

Supplemental table. Clinical course of patients with idiopathic orbital inflammation emerging de novo from other ocular adnexa

| Patient number | Clinical course | Clinical characteristics and duration between initial to last onsets |
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| 1 | <p>Patient 1 initially had orbital myositis in IO, LR, and SO muscles on the left eye, which was recalcitrant to following steroid pulse therapy. Then, migrations and recurrence of myositis emerged; first: SR, LR, and SO muscles on the left eye, second: SO muscles on the right eye, third: right LR, MR, and IR muscles, on the right eye. The fourth affected muscle was IO muscle on the right eye after 120 months from initial onset. The patient had no novel inflammatory lesions during 72 months of follow-up following the fourth migration.</p> | <p>During 120 months, fat suppressed MR imaging exhibited four times migrations of inflammatory extraocular muscles to bilateral eyes. (Reference 13)</p> |
| 2 | <p>Patient 2 suddenly noted headache, diplopia, left periocular pain, and swelling before 10 days of initial visits. CT scan showed LR muscle enlargement on the left eye. Steroid pulse therapy dramatically responded, but bilateral orbital myositis with SR, IR, MR, and LR muscle enlargement emerged during maintenance 5 mg prednisolone. Following prednisolone at 0.4 mg/Kg/day with a slow taper, the patient achieved complete remission.</p> | <p>Orbital myositis recurred in a more severe condition after 41 days from the initial onset and during maintenance low dose corticosteroids following steroid pulse treatment. This case illustrates that previous corticosteroids may not prevent inflammatory ocular adnexal lesions. (Reference 13)</p> |
| 3 | <p>Patient 3, who suddenly noticed left lid swelling and diplopia, was referred to our hospital after 10 months from initial onset. CT scan and fat suppressed T2-weighted MR imaging showed poorly enlargements but hyperintense in SR and IR muscles on the left eye. The patient was recalcitrant to steroid pulse therapy. In addition, migrations of inflammation emerged: second: MR and SO muscles on the left eye, third: MR muscle on the right eye. The fourth affected muscle was IR muscle on the right eye after 82 months from initial onset. After the fourth migration of inflammation, the patient had no novel inflammatory lesions during 72 months of follow-up.</p> | <p>During 82 months, fat suppressed MR imaging exhibited migrations of inflammatory extraocular muscles to bilateral eyes four times.</p> |

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| 4 | <p>Patient 4 suddenly noticed ocular pain and blurred vision (corrected visual acuity; 6/15) on the right eye 7 days ago. Clinical examinations showed optic perineuritis on the right eye. Prednisolone at 0.4 mg/kg/day with a slow taper was started and ocular pain and blurred vision improved completely, but suddenly noticed ocular pain and blurred vision (corrected visual acuity; 6/15) on the left eye after 137 days at initial visits. Following the diagnosis of optic perineuritis on the left eye, prednisolone at 0.4 mg/kg/day with slow taper was started. The patient achieved complete remission and did not relapse during 48 months of follow-up after discontinuing corticosteroids.</p> | <p>Bilateral optic perineuritis emerged at different times (144 days).</p> |
| 5 | <p>Patient 5 suddenly noted an inflamed eye and blurred vision in the left eye. Scleritis with serous detachment on the left eye was revealed. After prednisolone at 0.6 mg/kg/day with slow taper, the patient achieved complete remission, but scleritis on the right eye emerged. Prednisolone at 0.4 mg/kg/day with slow taper was re-started and the patient achieved complete remission. Further, scleritis on the left eye recurred 12 months after initial visits. Prednisolone at 0.4 mg/kg/day with slow taper was started again. The patient achieved complete remission on bilateral lesions and didn't relapse for 12 months after discontinuing the treatment.</p> | <p>Bilateral posterior scleritis emerged at different times (173 days). In addition, this case illustrates that previous corticosteroids may not prevent inflammatory ocular adnexal lesions.</p> |
| 6 | <p>Patient 6 had suddenly left chemosis, upper eyelid swelling, periocular pain, and ptosis four days before the initial visit. CT scan showed diffuse inflammatory lesion around the left eye. Complete remission was achieved after prednisolone at 0.4 mg/kg/day with a slow taper. However, seven days after discontinuing the treatment, the patient noticed a worse condition in the right eye. CT scan revealed a diffuse lesion around the right eye.</p> | <p>Bilateral diffuse lesions around the eyes (possible scleritis) at different times (77 days). This case also illustrates that previous corticosteroids may not prevent inflammatory ocular adnexal lesions.</p> |

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| 7 | Patient 7 suddenly noticed lid swelling, ocular pain in the left eye and diplopia 70 days ago. At the initial visit, Fat suppressed T2-weighted MR imaging findings revealed high signal intensity in the fascial structure of IR and LR muscles on the left eye. Complete remission was achieved after prednisolone at 0.4 mg/kg/day with a slow taper. However, the patient noticed ocular pain and visual field defect on the left eye and diplopia again. Fat suppressed T2-weighted MR imaging findings revealed high signal intensity around the optic nerve, unlike the first fat-suppressed T2-weighted MR imaging. | Migration of inflammation on ipsilateral ocular adnexal lesions at different times after 252 days. |
| 8 | Patient 8 suddenly noticed chemosis, lid swelling, ocular pain, and blurred vision in the right eye 4 days ago. Optic perineuritis on the right eye was diagnosed and complete remission was achieved after prednisolone at 0.2 mg/kg/day for 7 days. However, orbital myositis with MR muscle enlargement on the left eye emerged after 37 days from the initial onset and the lesion achieved spontaneous regression. | Optic perineuritis and orbital myositis emerged contralaterally at different times respectively (41 days). |

Abbreviation CT scan; computed tomographic scan, MR imaging; magnetic resonance imaging, SR; superior rectus, IR; inferior rectus, MR; medial rectus, LR; lateral rectus, SO; superior oblique, IO; inferior oblique