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More than meets the eye

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A 28-year-old man presented with a 4-month history of a prednisolone-sensitive, painful and progressive right-sided exophthalmos and consecutive diplopia. On examination, the ophthalmologist suspected a pseudotumour of the right eye socket with lid closure defect, hypertropia and exotropia and protrusion bulbi with an intraocular pressure of 21 mm Hg (range; 10–21 mm Hg) and no signs of compressive optic neuropathy (cup-to-disc ratio of 0.4; ≤ 0.7) (figure 1a). Antiproteinase-3-antibodies were found both by positive immunofluorescence and by ELISA (165 U/mL; <10 U/mL). An MRI with a gadolinium-based contrast agent identified a destructive mass within the right eye socket (figure 1b). A subsequent debulking and biopsy confirmed small-vessel vasculitis (figure 1c) and ruled out IgG4-related disease, sarcoidosis, tuberculosis, orbital cellulitis, thyroid-associated orbitopathy or rhabdomyosarcoma as differential diagnoses of orbital pseudotumour.

History and physical examination were otherwise unremarkable with no signs of sinusitis, nose bleeds or stuffiness. Creatinine and urine analysis showed no evidence of renal involvement, and chest CT and pulmonary function tests showed no evidence of pulmonary involvement. Hence, we confirmed this unilateral ocular manifestation to be an isolated initial manifestation of a granulomatosis with polyangiitis. A course of high-dose methylprednisone and rituximab infusions (500 mg every 6 months) resulted in a sustained reduction in the size of the pseudotumour and a clinical improvement of the patient's diplopia, pain and lid closure defect over the follow-up of 12 months.

Collaborators Not applicable.

Contributors ES, GK and RB took over the clinical care. WS is the neuropathologist who contributed the histology and DSK is the cotreatment ophthalmologist. All authors were involved in the correction and improvement of the manuscript. RB is the guarantor.

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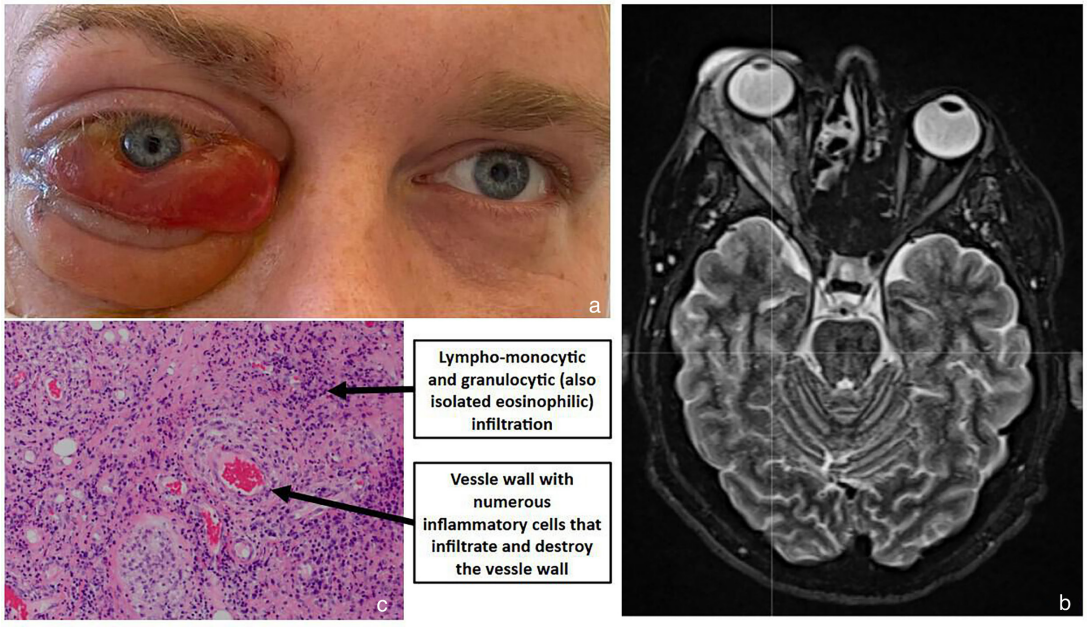

Figure 1 Clinical findings with (a) pseudotumour of the right orbit with lid closure defect, hypertropia and exotropia and protrusion bulbi, (b) corresponding MRI examination with a gadolinium-based contrast agent and (c) histologically proven small-vessel vasculitis.

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