

# The missing picture: blindness in giant cell arteritis

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In the era of evidence-based medicine there are key facts that need to be established in order to form decisions about appropriate treatment. These facts include the risk from both the disease being treated and the proposed treatment. It is perhaps surprising how much of this key information is lacking for the most common form of vasculitis.

Giant cell arteritis (GCA) is the most common form of large-vessel vasculitis, affecting an estimated 2 individuals over 50 years of age per 10,000 per year (1). The eye is at particular risk in this disease. Irreversible visual loss, one of the most well-known and devastating complications of GCA, is reported to occur in 23% of cases in hospital series (2). To date, information on the occurrence of eye disease in GCA has been based almost exclusively on small hospital-based patient series (3, 4). The most common reported outcome is of visual loss. Permanent visual loss occurs in 12% to 32% of patients (5, 6). Visual loss is reported in a number of ways, including transient or permanent, monocular or biocular involvement. Definitions of severity of visual loss are also equally muddled with visual loss reported as reduced visual acuity or as loss of visual field. Furthermore the absence of control groups for these studies results in a lack of relative and absolute risk estimates for visual loss in patients with GCA. In addition, risk for eye complications appears to differ in subsets of patients with GCA, with those with isolated aortitis proving a particular diagnostic challenge (7-9).

There are no accurate data on the prevalence and nature of eye complications among patients in the community. Patients with GCA may be exclusively managed in the primary care setting without referral for either temporal artery biopsy or ophthalmic department examination. Currently, the incidence

and prevalence of eye complications within this group are unknown.

Moreover there have been no comprehensive assessments of the full range of ocular morbidity in patients with GCA due to both the disease process and treatment with glucocorticoids. The focus to date has been on the disease process itself rather than any iatrogenic associated outcome due to long term exposure with glucocorticoids.

The most common ocular pathology from hospital case series analysis is anterior ischaemic optic neuropathy (AION). Central arterial or branch retinal occlusion (CRAO), choroidal ischaemia and posterior optic neuropathy (PION) are also reported (10). In a prospective case series of 170 patients with biopsy proven GCA, 69 had AION (40%), 12 had CRAO (7%) and 6 had PION (3%). Of these 170 patients, 55 had fluorescein angiography and 12 of these had cilioretinal artery occlusion (21%). Nearly all the patients who underwent fluorescein angiography had evidence of posterior ciliary artery occlusion (10). In another retrospective study of 47 patients with GCA who had fluorescein angiography, 22 had AION (47%), 17 had choroidal ischaemia (36%) and 7 had CRAO (15%) (11).

The use of MRI has revealed further changes affecting the eye. A study of 43 patients with GCA using high resolution MR imaging of the ophthalmic artery demonstrated that 20 (46%) of patients had mural enhancement suggesting inflammation. Of the 43 patients, 11 had their MRI scan prior to treatment with glucocorticoids, but there was no correlation between treatment prior to the scan and mural enhancement. Ophthalmic examination comprising of fundoscopy in the 20 patients with mural enhancement revealed fundal changes in seven patients. Changes seen on fundoscopy included AION and CRAO. In

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fifteen patients the MRI and ophthalmic examination were normal. Nine patients had AION (20%) 4 CRAO (9%) and 1 PION (2%). However, in nine patients the MRI showed vessel inflammation but the patients had no symptoms of visual disturbance, and another eight participants were MRI-negative but had typical changes of arteritis on ophthalmic examination. The authors suggest that the differences in agreement between symptoms, ophthalmic examination and MRI may be due to the imaging protocol used in their study. They could only detect inflammation of the ophthalmic artery and not the small posterior ciliary arteries. The utility and significance of MRI changes requires further evaluation (12).

Treatment for GCA is also a risk to ocular health, with the currently accepted treatment for GCA involving the use of high doses of glucocorticoids with consequent risk of glaucoma or cataract development. However, glucocorticoid treatment regimens themselves are unstandardised and as yet no steroid-sparing agents have been found to be more effective (13). Courses of glucocorticoid treatment are often prolonged (possibly because of clinicians' overriding concern to prevent the onset of AION) increasing the risk of cataract, diabetes and glaucoma.

There are no accurate data on the incidence and significance of subclinical ocular involvement; or on the risk of progression of eye disease over time; neither has the relative frequency of eye morbidity related to treatment been evaluated. As a result, it is difficult to gauge the true risk of eye complications in GCA; the factors that predict the onset and progression of eye disease in GCA are unknown, and there is little understanding of how best to monitor patients over time for the development of eye disease.

In conclusion, there is an urgent need to develop an agreed objective assessment of visual loss in GCA to permit determination of the incidence and prevalence of visual loss in GCA, its prognosis and to provide objective assessments for the development of new therapies for GCA.

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