

Loss of vision, right eye: an optometry case study

BACKGROUND

An eighty year old female patient, Mrs S, has arrived for examination complaining of an absence of vision in her right eye on several occasions over the last 3 days.

Visual acuities show the right vision to be 6/15 and the left 6/9. The pupils show no relative afferent pupillary defect (RAPD) and are round and reactive.

Examination of the anterior eyes is unremarkable with palpable temporal artery pulses. Intraocular pressures are fine at 10 mmHg OU. Fundus photos show cottonwool spots and a possible retinal haemorrhage superiorly, between the macula and disc.

The disc may be slightly pale but it is not possible to confirm due to the quality of the photo. The left fundus does not exhibit any cottonwool spots or haemorrhages.

Mrs S reports severe pain in the neck, upper back and arms for the last month, worse at night. The pain is enough to hinder her mobility. There is no past history of pain. She suffers from hypertension and arthritis. There is no history of fever or trauma.

Systemic medications being taken by Mrs S are Vasotec, Synthroid, and Premarin.

Mrs S reports loss of vision and also has severe pain in the neck, upper back and arms, worse at night ...

DIFFERENTIAL DIAGNOSIS:

The presentation of these symptoms in a patient would require a differential diagnosis including hypertensive retinopathy, non-arteritic anterior ischaemic optic neuropathy, polymyalgia rheumatica and arteritic anterior ischaemic optic neuropathy.

Hypertensive Retinopathy: This condition exhibits few symptoms but patients may have reduced vision. The ocular signs include binocular generalized or localized arteriolar narrowing (Wills Eye Manual, 1999). Vasoconstriction is the primary response of the retinal arterioles to raised blood pressure. It is related the severity of hypertension and occurs in pure form in young individuals but is affected by pre-existing arterial sclerosis in older patients. In sustained hypertension the blood retinal barrier is disrupted in localized areas resulting in increased vascular permeability. Thus the fundus shows vasoconstriction (either general or focal), haemorrhages (leakage), and arteriosclerosis with "copper or silver wiring" of arteries. The leakage results in haemorrhages, microaneurysms due to ischemia and obstruction of arterioles, and cottonwool spots (Kanski, 2002).

The occurrences of central or branch retinal vessel occlusions are more prevalent in this situation (Wills Eye Manual, 1999). Acute hypertension gives rise to optic disc oedema (the

hallmark of malignant hypertension), retinal oedema, flame haemorrhages and hard exudates in the Henle layer around the fovea forming a macula star.

Arteriosclerosis or thickening of the vessel walls causes arteriovenous nipping which can be graded from 1 to 4 depending on severity (Kanski, 2002). Other manifestations can include choroidal infarcts and anterior ischemic optic neuropathy (AION). This condition would most likely be excluded in this patient mainly due to the unilateral nature of the symptoms and signs. Hypertensive retinopathy is always bilateral and this patient does not exhibit any signs in the left fundus.

Non-arteritic Anterior Ischemic Optic Neuropathy (NAION): This would need to be excluded from the differential diagnosis in this case. This disease most likely can be excluded based on the existence of the preceding transient visual loss. In addition, there is no afferent pupil defect (a critical sign of NAION) present in this case, and there is no reduction in colour vision (Wills Eye Manual, 1999).

NAION symptoms include a sudden, painless, non-progressing moderate vision loss, initially unilateral but may become bilateral. The general age group for NAION is 40 to 60 years. Critical signs include afferent pupil defect, pale swollen disc (often only one segment of the disc), flame haemorrhages and a normal erythrocyte sedimentation rate (ESR).



Other signs include abnormal colour vision, optic neuropathy after oedema resolved and visual field defects (Wills Eye Manual, 1999).

Inflammation occurs within the prelaminar or laminar portion of the optic nerve and is caused by occlusion of the short posterior ciliary arteries. NAION is usually an isolated event with monocular sudden loss of vision without premonitory transient vision loss. The acute phase oedema resolves into pale disc neuropathy, commonly sectoral. Vision loss is slight in one third of cases and severe in other two thirds. Typically visual field defects are altitudinal, commonly involving the inferior field. Colour vision is diminished in proportion to the vision loss.

Investigations required include ESR, fasting lipid profile, blood glucose and viscosity (Kanski, 2002).

Treatment involves treating any underlying disease process and instituting long term, low dose, aspirin to prevent fellow eye involvement. Though prognosis of recovery is poor, most suffer no further visual loss. However approximately one third will develop anterior ischemic optic neuropathy in the fellow eye in the months to years that follow (Kanski, 2002).

Arteritic Anterior Ischemic Optic Neuropathy (AAION): This is the ocular manifestation of the condition Giant Cell Arteritis. Occurrence of AAION is considered an ocular emergency not only due to the ocular developments but also the fact that Giant Cell Arteritis (GCA) has a high morbidity rate.

Polymyalgia rheumatica (PMR) is a major symptom of Giant Cell Arteritis (GCA) but also occurs in isolation (Gurwood, 2001). It is suggested that PMR and GCA may be different manifestations of the same disease process.

Polymyalgia rheumatica is characterized by aching and stiffness of proximal joints, such as shoulders and neck, for more than a month. It is exacerbated in the mornings. PMR mainly occurs in people over 50, as does GCA and is associated with elevated ESR and treated with systemic steroids. PMR can be excluded in this case because there are ocular symptoms suggesting the involvement of GCA rather than PMR.

Giant Cell Arteritis can occur without ocular signs. The symptoms tend to be insidious, often causing delay in seeking medical attention until catastrophic vision loss occurs. Systemic symptoms include most commonly headache, jaw claudication, temporal artery tenderness, scalp pain, PMR, fever and malaise (Gurwood, 2001).

Ophthalmic involvement is reported in up to 50% of cases. Visual loss as the first symptom is reported in up to 36% of cases. Signs include conjunctival hyperaemia and episcleral vessel engorgement. A relative afferent pupil defect is common. Corneal oedema, decreased intraocular pressure and iritis are prevalent. AAION involves the posterior arteries supplying the optic nerve. The optic disc may appear oedematous with indistinct borders. Central and branch vessel occlusions are amongst potential occurrences with this situation (Gurwood, 2001). Cottonwool spots are observed in 33% of eyes effected by GCA indicating local ischemia.

An important symptom of GCA is amourosis fugax (momentary loss of vision). Approximately 30% of patients experience this symptom. The transient optic nerve ischemia may go on to produce AAION. Gurwood (2001) reports of 63% of cases with Amourosis fugax go on to experience permanent visual loss.

AAION is considered an infarction of the optic nerve head at the level of the optic disc. It is theorized that the optic nerve infarction is caused by a thrombus. The mechanism in AAION as with GCA is granulomatous thrombus formation. In arteritic cases the second eye often becomes involved in a few days or weeks, with severe visual loss occurring in the fellow eye despite prompt and immediate systemic treatment (Gurwood, 2001).

Diagnosis of GCA has been established by the American College of Rheumatology to include five clinical criteria; 1) age greater than 50 years, 2) new onset localized headaches, 3) temporal artery tenderness or decreased temporal artery pulse, 4) elevated ESR levels, 5) superficial temporal artery biopsy consistent with GCA. The presence of 3 of these 5 criteria has sensitivity of 93.5% and specificity of 91.2% (Bhatti, 2001).

Laboratory testing includes ESR and the C-reactive protein (CRP) test. Though elevated ESR is not pathognomonic of CGA it does indicate an inflammatory process is present and markedly elevated ESR is a sign of GCA. The C-reactive protein (CRP) test is thought more sensitive than ESR for early detection of GCA. Other tests include anticardiolipin antibody (aCL) levels and CD8+ lymphocyte levels.

The definitive means of confirming CGA is by temporal biopsy. A pathological diagnosis of CGA is indicated by the presence mononuclear or granulomatous inflammation with histocytes, lymphocytes and multinucleated giant cells in the arterial wall (Gurwood, 2001). Elevated ESR levels, CRP levels and a positive biopsy would confirm diagnosis of GCA. A dilated fundus examination to view both optic discs would be useful in confirming AAION. Visual field defects are not specific but commonly altitudinal or involving the central field. These defects are presumably due to direct damage to the nerve fibre axons at the anterior optic disc level.

MANAGEMENT:

The management of GCA is through the use of systemic steroids. Wills Eye Manual (1999) suggests administration of intravenous methylprednisolone (250mg IV q 6 h) for 12 doses in a hospital setting; then changing to oral prednisone daily (80 - 100 mg po). The treatment with oral prednisone to continue if GCA is confirmed by biopsy.

Kanski (2002) suggests intravenous methylprednisolone in combination with oral prednisone for 3 days initially. After three days the oral dose is reduced for another 3 days and then again for the next four days. The daily dose is reduced to 10mg per day and maintained at that level.

Chan, Paine & O'Day (2001) reviewed treatment regimes comparing intravenous methylprednisolone within 24 hours of visual loss compared to oral prednisone and found greater visual improvement with intravenous rather than oral use, suggesting intravenous steroids are superior to oral steroids in treating GCA.

The use of steroids and non-steroidal anti-inflammatory drugs (NSAIDs) will moderate inflammatory reactions via two different pathways. Steroids have an inhibitory influence on the inflammation responses mediated by the T-cells. The inflammation via the T-cells relies on the antigen-presenting cell presenting the antigen to the T-cell receptor. This stimulation causes a change in calcinevrin activation, which in turn dephosphorylates NFAT enabling it to enter the nucleus and bind to the interleukin-2 promoter. For T-cell activation to be optimized interleukin-2 gene transcription occurs preventing T-cell anergy and inhibiting T-cell apoptosis. The interleukin-2 receptor is stimulated causing progression into the cell cycle and denovo purine synthesis occurs which is selective for lymphocytes (Forrester, 2002). Corticosteriods act on cytosolic receptors and block transcription of cytokine genes thus preventing T-cell activation. There are a large number of steroid receptors thus steroids have a multiplicity of actions in the body.

The NSAIDs however involve a different pathway of action. The NSAIDs owe some of their activity to the inhibition of synthesis of eicosanoids. The principle eicosanoids are Prostaglandins, thromboxanes and leukotrienes. The main source of these substances is arachidonic acid found in cell membranes. Stimuli that release the enzymes to create arachidonic acid include thrombin in platelets, C5a from compliment, bradykinins, as well as cell damage. The free acid is then metabolized via two pathways mediated by either cyclo-oxygenase producing Prostaglandins, or lipoxygenase producing leukotrienes. NSAIDs inhibit cyclo-oxygenase thus reduce prostaglandin production (Forrester, 2002).



FINAL CONCLUSIONS:

In this case study the presence of amaurosis fugax with the monocular visual reduction and symptoms of PMR suggest the most likely diagnosis to be arteritic anterior ischemic neuropathy.

Considering the mild to moderate vision loss at this point, the absence of an afferent pupil defect and the presence of palpable temporal artery pulses, it would seem that this case presents a very early stage of disease process.

This might possibly improve the outcome achieved by systemic steroid treatment. Currently intravenous steroids would be preferred to oral steroid use alone. (Chan, Paine & O'Day, 2001).

The immediate initiation of systemic steroids would be strongly recommended for Mrs S.

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