Unilateral proptosis in a woman with asthma

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DESCRIPTION

A 47-year-old woman with a history of bronchial asthma presented with progressive proptosis of the right eye and diplopia. Her right eye showed apparent exophthalmos, impaired movement, conjunctival hyperaemia and episcleritis. Laboratory analysis revealed an increased titre of MPO-ANCA (4.3 IU/mL) and hypereosinophilia (1900 cells/μL, 22.5% on white cell count). MRI showed significant swelling of the extraocular muscles of the right eye and right ethmoid sinusitis (figure 1). Since a biopsy of extraocular muscles is unfeasible due to a risk of complications, the right bulbar conjunctiva was biopsied and dense infiltration of eosinophils with a granuloma formation (figure 2, arrows) was found. There were several phagocytised Charcot-Leyden crystals (figure 2, arrowheads) that were created by the destruction of eosinophils and release of their inner granules. The patient was diagnosed with eosinophilic granulomatosis with polyangiitis (EGPA) based on this histology, along with asthma, sinusitis and eosinophilia. It was speculated that unilateral proptosis was caused by the eosinophilic orbital myositis. Thirty milligrams of prednisone dramatically improved her proptosis and conjunctivitis.

Orbital myositis is an inflammatory process that involves the extraocular muscles. The typical symptoms are orbital pain, ocular movement impairment, diplopia and proptosis. Orbital myositis is mainly idiopathic but sometimes caused by infection, thyroid dysfunction or systemic inflammatory diseases including antineutrophil cytoplasmic antibody-associated vasculitis. However, the prevalence of orbital manifestations in EGPA is much lower than that in granulomatosis with polyangiitis. Our case demonstrates the diagnostic utility of bulbar conjunctival biopsy in such a major diagnostic challenge.

Learning points

▸ Orbital myositis is sometimes caused by systemic diseases including infection, thyroid dysfunction or systemic inflammatory diseases such as antineutrophil cytoplasmic antibody-associated vasculitis.

▸ Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare but important differential diagnosis in a patient with orbital myositis.

▸ Biopsy of bulbar conjunctiva is a useful diagnostic technique in EGPA with an eye presentation.

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REFERENCES


