Anterior Ischaemic Optic Neuropathy presenting with bilateral blindness due to Giant Cell Arteritis

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Introduction

Newly recognised giant cell arteritis (GCA) should be considered a true neuro-ophthalmic emergency. Prompt treatment with steroids can prevent blindness and other vascular sequelae of GCA.

Case history

A 78-year-old woman with a six week history of headache complained of sudden bilateral visual loss. Her vision became acutely blurred whilst watching television and the next day her vision remained poor.

She had a right cataract removed with IOL (intraocular lens) three years earlier and a total hip replacement two years earlier following which she developed a DVT for which she was warfarinised for 6 months.

On examination she was mildly pyrexial at 37.8^{o}C with a BP of 178/72 and heart rate of 108. She had bilaterally visual acuity of hand movements only with poorly reacting pupils, a weak left temporal pulse and no palpable pulse on the right. Fundoscopy revealed bilateral swollen discs with a small flame shaped haemorrhage inferiorly at the disc of the right eye (fig. 1).

Cardiovascular examination revealed no obvious pathology with normal heart sounds and no carotid bruits. Her ESR was 131, CRP 145 and WCC 13.9.

She was given high dose intravenous methyl-prednisolone pulses and a dose of cyclophosphamide and then she was established on high dose oral steroids.

Her visual acuity on the right improved to 6/60 with pin hole assistance. However, on the left it remained hand movements only.

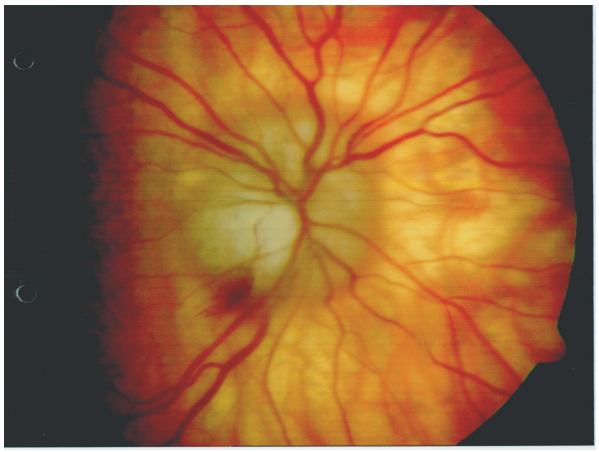

Figure 1. Right Optic disc: Swollen disc with a small flame shaped haemorrhage inferiorly.

A left temporal artery biopsy four days after admission revealed a vasculitis. MRI head showed no abnormalities.

She was discharged with prednisolone 60mg once a day for two weeks, and advised to reduce her dose of prednisolone by 10 mg every week so that by the eighth week post discharge she would be off the steroids. She also received ranitidine 150mg once a day, alendronate 70mg once a week and adcal D3 (calcium carbonate and cholecalciferol) twice a day.

A diagnosis of arteritic anterior ischemic optic neuropathy (A-AION) secondary to GCA was reached and she was discharged with both rheumatology and ophthalmology follow up. Interestingly she was admitted 2 months later for a DVT and is now on lifelong warfarin, with no improvement in her vision.

Discussion

This case highlights the importance of identifying various presentations of GCA. Headache is the cardinal symptom of giant cell artertitis. It is present in 63-85% of people. The diagnosis of GCA is largely based on symptoms

and signs, with subsequent positive histological findings on temporal artery biopsy.

This disease affects those over the age of 55 and is three times more common in women than men. About 80% of GCA patients report feeling ill for some time prior to the episode of vision loss. Common signs and symptoms may include general fatigue, weight loss, fever, temple pain, neck pain, pain on chewing, scalp tenderness when combing their hair, headache, anemia and aching joints. Unfortunately, there is also a variant of GCA that produces none of the above mentioned symptoms, and the patients generally are in good health.

A very important visual symptom in an arteritic AION is a transient blurring or loss of vision that precedes the permanent vision loss. Given that our patient complained of a six week history of headache, the question arises as to whether an ESR should have been measured in the primary care setting.

This woman's long history of headache and subsequent ophthalmic problems were the only real clues that lead to a suspicion of GCA. She denied any of the other common symptoms such as temporal pain, jaw claudiaction and symptoms of polymyalgia rheumatica (PMR). Her case stands out because of the way in which she presented with bilateral AION on admission.

It has been reported that the delay between the first symptoms of GCA and blindness in one eye can be on average 7 weeks.[1]

In older age groups, anterior ischemic optic neuropathy (AION) is the most common cause of acute optic neuropathy. AION is caused by infarction of the short posterior ciliary arteries supplying the anterior optic nerve. The blood supply and blood flow patterns in the optic nerve head have a marked inter-individual variation which exercises a profound influence on the mechanism of development and clinical features of AION.[2]

Clinically two forms exist, the non-artertic form and arteritic form. In the non-arteritic form, these vessels are compromised by vascular disease and arteriolosclerosis. In the case of arteritic AION, these vessels, as well as the ophthalmic and central retinal arteries, are compromised by an idiopathic infiltration of the vessels walls by inflammatory macrophages, lymphocytes, and multinucleate giant cells. As most arteries are affected in GCA, there usually is a constellation of systemic symptoms. Due to the widespread involvement in GCA, there is a propensity for the fellow eye to become similarly involved within days, causing severe bilateral vision loss.

As well as AION, other ocular manifestations of GCA include posterior ischemic optic neuropathy (PION), central retinal artery occlusion (CRAO), cilioretinal artery occlusion, ophthalmic artery occlusion, amaurosis fugax, diplopia and ophthalmoplegia.

This woman's fundoscopy findings are very similar to those documented in previous cases. The visual loss associated with optic disc swelling of a pallid nature, flame hemorrhages on the swollen disc or nearby neuroretinal layer, and sometimes with nearby cotton-wool exudates are all well documented signs. Visual loss usually is sudden, or over a few days at most, and usually is permanent, with some recovery possibly occurring within the first weeks or months.

There is limited trial evidence to support optimum starting dosage of corticosteroids and duration of treatment. According to the PRODIGY quick reference guide issued in April 2006, cases of GCA with new visual disturbance should be given oral prednisolone 60mg with an urgent referral to the Ophthalmology department.

Following a two month course of oral

prednsiolone her ESR and CRP came down to 17 and 11 respectively. In 56% of people, the ESR returns to normal by two weeks, and in 76% of people by five weeks. CRP decreases more rapidly, with 67% normalising in 2 weeks and 75% in three weeks.[3]

Hayerh et al. concluded that early diagnosis and prompt steroid therapy are the keys to any chance of visual improvement, but that improvement will still be rare. They found that only 4% of patients showed real visual improvement (i.e improvement in both visual acuity and central visual field). Their study also addressed the frequent reporting of visual improvement with steroids. They suggest that visual improvement in GCA patients may have been due to VA testing artefacts, particularly eccentric fixation learned by the patient over time.[4]

Chan et al also concluded that prompt treatment of GCA with steroids leads to improvement of visual acuity in a significant number of cases.5 In a retrospective study of 100 consecutive patients with biopsy-proven giant cell arteritis, they concluded that intravenous steroids may offer a greater prospect of improvement compared with oral steroids.[5] A prospective trial comparing intravenous with oral steroids is needed to validate these findings.

GCA is a chronic disease that may last for years. Although the overall course of the disease is one of progressive improvement and eventual complete resolution, the clinical course is highly variable, and, in some patients, it may be protracted for months or years.

Reference:

1: Schmidt D, Vaith P Blindness in both eyes due to late diagnosis of giant cell arteritis. Dtsch Med Wochenschr. 2005 Aug 19;130(33):1874-6.

2: Hayreh SS. Acute ischemic disorders of the optic nerve: Pathogenesis, Clinical Manifestations and Management. Ophthalmol Clin North Am 1996;9:407-42.

3: Leeb BF, Bird HA. A disease activity score for polymyalgia rheumatica. Ann Rheum Dis. 2004 Oct;63(10):1279-83

4: Hayreh, Sohan Singh, Zimmerman, Bridget & Kardon, Randy H. Visual improvement with corticosteroid therapy in giant cell arteritis. Report of a large study and review of literature. Acta Ophthalmologica Scandinavica 80 (4), 355-367

5: Chan CC, Paine M, O'Day J Steroid management in giant cell arteritis. Br J Ophthalmol. 2001 Sep;85(9):1061-4.