Complete recovery of visual acuity in two patients with giant cell arteritis

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Summary

Principles: Report on two patients with visual loss due to inflammatory anterior ischaemic optic neuropathy (IAION) and cotton wool retinopathy respectively, caused by biopsy-proven giant cell arteritis (GCA), in whom complete recovery of visual acuity (VA) was observed following therapy with intravenous methylprednisolone.

Methods/patients: A 73-year-old male and a 65-year-old female presented with visual loss in the left eye (20/50, 20/60 respectively) for two days at most before admission. Jaw claudication and headache had been present for several weeks before onset of visual symptoms. An extensive laboratory workup (including erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), platelet count), fluorescence angiography and a temporal artery biopsy were performed.

Results: ESR, CRP and platelet count in both patients were elevated. Fluorescence angiography showed early hyperfluorescence of the pale swollen optic disc in one patient and late hyperfluorescence of the fundoscopy unremarkable optic disc and retinal ischaemic spots (cotton wool) in the other. Biopsy of the temporal arteries revealed GCA in both patients. 48 hours at most after the initiation of intravenous high dose methylprednisolone (HDMP) treatment VA improved to 20/20 in both patients and remained stable over observation periods of three and six months respectively.

Conclusion: Visual recovery in GCA patients is rare even when treated with HDMP. Treatment is therefore believed to be effective mainly in saving the unaffected eye. We report on two cases with biopsy-proven GCA in which complete recovery of VA occurred under intravenous HDMP.

Key words: giant cell arteritis; visual loss; visual recovery; high dose methylprednisolone; ischaemic optic neuropathy

Introduction

Visual loss is often the most striking and disabling manifestation of giant cell arteritis (GCA). Due to the non-specificity of a single sign or symptom of GCA, such as general malaise, fever, weight loss, jaw claudication and elevation of erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) (usually present before visual symptoms occur), the diagnosis is frequently missed initially [1]. However, a combination of several signs and symptoms, such as jaw claudication, scalp tenderness, headache and CRP elevation are highly suggestive of GCA. Ocular involvement in GCA with reduced visual acuity (VA), visual field loss (VF) and deficiency of colour vision occurs in 7–60% of patients [2]. Diplopia as a presenting symptom in GCA occurred in up to 5.9% of patients with GCA in a prospective study [3, 4]. The most frequent and devastating ocular manifestations of GCA, however, are inflammatory anterior ischaemic optic neuropathy (IAION), occurring in approximately 75%, and central retinal artery occlusion (CRAO) in approximately 15% [5]. Less frequent are choroidal ischaemia or occipital lobe infarction (due to involvement of the vertebral arteries) [4, 6]. IAION and choroidal ischaemia are thought to be caused by occlusion of the posterior ciliary vessels.

<table>
<thead>
<tr>
<th>Abbreviations</th>
<th>Description</th>
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<td>CRAO</td>
<td>Central retinal artery occlusion.</td>
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<td>CRP</td>
<td>C-reactive protein</td>
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<td>ESR</td>
<td>Erythrocyte sedimentation rate</td>
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<td>GCA</td>
<td>Giant cell arteritis</td>
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<td>HDMP</td>
<td>High dose methylprednisolone</td>
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<td>IAION</td>
<td>Inflammatory anterior ischaemic optic neuropathy</td>
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<td>VA</td>
<td>Visual acuity</td>
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<td>VF</td>
<td>Visual field</td>
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GCA: Uncommon visual recovery

Even partial recovery of vision in GCA is highly uncommon [9–11]. Up to 27% of GCA patients suffer progressive visual function loss despite high dose methylprednisolone (HDMP) treatment within the first six days of the onset of visual symptoms [10]. A few cases have been described in the literature in which visual function improved slightly under HDMP [12–15]. In a prospective study of biopsy-proven GCA in 34 patients treated with intravenous HDMP (1g/d) within 2 days (mean) of the onset of visual symptoms, 83% of the involved eyes had a final VA of 20/200 or worse [10]. Our report describes two cases with biopsy-proven GCA, one with IAION and the other with cotton wool retinopathy, in which the loss of VA completely reversed within 48 hours of the initiation of intravenous HDMP treatment.

Case reports

Case 1

A 73-year-old male patient presented with headache and claudication of the left jaw and substantial weight loss over the previous eight weeks. Twenty hours before admission he noticed loss of VA and loss of the temporal VF of the left eye. VA (Snellen test) measured 20/20 on the right and 20/50 on the left. The right VF (Goldmann perimetry) was unremarkable, while there was partial temporal VF loss on the left (Fig. 1). 15 out of 15 Ishihara plates were identified with both eyes. Fundoscopy demonstrated optic disc swelling on the left (Fig. 1). Fluorescence angiography showed early hyperfluorescence of the left optic disc without any other abnormalities (Fig. 1). Erythrocyte sedimentation rate (ESR) measured 121 mm in the first hour (Westergren), the platelet count was 426'000/μl and the C-reactive protein (CRP) measured 79.3 mg/l. IAION was diagnosed on the basis of the fundoscopic appearance of the optic disc and the laboratory results. Intravenous HDMP (1 g/d) treatment was administered for three days, followed by long term oral prednisolone beginning at 1 mg/kg over six months. The patient was already under antiplatelet treatment (acetylsalicylic acid) at the time of admission. Within 24 hours of initiating treatment the VA in both eyes measured 20/20 and temporal VF loss on the left partially recovered. All laboratory parameters normalised within two weeks. Biopsy of the left temporal artery performed one week after initiation of HDMP treatment showed acute, persistent, severe GCA (Fig. 1). Corticosteroid treatment was tolerated without adverse reactions and VA and VF on both sides remained unchanged for over six months. When the corticosteroid dose was reduced to 15 mg/d after seven months, the patient again developed headache and chest pain without deterioration of visual function. Headache ceased after the corticosteroid dose was increased to 50 mg/d, but remanifested when the dose was reduced. Methotrexate treatment was therefore started in order to allow reduction of the corticosteroid dose without risking remanifestation of symptoms. All visual parameters remained stable following the methotrexate treatment.

Case 2

A 65-year-old female patient presented with a 20-day history of scalp hypersensitivity, occipital headache and bilateral jaw claudication. Transient visual obscurations, graying out, foggy vision and incomplete visual loss were present for 48 hours before admission. The ocular history was remarkable for bilateral open angle glaucoma. VA measured 20/60 in the left eye and 20/20 in the right eye.

Figure 1

Case 1: Normal disc in the right eye (A). Pale swelling of the left disc (more on the nasal side) due to IAION (B). Goldmann perimetry of the left eye (before treatment) demonstrating loss of the temporal visual field (C). Fluorescein angiography (arteriovenous phase): hyperfluorescence of the optic disc, sectorial blockage of adjacent choroidal pattern on the nasal side (D). Histological section (hematoxylin-eosin, x20) of the temporal artery shows thickened intima and a marked mixed inflammatory cell infiltrate consisting of lymphocytes, histiocytes and eosinophils (E). Microscopically, partial destruction of the wall by an inflammatory infiltrate containing multinucleated giant cells (arrow) is present. Some of the multinucleated giant cells are of Langhans type and others are of foreign body type. Many of them are intimately associated with the internal elastic lamina and some may even contain fragments of phagocytosed elastica in their cytoplasm (hematoxylin-eosin, x200) (F).
Intraocular pressure measured 10 mm Hg in both eyes. Fundoscopy revealed several cotton wool spots (arrows) near the optic disc in the left eye, while the optic disc itself looked normal (Fig. 2). ESR was 118 mm in the first hour, CRP 72.3 mg/l and platelet count 486'000/μl. Haemoglobin was 110 g/l. Glucose and lipid concentrations were within the normal range. Serology for HIV, VDRL and cytomegalovirus was negative. A protein immune electrophoresis was unremarkable. Fluorescence angiography showed fluorescence blockage corresponding to the sites of the cotton wool spots (Fig. 2). Histological examination of the left temporal artery biopsy was compatible with the diagnosis of GCA (Fig. 2). The patient was treated with intravenous HDMP (1 g/d) for three days, followed by oral prednisolone. Within twelve hours the headache disappeared completely and VA began to improve. Within 48 hours VA recovered to 20/20. There were no adverse reactions to the corticosteroid treatment and VA remained unchanged over a 3-month observation period. In addition to corticosteroid treatment, antiplatelet treatment (acetylsalicylic acid) was prescribed. No further immunosuppressive medication was needed and the corticosteroid dose was reduced to 2.5 mg/d within another six months without deterioration of visual function. The cotton wool spots disappeared completely within four weeks.

Discussion

Treatment of visual loss due to GCA is unsatisfactory in most cases, with 27% of patients suffering progressive visual loss even under HDMP treatment [10]. VA recovery is uncommon, and, left untreated, GCA causes visual loss in the fellow eye in a significant number of patients within 1 to 10 days [9–15]. HDMP treatment is often considered to be effective only in preventing irreversible binocular blindness by saving the non-involved eye [6]. We describe two patients with biopsy-proven GCA suffering from visual loss due to IAION and cotton wool retinopathy. The patients were admitted within 20 and 48 hours of the onset of VA loss, which improved from 20/50 and 20/60 respectively to 20/20 after treatment with intravenous HDMP. Compared to large studies, loss of vision was minor to medium [5]. In both patients headache and jaw claudication were presenting symptoms several weeks prior to onset of visual function loss, before disappearing spontaneously, and laboratory parameters for ESR, CRP and platelet count were typically elevated. Although the specificity of a single sign or symptom of GCA is low, the combination of several signs and symptoms is strongly indicative for the presence of GCA. Jaw claudication, scalp tenderness, headache, weight loss and elevated CRP, especially in elderly patients exhibiting other rheumatic symptoms such as shoulder or hip pain (consistent with polymyalgia rheumatica), should alert the primary care physician to the possibility of GCA and prompt him to force the diagnosis. In cases of visual function deterioration, early referral to the ophthalmologist is highly mandatory.

The vasculitis in GCA is segmental, initially involving the vessel wall. At this stage patients may experience transient visual obscuration due to functional stenosis during eye movements. If the
inflammation of the vessel wall progresses, the vessel lumen narrows. This allows thrombus formation, which may eventually result in occlusion of the affected vessels. Treatment with HDMP may only be effective before thrombus formation occurs. Unlike the majority of patients with visual loss due to GCA, visual loss in our patients was minor and medium, probably reflecting early involvement of the ocular circulation. HDMP may have prevented substantial thrombus formation in the affected vessel via reduction of the inflammatory-mediated thickening of the affected vessel wall.

To optimise the chance of a favourable outcome for the acutely affected eye, combined treatment with HDMP and heparin may represent the best rationale since it targets both the inflamed vessel wall and luminal thrombus formation [16]. A favourable effect of antiplatelet and anticoagulation treatment in preventing ischaemic complications in patients with GCA, for example IAION, has been described in the literature [17, 18]. Whether antiplatelet treatment in combination with HDMP therapy in acute ischaemic disorders due to GCA is an adequate therapy is still not clear. Although no significant increase in bleeding complications was reported in one study, we feel that in acute disorders heparin treatment could be more effective and safer, due to the fact that heparin treatment is highly controllable. Transient visual obscuration and minor visual loss should therefore always prompt the physician to consider the possibility of GCA and to start treatment with steroids and probably heparin immediately. Although a favourable visual outcome for GCA-associated visual loss is rare, our cases should encourage physicians to adopt a proactive approach in ensuring rapid diagnosis and initiation of therapy for GCA-associated disturbances.

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