditions have been excluded.

Keywords: Diplopia, orbital myositis, systemic lupus erythematosus

INTRODUCTION

We present a case of diplopia and eye pain due to orbital myositis in a patient with a de novo diagnosis of systemic lupus erythematosus.

CASE REPORT

A 33-year-old Nigerian lady presented with left eye pain, swelling, and horizontal binocular diplopia on looking to the left. She worked as a psychiatric nurse and 5 days previously had been struck just above the left eye with a stick by one of her patients. Since that time she had developed a severe headache, with the localised eye symptoms developing over the following few days.

She had a past medical history of a spontaneous subarachnoid haemorrhage 6 months prior to this, secondary to a left middle cerebral artery aneurysm that was treated with endovascular coiling.

On admission she had left peri-orbital swelling with a red eye (mild conjunctival injection). She had normal visual acuity, visual fields, colour vision,

pupillary function, and retinal examination on fundoscopy. She had impairment of right eye adduction but otherwise normal eye movements (Figure 1). She reported pain around the left eye that was exacerbated by movement. There was no proptosis or ptosis.

Routine blood tests revealed a microcytic anaemia (and subsequently sickle cell trait), mild lymphopaenia ($0.9 \times 10^9 \text{ L}^{-1}$; range: 1×10^9 to $4 \times 10^9 \text{ L}^{-1}$), renal impairment with a creatinine of $132 \mu\text{mol L}^{-1}$ (range: $60\text{--}110 \mu\text{mol L}^{-1}$), slightly elevated C-reactive protein at 12 mg L^{-1} (normal $<10 \text{ mg L}^{-1}$), and raised erythrocyte sedimentation rate of 43 mm h^{-1} , but normal liver function tests, electrolytes, thyroid function, thyroid antibodies, creatine kinase, and clotting screen. She was initially treated for orbital cellulitis and admitted for further investigations.

Computed tomography (CT) imaging of her brain and orbits identified enlargement of the right medial rectus and left inferior rectus muscles (Figure 2), with evidence of involvement of the tendinous insertion of the right medial rectus on axial slices (Figure 3); findings that are consistent with a focal myositis. Further, more specific, blood tests went on to reveal a

FIGURE 1 Photographs demonstrating impairment of right eye adduction on presentation.

FIGURE 2 Computed tomography (CT) imaging of orbits demonstrating enlargement of the right medial rectus (thin arrow) and left inferior rectus muscles (arrow head). There is an incidental finding of an old fracture through the medial wall of the left orbit (seen on previous imaging 6 months before this episode).

positive antinuclear antigen, a positive *Crithidia* immunofluorescence test, with strongly positive anti-dsDNA (23.0 IU/mL [normal range: 0.0–9.9 IU/mL]), SSA and SSB autoantibodies, and reduced C3 and C4 levels (C3: 0.47 g/L [normal range: 0.90–1.80 g/L], C4: 0.07 g/L [normal range: 0.10–0.40 g/L]). These indicated an underlying diagnosis of systemic lupus erythematosus. A renal biopsy demonstrated class 4 lupus nephritis with focal necrotising lesions with C1q positivity on

FIGURE 3 Axial CT slices demonstrating enlargement of the right medial rectus with involvement of the tendinous insertion (arrow).

immunohistochemical stains (Figures 4 and 5). Electron microscopy showed segmental subepithelial electron-dense deposits.

She was treated with high-dose steroids and mycophenolate, and the diplopia and extraocular eye muscle swelling resolved. By follow-up, 2 months later, her eye movements had returned to normal (Figure 6), and a follow-up magnetic resonance imaging (MRI) scan showed resolution of the swelling of the extraocular muscles (Figure 7).

DISCUSSION

Orbital myositis is a rare form of orbital inflammatory syndrome (previously known as orbital

pseudotumour).¹ All forms of orbital inflammatory syndrome have been described in relation to systemic lupus erythematosus,² with orbital myositis being recognised as early as 1983³ and further reported in subsequent cases.^{4,5} In this case, however, the patient presented with an acute onset of painful diplopia and

FIGURE 4 Haematoxylin and eosin stain of a single glomerulus, showing fibrinoid necrosis (arrow) in one segment and early crescentic nephritis in evolution.

FIGURE 5 Immunohistochemical stain showing granular C1q positivity in capillary loops and the mesangia. (C3, IgG, IgM, and IgA stains were also positive in other stains.).

without a known diagnosis of systemic lupus erythematosus. Systemic inflammatory processes should therefore be borne in mind when assessing the possible causes of a localised orbital myositis. Our patient presented with pain in the left eye that was exacerbated by movement and a painless right eye with restricted movement. The imaging demonstrated involvement of the extraocular muscles in both eyes, hence demonstrating a bilateral process that manifested as pain on eye movement in one eye and restricted movements in the other. Contrast-enhanced orbital MRI is the most sensitive imaging modality and can help determine the underlying diagnosis. As in our case, orbital myositis classically involves the entire length of the extraocular muscle including the tendon, as opposed to thyroid orbitopathy where the tendons are usually spared.⁶ In this case, the recent trauma was most probably

FIGURE 7 Magnetic resonance imaging of orbits demonstrating improvement in swelling of extraocular muscles.

FIGURE 6 Photographs demonstrating normal eye movements following treatment.

unrelated given the clinical and radiographic evidence of bilateral involvement. Localised swelling and conjunctival injection are common symptoms of orbital myositis¹ and, in this case, were unrelated to the trauma.

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Note: Figure 1, 4, 5, and 6 are available in colour online at <http://informahealthcare.com/oph>.

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