



Uncommon but not Rare Ocular Manifestation of Giant Cell Arteritis

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Abstract

Giant cells arteritis (GCA) is the most common form of systemic vasculitis in adults. It is potentially blinding diseases, which needs immediate diagnosis and treatment. The most common ocular manifestation that ophthalmologist see in their practice are visual loss (97.7%), amaurosis fugax (30.6%), diplopia (5.9%). Arteritic anterior ischemic optic neuropathy is the most common cause of blindness in GCA by (81.2%), followed by central retinal artery occlusion (14.1%), and cilioretinal artery occlusion (21.8%) [1].

We will discuss in this article two confirmed GCA cases with uncommon ocular presentation. Namely, Ocular hypotony and isolated cotton wool spots.

Aim: To withdraw the attention of ophthalmologist that uncommon ocular manifestation of Giant cell arteritis is not rare. It might be isolated or associated with Anterior ischemic optic neuropathy. Urgent action may prevent severe visual loss.

Keywords: Giant Cells Arteritis (GCA); Arteritic Anterior Ischemic Optic Neuropathy; Giant Cell Arteritis

Introduction

Giant cells arteritis (GCA) is the most common form of systemic vasculitis in adults, preferentially involves large and medium-sized arteries in patients over the age of 50 [6]. It is potentially blinding diseases, which needs immediate diagnosis and treatment. The most common ocular manifestation that ophthalmologist see in their practice are visual loss (97.7%), amaurosis fugax (30.6%), diplopia (5.9%). Arteritic anterior ischemic optic neuropathy is the most common cause of blindness in GCA by (81.2%), followed by central retinal artery occlusion (14.1%) and cilioretinal artery occlusion (21.8%) [1].

We will discuss in this article two confirmed GCA cases with uncommon ocular presentation.

Aim of the Study

To withdraw the attention of ophthalmologist that uncommon ocular manifestation of Giant cell arteritis is not rare. It might be isolated or associated with Anterior ischemic optic neuropathy. Urgent action may prevent severe visual loss.

Methods

Two case reports of confirmed Giant Cell arteritis patients.

Case Series

Case 1: Ocular hypotony and AION in one eye, followed by cotton wool spots in the other eye.

87 years old lady presented with one day history of loss of vision in her left eye. She reports temporal headache for 1 month. She

was diagnosis with Temporomandibular joint syndrome (TMJ) by General Practitioner (GP), as her blood test showed normal inflammatory markers. Apart from that she is fit and healthy.

On examination, there is left relative afferent pupillary defect. Visual acuity was 6/6 on Snellen chart in the right eye and perception of light in the left eye. Clinical examination found Descemet membrane folds in the left eye with Intraocular pressure of 4 mmHg in the left eye and 9 mmHg in the RE. Anterior chamber was clear and deep, there was no evidence of ischemia. Gonioscopy showed open angle. Dilated funduscopy, showed pale, raised, swollen optic disc in the left eye. There is no evidence of choroidal folds, retinal haemorrhage, or any other lesion. The right eye examination is satisfactory with healthy, not crowded, optic disc.

Blood test showed normal inflammatory markers. However, Intravenous Methylprednisolone was commenced due to the highly suspicious of GCA. Urgent Temporal artery ultrasound doppler was conducted by Rheumatologist, which confirmed the diagnosis. Accordingly, the patient received 1 g of methylprednisolone for 3 days followed by 60 mg of oral prednisolone.

Interestingly, one week later, intraocular pressure went up to 9 mmHg in Left eye.

After few days of tapering down the steroid, the patient came complaining of visual disturbance in her RE (the good eye). Clinical examination, was satisfactory with stable visual acuity in the RE. However, few cotton wool spots around the optic nerve were noted. Immediately the dose of steroid was raised again, and the symptoms and signs has significantly improved few days later.

Case 2: Confirmed GCA with AION and cotton wool spot in left eye, and isolated cotton wool spots in the other eye.

85 years old man referred by optician with sudden painless loss of vision in the left eye since 2 weeks. The patient reported visual disturbance in the right eye as well. He denied any headache, scalp tenderness or jaw claudication. His general health is fine apart from history of hip replacement. There is no history of Hypertension or diabetes.

On examination: Visual acuity is 6/9 in the RE on Snellen chart (similar to his vision in 2018) and counting finger in the LE. There is Left relative afferent pupillary defect. Anterior segment examina-

tion is satisfactory. Dilated funduscopy showed Left swollen optic disc surrounded by cotton wool spots, the right eye showed few cotton wool spots nasal to the macula with healthy looking crowded disc. There was no cherry red spot sign or retinal swelling.

Urgent blood test showed C- reactive protein of 37 and erythrocyte sedimentation rate (ESR) of 90. Temporal artery ultrasound doppler confirmed the diagnosis of Giant Cell Arteritis. Immediate IV methylprednisolone was already commenced and continued for 3 days followed by oral prednisolone.

Few days later, the visual symptoms of the right eye had improved and funduscopy did not show evidence of cotton wool spots in both eyes.

Discussion and Conclusion

Ocular hypotony has been documented earlier in many articles as a presentation of Giant Cell Arteritis. It occurs probably because of restricted production of aqueous humour secondary to ciliary body's arteries involvement [2].

Other study suggested that Hypotony occurred in one third of GCA patients without other signs of anterior ocular ischemia. In addition, low IOP may be helpful as distinguishing factor between GCA related Arteritic and Non- Arteritic Anterior Ischemic Optic Neuropathy [3,5].

The presence of cotton wool spots, which are signs of ischemia, is not surprising when it comes as a part of Arteritic AION or ocular ischemia manifestation. However, isolated cotton wools spots are uncommon. It might be early manifestation preceding severe visual loss. Hence, ophthalmologist should be aware of it, to commence urgent blood test and treatment [4].

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