Saudi Journal of Ophthalmology (2015) 29, 48–52

Neuro-ophthalmology Update

Review of Giant cell arteritis

Joseph G. Chacko [a,*], J. Anthony Chacko [b], Michael W. Salter [a]

Abstract

Giant-cell arteritis (GCA) is a systemic autoimmune disease affecting primarily the elderly. Giant cell arteritis can cause sudden and potentially bilateral sequential vision loss in the elderly. Therefore, it is considered a medical emergency in ophthalmology and a significant cause of morbidity in an increasingly aging population. Ophthalmologists need to be able to recognize the classic symptoms and signs of this disease, and then be able to work-up and treat these patients in an efficient manner. An in-depth review of GCA from the literature as well as personal clinical experience follows.

Keywords: Giant cell arteritis, Temporal arteritis, Cranial arteritis, Granulomatous arteritis, Arteritic ischemic optic neuropathy

http://dx.doi.org/10.1016/j.sjopt.2014.10.001

Introduction

Giant cell arteritis (GCA), also known as temporal or cranial or granulomatous arteritis, is a systemic autoimmune disease affecting primarily the elderly. It is characterized by granulomatous inflammation of the large and medium-sized arteries. GCA is most prevalent in the white population of European origin. There is an association with HLA-DR4 and HLA-DRB1 which suggests a genetic predisposition.[1] Most patients affected are over the age of 60 years. The mean age is 70 years old. Women are affected more often. The incidence can be as high as 27 cases in 100,000 people aged greater than 50 years old. With the rapidly growing proportion of older persons in most developed countries, the incidence of GCA will most likely increase.[2]

Permanent visual loss has been reported to occur in as high as 15–20% of these patients,[3] making early and correct diagnosis critical.[4] Several attempts, such as the American College of Rheumatology (ACR) criteria,[1] have been made to diagnosis GCA without temporal artery biopsy. However, Murchison et al. found that the use of ACR criteria alone could miss up to 25% of GCA diagnoses.[5] Thus, temporal artery biopsy remains the diagnostic gold standard for GCA.

The symptoms of giant cell arteritis can overlap with its cousin disease polymyalgia rheumatica (PMR). PMR similarly is an autoimmune disease that affects the elderly. Its hallmark symptoms include muscle pain and weakness affecting the large muscle groups especially in the hips and shoulders. Patients with PMR have trouble getting out of a chair and reaching for objects in cupboards. PMR symptoms also include low-grade fever, malaise, poor appetite, and weight loss. When symptoms affect the neck and higher, giant cell arteritis can be at work. Five to 15% of PMR patients will be diagnosed with giant cell arteritis, and 50% of giant cell arteritis patients have PMR symptoms.

The medium-sized extracranial arteries are most frequently affected clinically in GCA. However, occasionally, the aorta and its branches to the arms and neck or elsewhere are involved.[1]

Received 16 August 2014; received in revised form 16 September 2014; accepted 1 October 2014; available online 1 November 2014.

[a] Jones Eye Institute, Department of Ophthalmology, University of Arkansas for Medical Sciences, 4301 W. Markham, #523, Little Rock, AR 72205, USA
[b] University of Arkansas, Fayetteville, AR, USA

* Corresponding author. Tel.: +1 501 686 5150; fax: +1 501 603 1289.
e-mail address: jchacko@uams.edu (J.G. Chacko).

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online:
www.saudiophthaljournal.com
www.sciencedirect.com

Systemic symptoms

The classic systemic symptoms of GCA include headache, scalp tenderness, and jaw claudication. Headache especially located at the temples is the most common symptom. Jaw claudication is the most specific symptom. It is important to remember that jaw claudication means pain with chewing. This symptom is not constant jaw pain. Patients develop a significant jaw ache while chewing, so they decrease their food intake and thereby lose weight and feel weak. A good way to ask about scalp tenderness is to ask whether combing or brushing the hair hurts the scalp. Infrequently, a patient may notice an inflamed artery on the scalp or temple and tell his or her doctor that it is tender leading to the diagnosis. GCA patients may also suffer from ear pain and neck pain. Again they may share the symptoms of PMR with intermittent low-grade fever, weight loss, malaise, and joint/muscle pain.

Visual symptoms

Sudden, severe, and sequential vision loss is the hallmark of giant cell arteritis. The vision loss is usually discovered upon awakening in the morning. Visual acuity is usually less than 20/200 in greater than 60% of patients who lose vision. The fellow eye usually gets involved within days to weeks of the initial eye. In addition to causing a sudden permanent vision loss, GCA can present weeks earlier with amaurosis fugax or a temporary loss of vision which is due to partial occlusion of the short posterior ciliary arteries or central retinal artery causing transient ischemia. GCA may initially also present with diplopia or eye pain.[6] Cranial nerve palsies (3, 4, or 6) or ischemic myopathy may rarely occur. This reinforces the point that any elderly patient presenting to the eye clinic with visual symptoms or eye pain should be considered to be a GCA suspect until proven otherwise. This mode of thinking will help minimize permanent vision loss in GCA patients.

AION in GCA

Sudden vision loss in GCA occurs most often due to an inflammatory thrombosis of the short posterior ciliary arteries. The short posterior ciliary arteries form a fine vascular network that supplies the optic disk. When these vessels become thrombosed with inflammation, a stroke to the optic disk occurs. This is called anterior ischemic optic neuropathy or AION (Fig. 1). AION is characterized by a swollen optic disk accompanied by hemorrhages and sometimes exudates. The swollen optic disk may have a chalky white appearance in GCA. This pallid swelling (Fig. 2) is due to the extreme ischemia of GCA. Rarely, the ischemia to the optic nerve occurs posteriorly, and therefore there is no disk swelling. In this instance, it is called posterior ischemic optic neuropathy or PION.

GCA may also sometimes cause a central retinal artery occlusion (CRAO). About 5% of patients over age 50 with CRAO have GCA. A classic cherry red spot in the macula occurs, but no cholesterol or calcific embolus will be seen since again this is due to an inflammatory thrombosis. GCA may also rarely cause a cilio-retinal artery occlusion or ocular ischemic syndrome. The ocular ischemic syndrome is characterized by eye pain, iritis, and hypotony.

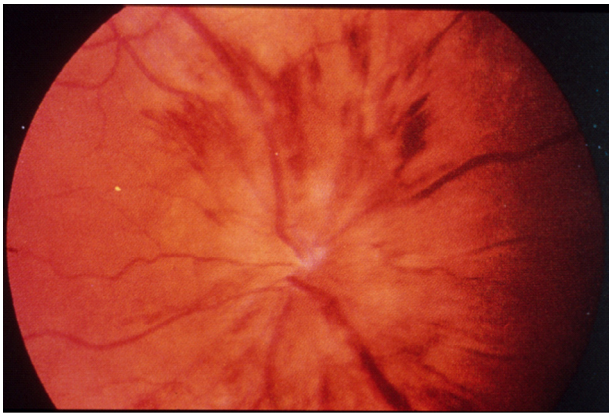

Figure 1. Anterior ischemic optic neuropathy (AION) is characterized by a swollen optic disk accompanied by hemorrhages.

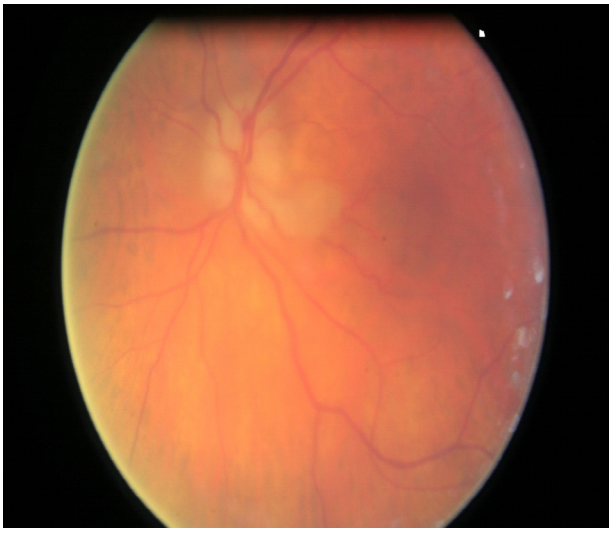

Figure 2. The pallid disk swelling may have a chalky white appearance in GCA.

About 5–10% of anterior ischemic optic neuropathy (AION) cases over the age of 60 are due to giant cell arteritis. The other 90–95% are due to garden-variety non-arteritic ischemic optic neuropathy (NAION). The risk factors for NAION include the disk-at-risk appearance of the optic disk (cup-to-disk ratio < 0.1), nocturnal hypotension (taking blood pressure medicines at night), sleep apnea, uncontrolled hypertension, diabetes, and tobacco abuse.

Bilateral involvement of anterior ischemic optic neuropathy in temporal arteritis is not uncommon. In untreated cases, it occurs 54–95% of the time. Cotton wool spots may also be seen in the retina and indicate concurrent retinal ischemia. Fluorescein angiography (FA) may be helpful to identify choroidal hypoperfusion and aid in the timely diagnosis of GCA.[7]

Laboratory diagnosis

The two most important labs to order to help make the diagnosis of GCA are the Erythrocyte Sedimentation Rate (ESR) and the C-Reactive Protein (CRP). These two lab values, if elevated, indicate systemic inflammation. If either blood value is high, this may point you to a diagnosis of GCA, but