Giant cell arteritis mimicking ocular myasthenia gravis

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Introduction: Giant cell arteritis (GCA) is infrequently reported as a cause of acute isolated monolateral third nerve palsy.

Case report: A 77-year-old man who complained of 4-5 days of left palpebral ptosis and one day of low-grade fever. No orbital pain was reported. Neurological examination revealed fatigable left eye ptosis and diplopia after 5 seconds of ocular fixation, as well as partial left third nerve palsy with moderate adduction and depression deficits. Pupils were equal and reactive to light, limb or bulbar weakness/fatigability and dyspnoea were absent. Routine haematological investigations showed raised erythrocyte sedimentation rate (ESR) 71 mm/h (nv <30) and C-reactive protein (CRP) 11.10 mg/dl (nv <1). Repetitive nerve stimulation (RNS) of the left facial nerve at 3 Hz did not yield a decremental response nor an incremental response at 20 Hz. Brain CT scan was normal. MG was suspected and therapy with pyridostigmine was started with partial improvement. In the following days diplopia disappeared and left palpebral ptosis was less evident. AChR-Ab turned out within the normal limits. Single-fibre electromyography (SFEMG) of the orbicularis oculi was negative for increasing jitter. In a clinical reassessment thickened left temporal artery with reduced pulsation was noted: for this reason, because GCA was suspected, a temporal artery biopsy was performed. Pending biopsy results, the patient was started on steroid treatment. Histological examination of temporal artery confirms Horton's disease. 3 months later, neurological examination was normal and no thickening of temporal artery was present.

Discussion: Ophthalmologic symptoms are common in GCA, appearing in 15–70% of patients, and may occur without other systemic symptoms in about 5–40% of cases. The most frequent ocular symptoms are ocular pain, monocular or binocular vision loss and diplopia. Co-occurrence of MG and GCA is extremely rare. Despite fluctuating symptoms and partial response to anticholinesterase treatment, we did not find any neurophysiological or serological data supporting the diagnosis of ocular MG. A correct diagnosis of GCA was delayed for cloudy symptoms, thankfully without any negative consequences. Prompt initiation of corticosteroid therapy actually ensures the minimization of the risk of irreversible vision loss.

Conclusion: We emphasize that in elderly patients presenting third cranial nerve palsy and/or apparently ocular myasthenic symptoms together with increased inflammatory marker levels and/or fever, a possible diagnosis of GCA should be considered. Temporal artery biopsy, the gold standard diagnostic test for GCA, should be mandatory.

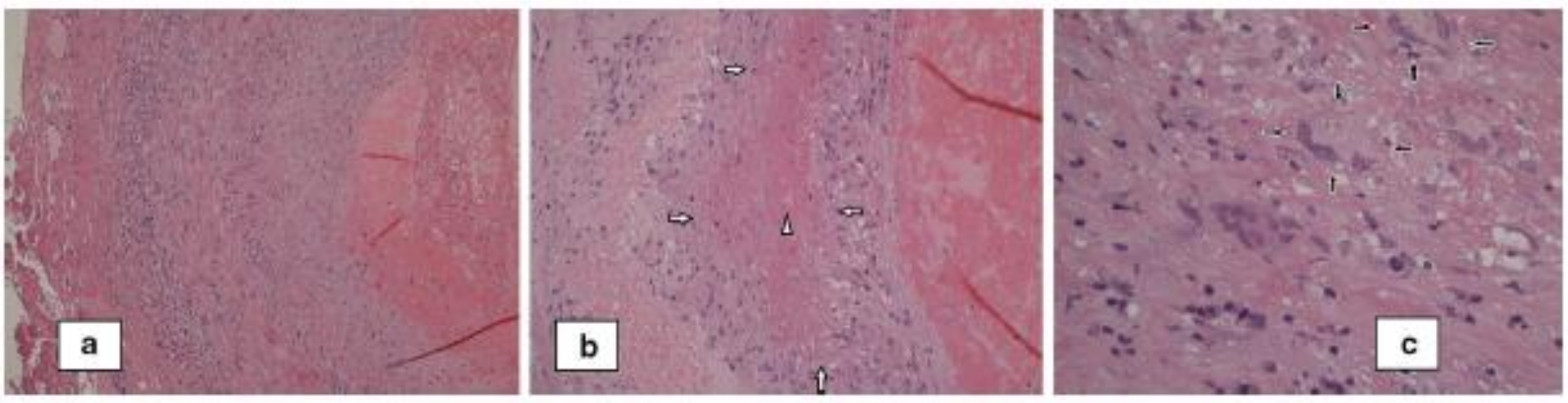

Fig. 1 **a** Arterial wall with the presence of lymphocytic inflammatory infiltrate, most evident in the adventitia (Hematoxylin–Eosin ×20); **b** granulomatous inflammation (*white arrows*) with central fibrinous area (*white arrowhead*), inflammatory lymphoplasmacytic and epithelioid cells (Hematoxylin–Eosin ×40); **c** granulomatous inflammation with multinucleated giant cells (*black arrows*) (Hematoxylin–Eosin ×60)

References:
Kale N, Eggenberger E (2010) Diagnosis and management of giant cell arteritis: a review. Curr Opin Ophthalmol 21:417-422
Liozon E, Loustaud-Ratti V, Soria P, et al (2004) Disease associations in 250 patients with temporal (giant cell) arteritis. Presse Medicale 33:1304–1312
Kertya E, Elsais A, Argov Z, et al (2014) EFNS/ENS Guidelines for the treatment of ocular myasthenia. Eur J Neurol 2:687–693