

A Case of Temporal Artery Dissection in a Patient with Giant Cell Arteritis

Shubha Kollampare*, Wendy Mackerricher, Dominick Sudano, Rafael Grau and Jeffrey Lisse

University of Arizona, Tucson, USA

Abstract

Giant cell arteritis (GCA) is a chronic vasculitis involving medium and large sized arteries. A 73-year-old male presented with right eye visual loss and jaw claudication associated with photosensitivity, supraorbital and retrobulbar pain. Evaluation demonstrated bitemporal tenderness, elevated ESR, CRP, leukocytosis and positive PANCA. Stranding and abnormal enhancement of right retrobulbar fat along the optic nerve sheath complexes was noted on MRI. Emergent biopsy of right temporal artery showed giant cell infiltration, disruption of internal elastic lamina with medial dissection. Despite steroid therapy, blurry vision in the left eye progressed to blindness. Tocilizumab infusion was initiated without much improvement. Interleukin 6 levels correlate with disease activity and severity in GCA. Glucocorticoids reduce inflammation by decreasing IL-6; however in refractory cases, the use of interleukin 6 antagonists may aid in rapid induction of remission. Blindness is a serious and irreversible complication of GCA, which makes early detection and treatment paramount.

Keywords: Temporal arteritis; Vasculitis; Blindness; Glucocorticoids; Giant cell arteritis

Introduction

Giant cell arteritis (GCA) is a chronic vasculitis involving the medium and large sized arteries that typically occurs in individuals over 50 years of age. This condition usually affects the extra-cranial branches of the arteries originating from the aortic arch [1]. Failure to diagnose this entity may lead to anterior ischemic optic neuropathy secondary to occlusive arteritis of the posterior ciliary artery. This compromises the blood supply to the optic nerve there by causing irreversible visual loss. Thus, a new onset headache with sudden onset of visual disturbances and jaw claudication in a patient older than 50 years should be taken seriously. We present a 73-year-old man with a classic presentation of temporal arteritis complicated by acute medial dissection of the temporal artery leading to acute and progressive visual loss. This is a novel mechanism for visual loss in GCA.

Case Report

A 73-year-old male presented with the chief complaint of loss of right eye vision for five days. Prior to presentation, he had a two-month history of bilateral ocular pain and jaw claudication, worse on the right side. Over the two-month period, he had multiple hospitalizations for recurrent ocular and facial pains associated with an increased feeling of pressure in his right ear. He was diagnosed with sinusitis and was discharged on antibiotics.

Without much relief in his pain, he presented to an outside hospital with ongoing neck pain and new right eye pain associated with worsening occipital headaches. An MRI demonstrated an ill-defined stranding and enhancement in the retrobulbar fat surrounding the bilateral optic nerves and optic sheaths. He was diagnosed with pseudotumor cerebri, right-sided optic neuritis, and was started on oral prednisone 60 mg once a day. His symptoms improved initially; however, they returned once the steroid dose was tapered. He then returned to the same hospital with new right-sided vision loss, and was subsequently transferred to our University for further evaluation.

On evaluation, he endorsed photosensitivity, supraorbital and retrobulbar pain bilaterally. He denied any lacrimation, scleral changes, or eye discharge. Other associated symptoms included jaw claudication, dizziness, dorsal neck pain, and profuse sweating in the absence of shoulder or hip girdle weakness. He complained of vision loss in the right eye with preserved left eye vision.

His past medical history was notable for hypertension and

hyperlipidemia. His medications included carisoprodol 350 mg tablet three times daily hydrocodone-acetaminophen 5-325 mg tablet every 6 hours as needed, ibuprofen tablet 600 mg by mouth every 6 hours as needed, tramadol tablet 50 mg by mouth every 8 hours as needed, aspirin 81 mg oral once daily, lisinopril 5 mg oral once a day and prednisone 60 mg tablet by mouth once a day.

On physical exam, the patient was in moderate distress with stable vital signs. He had jaw tenderness to palpation bilaterally. His pupils were round bilaterally and unequal on the right (4 mm on the right compared to 3 mm on the left). No conjunctival injection was noted. The patient did have bilateral temporal tenderness, right greater than left, with palpable temporal artery pulses. Fundoscopic exam revealed optic disc edema with blurred margins in the right eye. Cardiac and respiratory exam were normal. Abdomen was soft and non-tender with no evidence of bruits. The patient had no synovitis, erythema or joint effusions. Full range of motion observed in all extremities. The skin exam was unremarkable.

Laboratory studies showed an erythrocyte sedimentation rate of 67 mm/hr and a c-reactive protein of 16.40 mg/L. Leukocyte count was 23,200 per μm^3 . Hemoglobin and hematocrit were 12.1 mg/dL and 36.6%, respectively. Platelet count was 398,000 per μm^3 . Urinalysis had no significant findings. Rheumatoid factor and Anti-nuclear antibodies were both negative. A repeat MRI confirmed the stranding and abnormal enhancement of the right retrobulbar fat along the optic nerve sheath complexes. A 5 mm para-ophthalmic artery aneurysm was also observed. Based on this presentation, there was a high suspicion of giant cell arteritis, and the patient was immediately started on methylprednisolone 1 mg IV once daily.

Despite the treatment with IV steroids, the blurry vision in his left eye progressed to blindness on hospital day 2. Emergent biopsy of the right temporal artery was performed on the same day. A 1.5 cm

*Corresponding author: Shubha Kollampare, University of Arizona, Tucson, USA, Tel : +1 520-621-2211; E-mail: sh123pg@gmail.com

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segment of the artery obtained showed marked vasculitic changes with infiltration of inflammatory cells, giant cells, and disruption of the internal elastic lamina with medial dissection. Biopsy of the specimen can be seen in the Figures 1 and 2.

Further lab workup revealed a positive p-ANCA, so a PET scan was obtained to look for involvement of other arteries. The results were consistent with mild giant cell arteritis showing mild to moderate increase in activity in the arteries arising from the aortic arch, subclavian, and common carotids bilaterally. No abnormal activity was noted in the bilateral renal arteries. Mild atherosclerotic calcifications were noted in the abdominal aorta. No abnormal activity was detected in the brain. He was discharged on oral prednisone, and seen in rheumatology clinic. Since his discharge, he continued to have bilateral visual loss with one flare up over the course of 6 months. This consisted of right-sided facial pain that responded to an increase in his steroid dose.

Discussion

GCA causes vasculitis of large and medium vessels and is usually seen in patients older than 70 years of age; it seldom occurs in patients less than 50 years of age. About 15-30% of patients may have concurrent

polymyalgia rheumatica [2]. Interleukin 6, an inflammatory cytokine, is known to play a major role in GCA and thus correlates with disease activity and severity.

Our patient presented with classical symptoms of giant cell arteritis along with a biopsy of the temporal artery that showed focal medial dissection and invasion of the internal elastic lamina by giant cells. Focal dissection is a very rare occurrence. Ours is the second case with this complication in the literature; the first one having been reported in 2001.

In a study conducted at Brigham and Women's Hospital from May 1987 to November 1997, 447 temporal artery biopsies from 416 patients were obtained. A review of these biopsies showed the presence of giant cell arteritis in 59 patients and vasculitis in the absence of giant cells in 20 patients. No case of temporal artery dissection was noted [1]. Large and medium sized artery dissections have been known to occur in other forms of vasculitis, namely Takayasu's arteritis [3] and Systemic Lupus Erythematosus; however, our patient's age, ethnicity, and the absence of renal or other visceral involvement, including the results of a PET scan, made these diagnoses less likely. Arterial dissections are known to occur because of vessel wall weakness secondary to hypertension and atherosclerotic disease. Dissection of the temporal artery has been known to occur due to repeated palpation of the inflamed artery [4].

Permanent monocular or binocular vision loss has been reported in 15-20% of patients with GCA. Vision loss is known to occur from anterior ischemic optic neuropathy, central or branch retinal arterial occlusion, posterior ischemic optic neuropathy and very rarely from cerebral ischemia. Risk factors for the vision loss being age, hypertension, thrombocytosis [5]. Constitutional symptoms such as fever, weight loss along with laboratory abnormalities namely hemoglobin <11 g/dl, ESR >85 mm/hour help in early diagnosis and institution of timely treatment while preventing ischemic complication [6].

Glucocorticoids, when administered, decrease the levels of IL-6, thereby reducing inflammation. Since our patient had an aggressive disease course with vision loss in both eyes, he was given pulse dose glucocorticoids in the form of IV methylprednisolone and one dose of Tocilizumab. Tocilizumab is an IL-6 receptor antagonist that is used to cause rapid induction of remission in patients with aggressive disease as noted in our patient with bilateral vision loss [7].

Patients with giant cell arteritis are predisposed to cardiovascular events and hence need to be closely monitored [8]. Hypertension and presence of atherosclerotic changes usually lead to arterial dissection. Our patient had well controlled hypertension; however, the biopsy did show Monckeberg's calcification. Monckeberg's medial calcification may have weakened the vessel walls and this, with the invasion of inflammatory cytokines and giant cells, may have led to the dissection despite early, aggressive efforts.

Dreaded complication of temporal arteritis-permanent visual loss, may be prevented, if the ancillary staff are educated about the pathology and guided to identify the alarm symptoms such as maxillary/dental pain, facial edema, sore throat, jaw claudication and most importantly amaurosis fugax. Transient vision loss could be the harbinger of permanent vision loss and hence needs urgent attention.

A strong "partnership" between the nursing staff and the patient will enable in early identification of the alarm symptoms and thereby possibly prevent the catastrophic complication of permanent vision loss [9].

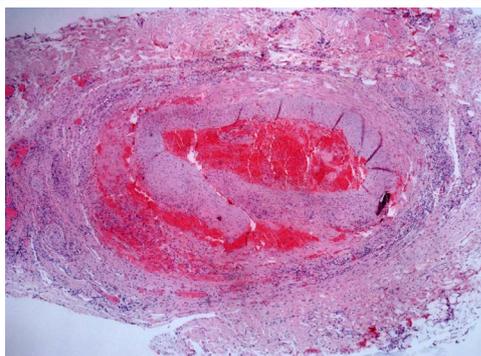


Figure 1: H and E stain: A dense inflammatory infiltrate (blue arrow) involves all arterial layers, and is composed of abundant histiocytes (confirmed by CD163 immunohistochemistry), lymphocytes, rare neutrophils, and rare giant cells, with extensive destruction of the internal elastic lamina, consistent with active giant cell arteritis. Accompanying this inflammatory process is acute medial dissection, with tracking of hemorrhage between the smooth muscle layers of the media (black arrows). Focal Mönckeberg medial calcification is also present (green arrow), with mild age-related intimal fibroplasia.

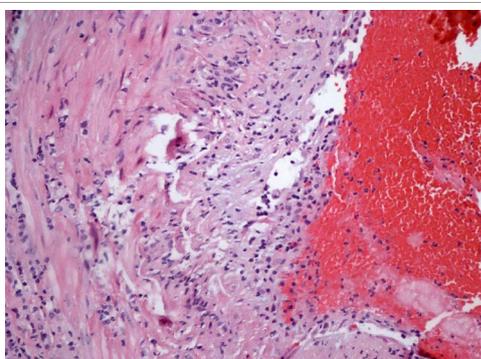


Figure 2: H and E stain: Dense lymphohistiocytic infiltration is present, with extensive destruction of the internal elastic lamina. Out of suspicion for refractory giant cell arteritis, he was given a dose of Tocilizumab IV infusion at a dose of 4 mg/kg in addition to a second pulse dose of methylprednisolone. He was also started on low dose heparin drip and aspirin for ischemic stroke prophylaxis secondary to giant cell arteritis. After receiving a 5-day course of IV steroids, the patient was tapered to prednisone 60 mg PO daily.

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