Internal carotid artery stenosis associated with giant cell arteritis: case report and discussion

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Abstract

Background—Cerebrovascular ischemic events associated with giant cell arteritis (GCA) are uncommon and have been reported in 3%–4% of patients. We describe a case report of GCA associated with intracranial stenosis and review various angiographic findings.

Case presentation—A 66-year-old man presented with worsening headache and vision loss. A recent magnetic resonance angiogram of the head and neck showed multiple intracranial stenosis. Cerebrospinal fluid (CSF) analysis demonstrated increased protein of 135.6 mg/dL, with two white blood cells/µL. No bacteria were observed in the CSF on gram staining, and cultures were negative for bacterial growth. Erythrocyte sedimentation rate was noted to be 14 mm/h, and C-reactive protein was 1.514 mg/L at admission. Human immunodeficiency virus (HIV) and hepatitis panels were negative. On digital subtraction angiography, patient had predominantly narrowing and irregularities in petrous and cavernous segments of the internal carotid arteries bilaterally. The diagnosis of GCA was confirmed by temporal artery biopsy. He was treated with steroids, and a follow-up angiogram 6 weeks later showed minimal resolution of the angiographic findings. Patient reported complete resolution of headaches and visual loss.

Conclusion—Bilateral internal carotid arteries stenosis may be seen in patients presenting with typical symptoms of GCA and may persist after steroid treatment despite resolution of clinical symptoms.

Keywords
Temporal arteritis; giant cell arteritis; intracranial stenosis; internal carotid artery

Introduction

Cerebrovascular ischemic events associated with giant cell arteritis (GCA) are uncommon and have been reported in 3%–4% of patients [1]. There is a paucity of data with regards to the angiographic appearance of intracranial stenosis associated with GCA particularly in the anterior circulation. We describe a case of GCA associated with intracranial stenosis and review angiographic and clinical findings. We supplement the case presentation with literature review of intracranial angiographic findings associated with GCA.

Case presentation

A 66-year-old man was referred to our emergency department due to a worsening headache, vision loss, and multiple intracranial stenoses in the internal carotid arteries diagnosed on magnetic resonance (MR) angiogram of the head and neck. He had a past medical history of type-II diabetes mellitus, hypertension, and atrial fibrillation and was being treated with warfarin. He had a previous episode of jaw claudication and a loss of peripheral vision which occurred 3 and 2 months prior to current presentation, respectively. He was evaluated in the emergency department and noted to have dull, bilateral fronto-temporal headaches, and mild patchy right temporal monocular vision loss. No other neurological deficits were noted. The dermatological examination was negative for icterus, rash, or other skin lesions.

Patient was admitted and started on 60 mg daily of prednisone for suspected GCA. He reported immediate resolution of his headache. Erythrocyte sedimentation rate
(ESR) was noted to be 14 mm/h, and C-reactive protein was 1.514 mg/L at admission. Human immunodeficiency virus (HIV) and hepatitis panels were negative. Lumbar puncture and cerebral angiography were performed to further investigate the etiology of intracranial stenosis. Cerebrospinal fluid (CSF) analysis demonstrated increased protein of 135.6 mg/dL, with 2 two white blood cells/µL.

No bacteria were observed in the CSF on gram staining, and cultures were negative for bacterial growth. A four-vessel cerebral angiogram was performed, which demonstrated severe multisegmental narrowing of both right and left internal carotid arteries, involving the petrous and cavernous segments (see Figure 1). The supraclinoid segment, middle cerebral artery and anterior cerebral artery were unaffected. Focal narrowing of the dural segment of the vertebral artery, with normal appearing basilar artery and posterior cerebral arteries, was also noted (Figure 1). There was also a mild narrowing of the superficial temporal artery. There was no renal artery disease noted on renal angiogram. A temporal artery biopsy was also performed on hospital day 5 due to previous history of jaw claudication, visual loss, and headache. The biopsy demonstrated patchy chronic inflammation within the arterial wall, which was compatible with GCA under therapy. The patient was followed up in clinic after discharge at 30 days and noted to have a resolution of headache with significant improvement in his vision with minimal residual deficit. A 6-week follow-up cerebral angiogram was also performed which showed minor resolution of the previously mentioned dural based arterial stenosis (Figure 2). Patient did not experience any new symptoms.

**Discussion**

GCA is a medium to large vessel arteritis which typically affects the elderly persons aged greater than 50 years. Histologically, it is indistinguishable from primary central nervous system vasculitis [1] and temporal artery biopsy remains the gold standard for the diagnosis of GCA. The sensitivity of unilateral temporal artery biopsy for diagnosis of GCA is 87% [2] According to another study, bilateral temporal artery biopsy improves the diagnostic yield to 90% or greater [3]. In biopsy negative cases, positron emission tomography (PET) using 18-fluorodeoxyglucose as a tracer is useful in diagnosing GCA. The use of this technology has also demonstrated the involvement of large arteries, such as the
aorta or the subclavian arteries, which occurs in 50%–80% of patients [4]. Computed tomography angiogram and MR angiography may show the extent of the disease, but are not helpful in the diagnosis of GCA and thus, are rarely used [4].

Previous reports estimate that there is a 3%–4% rate of ischemic cerebrovascular events in GCA patients [5,6]. Although rare, ischemic events in patients with GCA have been attributed to high grade stenosis or occlusion of the vertebral and/or carotid arteries [7–9]. The relative sparing of the intracranial cerebrovasculature is thought to be due caused by a lack of elastic fibers in the media and adventitia of the intracranial vessel wall [8,9]. It has been reported that patients with intracranial involvement may have a fulminant course of neurological decline, progressing to death [1]. Our patient presented with typical symptoms of GCA; however, around 40% of the patients with GCA may have an atypical presentation [10]. A single series involving 248 patients shows that 10 patients suffering ischemic events and were being treated as cases of atherosclerosis. These patients were actually suffering from GCA and the diagnosis was reached after performing cerebral angiography [11].

The vertebral artery is a common site of extracranial and intracranial involvement. Around 50%–75% of the strokes associated with GCA have been reported to occur in the vertebrobasilar circulation [12]. Internal carotid artery occlusion was noted in a case report, in which the patient underwent a superficial temporal artery and middle cerebral artery anastomosis. The patient was started on concurrent steroid therapy. Repeat angiography performed at 3 months, showed resolution of the superficial temporal artery beading, but the internal carotid artery remained occluded and showed lowering of vascular reserve on single-photon emission computed tomography [13]. The findings of this case and our observation of minimal resolution in our patient that did not resolve with steroids, suggests an irreversible component to GCA-induced arterial changes.

**Conclusion**

Patients presenting with typical or atypical symptoms of GCA may suffer concurrent stenosis of intracranial vessels and should undergo cerebral angiography to evaluate for stenosis or occlusion of the intracranial vasculature. Bilateral internal carotid arteries stenosis may be seen in patients presenting with typical symptoms of
GCA and may persist after steroid treatment despite resolution of clinical symptoms.

References