

Dacryocystitis and orbital pseudotumor in a patient with Granulomatosis with polyangiitis

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A 38-year-old male presented with decreased vision and diplopia in his right eye over the past month. On performing an examination, he had proptosis, periorbital puffiness, dilated episcleral vessels, sluggish pupillary reflex, external ophthalmoplegia, and diminished vision. He also had left dacryocystitis (Figure 1a). He had presented six years ago with bilateral hearing loss, recurrent sinusitis, and hemoptysis and was diagnosed with GPA (anti-PR3 titer >100 units/mL and a necrotizing granuloma in the nasal biopsy). He was initially treated with steroids and intravenous cyclophosphamide that were discontinued after the second dose as he developed pneumonia twice; he was subsequently treated with mycophenolate mofetil, followed by azathioprine and steroids. His current anti-PR3 titer was <3 units/mL. An MRI orbit showed an ill-defined T1/T2 isointense-to-hypointense lesion (pseudotumor) replacing the orbital fat that diffusely involved intra- and extraconal compartments (Figure 1b, arrows). The extraocular muscles were encased by the lesion and appeared bulky, with mild flattening of the posterior globe (Figure 1c, arrows). The lesion encased the optic nerve and mildly compressed the optic nerve in the orbital apex. The patient was administered three doses of methylprednisolone pulses and two doses of rituximab (500 mg) two weeks apart. He responded well to treatment; he showed resolution of dacryocystitis, periorbital puffiness (Figure 1d), and diplopia and improvement in his vision. GPA has a wide spectrum of orbital manifestations, which can occur in up to 52% of patients. Patients with GPA



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Figure 1. a-d. Right eye proptosis, periorbital puffiness, and dilated episcleral vessels with left dacryocystitis (a) T1/T2 MR image showing a isointense-to-hypointense lesion (pseudotumor) replacing the orbital fat that diffusely involved intra- and extraconal compartments, with mild flattening of the posterior globe (b, c) Resolution of dacryocystitis and periorbital puffiness (d)

may develop conjunctivitis, corneal ulceration, episcleritis/scleritis, optic neuropathy, retinal vasculitis, and uveitis. In addition, a retro-orbital pseudotumor and nasolacrimal duct obstruction may occur (1) (2).

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