Stroke and internal carotid dissection as the first manifestation of giant cell arteritis: a case report

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Abstract

Introduction: Giant Cell Arteritis (GCA) is a systemic vasculitis that involves medium-sized and larger arteries.GCA rarely can affect the brain-arteries, resulting in ischemic strokes and transient ischemic attacks, whereby the most affected region is vertebrobasilar. Also, it's very unusual that cocurate with large artery dissections. We describe a patient with anterior brain territory stroke and right internal carotid dissection as the first manifestation of GCA.

Case report: A 51-year-old man was presented with sudden onset of right-side blurred vision, frozen movements, and ptosis in the right eye and left side paresis. There was a history of right-sided frontotemporal headache, diabetes mellitus, and dyslipidemia. The Laboratory tests show erythrocyte sedimentation rate (ESR)=90, C-reactive protein (CRP)=15mg/L. CT scan and brain MRI indicated an acute ischemic infarction in the right frontal region. CT-Angiography has shown internal carotid artery dissection. Due to the presentation and lab tests, we started Methylprednisolone 1 gr for treating GCA, and temporal artery biopsy was positive for GCA in pathology findings. After ten days, inflammatory markers were reduced (ESR:40). Besides improving headaches, there was no significant change in eye deficits and reduction of left limb's force.

Conclusion: Due to the noisy symptoms in patients with Stroke and carotid dissection, the diagnosis of GCA may be neglected as an underlying cause. The association of high CRP levels and the rate of ESR with Stroke, although nonspecific, should draw some attention to vasculitis, topped by GCA.

1 Introduction

Giant Cell Arteritis (GCA) is the most prevalent systemic arteritis that influences medium-sized and larger extradural arteries ,usually in the fifth decade of life. Intracranial arteries are generally do not get involved. If the ophthalmic artery and its branches are involved, it can lead to irreversible and painless vision loss. (1, 2)

Patients with GCA can present to medical centers with various neurological symptoms such as jaw claudication, headache visual symptoms, or cerebrovascular events. GCA rarely can affect the brain-arteries, resulting in ischemic strokes and transient ischemic attacks, whereby the most involved region is vertebrobasilar. (1, 3-5) As an essential point, in GCA patients with stroke, the overall survival was decreased, with a 10th survival percentile of 4.4 months versus 221.7 months in GCA lonely. (6)

We present a middle-aged patient with sudden visual loss, frozen eye movements, ptosis, and left side paresis with acute ischemic infarction and internal carotid artery dissection in imaging and high ESR level in lab tests. The aim of writing this case report is to draw attention to the fact that GCA may develop new and previously unreported ways that require proper diagnosis and prompt treatment to improve the prognosis of the disease.

2 Case Presentation

A 51-year-old man was presented to the neurology ward of Rajaei Hospital affiliated to Alborz University of Medical Sciences in April 2021 with sudden onset of right-side blurred vision, frozen movements, and ptosis in the right eye. He also complained of sudden onset left side paresis. There was a severe recent history of right-sided frontotemporal headache. There were no symptoms of Covid19 infection, fever, seizure, respiratory problems, and jaw claudication. He also reported smoking, but he quit several years ago and had diabetes mellitus and dyslipidemia.

On examination, the patient was conscious, oriented, obeyed, and vital signs were stable with SpO2:94%, there was no tenderness over temporal arteries, and their pulses were normal. In appearance, the right eye had complete ptosis, and in ophthalmoscopy, we detected that the right optic disc was pallor. The right pupil was not responding to light, and the right eye's visual acuity was no light perception. Examination of ocular movements affirms a complete right ophthalmoplegia. Evaluation of the left eye was unremarkable. Also, the force of the left side limbs was reduced to 4/5. The first Laboratory tests shows: WBC=6500, Hb=15.1, HCT=47.5, platelet count=245000, erythrocyte sedimentation rate (ESR) =90, and c-reactive protein (CRP) =15mg/L. Spiral brain computed tomography (CT) scan and brain magnetic resonance imaging (MRI) indicated an acute ischemic infarction in the right frontal region ***(Figure 1)*** and some involvements in the maxillary sinus that spread to the cavernous sinus. The echocardiography was normal, but the Ultrasound of the right carotid artery was completely occluded. Due to the history of diabetes, complete right ophthalmoplegia, right side blurred vision, and imaging investigations, we were suspected of Mucormycosis. We started routine orders for cerebrovascular accident (CVA) and Amphotericin B and antibiotics for empirical therapy. We also performed an endoscopic sinus biopsy which was negative for Mucor, so we stopped the antifungal therapy. Brain magnetic resonance angiography (MRA) demonstrated significant stenosis with a large clot in the right internal carotid artery (ICA) that spreads to the sinus of the cavernous. ***(Figure 2)*** Cause of this report, we promote a CT-Angiography that shown internal carotid artery dissection. ***(Figure 3)*** Also, due to the history of ipsilateral frontotemporal headache and the high ESR level, we started pulse of Methylprednisolone 1 gr daily in suspicion of Giant Cell Arthritis (GCA), and temporal artery biopsy was done after one week, which was positive for GCA in pathology findings.

After ten days of undergoing corticosteroid therapy, inflammatory markers were reduced (ESR:40). Besides improving headaches, there was no significant change in ptosis and ocular movements, visual acuity, and reduction of left limb's force. After two weeks, we discharged the patient with Aspirin 80, clopidogrel 75mg, Atorvastatin 40 mg, and prednisone 50 mg daily. Also, he was referred to the rehabilitation center for further treatments.

3 Discussion

Diagnosis of GCA is critical because delay can cause irreversible loss of vision in patients. Also, it can be challenging in those without the classic symptoms, such as headaches. The American College of Rheumatology criteria for the classification of GCA is not very specific in diagnosing clinical cases. Therefore, vascular imaging (ultra- sonography, computed tomography, magnetic resonance imaging, or

positron emission tomography), temporal artery biopsy (TAB), or a combination of these tests help with a definitive diagnosis. (7)

GCA-related stroke is defined as if the stroke showed GCA or occurred between the onset of symptoms and four weeks after the beginning of treatment. A population-based study proved that GCA-related stroke mainly affects the vertebrobasilar territory and usually occurs in older men with a history of vascular risk factors. (8) Through cardiovascular risk factors, only having diabetes mellitus was significantly higher in people with stroke during follow-up. (6)As a point to consider in our case, stroke affects the internal carotid region, and diabetes was the main cardiovascular risk factor. In another study, patients with GCA who experience recent ophthalmic ischemic symptoms and present with low inflammatory variables were more susceptible to stroke. (9) Patients with stroke in vertebrobasilar regions had more commonly irreversible visual loss due to involvement of ophthalmic artery branches derived from the internal carotid than the other GCA patients. (10) In confirmation of the previous study, our patient had no significant change in visual acuity after treatment.

The GCA symptoms detected at the time of the CVA were: headaches, signs of polymyalgia rheumatica, and acute anterior neuropathy. Acute-phase reactants were increased by (83%) at the stroke event. At the time of the stroke, 22% of patients were on antiplatelet therapy. Of the 18 patients, only 5 had Magnetic resonance brain imaging that indicated ischemic lesions located in the carotid territories. Vascular stenosis or occlusion was observed in vertebral arteries in 11 individuals, with bilateral involvement in 6 patients; basilar artery in 2 cases, circle of Willis in 2 persons, and internal carotid in 1 patient. (6). According to the information mentioned in our case had a severe recent headache and sudden visual loss with moderately elevated CRP and ESR. Also, the brain's parenchymal territory involvement and the kind of vascular stenosis are rare and unusual.

Moghaddasi M et al. reported a 67-year-old man who was finally diagnosed with giant cell arteritis. The patient presented with a recently temporal headache, cranial nerves palsy, elevated ESR, and severe internal carotid artery stenosis. However, the biopsy of the temporal artery was normal. (11)A case report presented a 59-year-old man with multiple strokes who the conventional angiogram demonstrated stenosis of bilateral carotid and vertebral vessels. (12)Another case was presented with multiple acute infarctions in the territory of the vertebrobasilar arteries with the left vertebral artery stenosis in the MRI and an increase in inflammatory markers. (13). The cohort of all residents of Olmsted County, Minnesota, in whom GCA was diagnosed and followed up in the duration of 50 years, demonstrated that the incidence of large artery stenosis is 13%. (14). Several studies demonstrated that ischemic cerebrovascular attacks are mainly linked with the occlusion or stenosis of the extradural vessels instead of the intradural vasculitis. (15, 16) Siemonsen et al. assessed patients suspicious of GCA by using MRI protocol focused on evaluating the intradural arteries. Ten out of 25 cases presented with vessel wall enhancement (VWE) of the intradural ICA; eventually, all these cases were positive for GCA. Moreover, the involvement of the intradural vessels did not associate with the cerebral ischemic lesions. (17)

Few case reports published vertebral and carotid artery dissection and pseudo dissections with severe cerebral ischemic signs and symptoms without GCA. (18-22)A case report presented a 56-year-old man with arthralgia, weight loss, and progressing minor neurological symptoms over one month. Neurosoncological evaluation indicates occlusion in both internal carotid arteries (ICA) and intracranial segments of the left vertebral artery (VA) and the famous hypoechoic halo sign in both superficial temporal arteries. They confirmed GCA diagnosis by inflammatory markers and biopsy. During the treatment, brain MRI suggests watershed infarcts and intracranial dissections of both ICA and left VA. (18) In another report, Zheng X et al. presented a case that ICA dissection was the underlying reason for ION. They recommended that the threshold of suspicion of the ICA problems during ION and initiating treatment should be lowered. (23) As the same in our case CT-angiography indicated right internal carotid dissection.

A combination of corticosteroids and antiplatelets may help prevent stroke occurrence in patients with GCA. However, the combination of antithrombotic and corticosteroid therapy is less effective in GCA patients. Cause the stroke due to Giant cell arteritis is an unusual condition; there are no evidence-based recommendations or guidelines for the treatment. If GCA is suspected in patients with severe complications, high-dose glucocorticoids are the mainstay of treatment and should be started immediately. (9, 24)When we began the corticosteroid pulse, our patient dramatically responded to the treatment in the form of a reduction in the inflammatory markers.

4 Conclusion

Due to the noisy symptoms in patients with Stroke and carotid dissection, the diagnosis of GCA may be neglected as an underlying cause. When diagnosing stroke in elderly patients, based on the symptoms and paraclinical investigations, we should start treatment immediately before further diagnostic procedures if we suspect GCA. The association of high C-reactive protein levels and the rate of erythrocyte sedimentation rate with stroke, although nonspecific, should draw some attention to vasculitis, topped by GCA. In these cases, before biopsy, we can use superficial temporal artery ultrasound as an aid.

Declarations

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Conflicts of interest

The authors declare that there is no conflict of interest.

Ethics Approval

All procedures conducted herein were in accordance with the ethical standards of the institutional and national committees on human experimentation and the 1964 Helsinki Declaration, and later versions.

Consent to Participate

Written and orally informed consent was obtained from the patient.

Consent for Publication

Written and orally informed consent was obtained from the patient to publish images and case report.

Data Availability Statement

The main contributions provided in the study are included in the article; further questions can be referred to the relevant author/authors.

Code Availability

Not applicable

Author Contributions

1. Study concept and design: Leila Hashami, Arsh Haj Mohamad Ebrahim Ketabforoush, Matineh Nirouei
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The full article was not posted or published elsewhere.

References

1. Salvarani C, Giannini C, Miller DV, Hunder G. Giant cell arteritis: Involvement of intracranial arteries. Arthritis Rheum. 2006;55(6):985-9.

2. Borchers AT, Gershwin ME. Giant cell arteritis: A review of classification, pathophysiology, geoepidemiology and treatment. Autoimmunity Reviews. 2012;11(6):A544-A54.

3. Lu-Emerson C, Walker M, Huber BR, Ghodke B, Longstreth WT, Khot SP. Lethal giant cell arteritis with multiple ischemic strokes despite aggressive immunosuppressive therapy. Journal of the Neurological Sciences. 2010;295(1):120-4.

4. Zwicker J, Atkins EJ, Lum C, Sharma M. An atypical presentation of giant cell arteritis. Canadian Medical Association Journal. 2011;183(5):E301-E5.

5. Tang V, Fantaneanu T, Chakraborty S, Patel V, Dowlatshahi D. Intracranial Non-Occlusive Thrombus and Multiple Strokes in Giant Cell Arteritis. Canadian Journal of Neurological Sciences / Journal Canadien des Sciences Neurologiques. 2014;39(1):116-7.

6. Pariente A, Guédon A, Alamowitch S, Thietart S, Carrat F, Delorme S, et al. Ischemic stroke in giant-cell arteritis: French retrospective study. Journal of Autoimmunity. 2019;99:48-51.

7. Banz Y, Stone JH. Why do temporal arteries go wrong? Principles and pearls from a clinician and a pathologist. Rheumatology. 2018;57(suppl_2):ii3-ii10.

8. Samson M, Jacquin A, Audia S, Daubail B, Devilliers H, Petrella T, et al. stroke associated with giant cell arteritis: a population-based study. Journal of Neurology, Neurosurgery & Psychiatry. 2015;86(2):216-21.

9. de Boysson H, Liozon E, Larivière D, Samson M, Parienti J-J, Boutemy J, et al. Giant Cell Arteritis–related Stroke: A Retrospective Multicenter Case-control Study. The Journal of Rheumatology. 2017;44(3):297-303.

10. Gonzalez-Gay MA, Vazquez-Rodriguez TR, Gomez-Acebo I, Pego-Reigosa R, Lopez-Diaz MJ, Vazquez-Triñanes MC, et al. Strokes at Time of Disease Diagnosis in a Series of 287 Patients With Biopsy-Proven Giant Cell Arteritis. Medicine. 2009;88(4).

11. Moghaddasi M. Giant cell arteritis with internal carotid artery stenosis and third nerve palsy: a case report. GLOBAL JOURNAL FOR RESEARCH ANALYSIS. 2015;4.

12. Cox BC, Fulgham JR, Klaas JP. Recurrent Stroke in Giant Cell Arteritis Despite Immunotherapy. The Neurologist. 2019;24(4):139-41.

13. Elhfnawy AM, Bieber M, Schliesser M, Kraft P. Atypical presentation of giant cell arteritis in a patient with vertebrobasilar stroke: A case report. Medicine. 2019;98(32):e16737-e.

14. Nuenninghoff DM, Hunder GG, Christianson TJH, McClelland RL, Matteson EL. Incidence and predictors of large-artery complication (aortic aneurysm, aortic dissection, and/or large-artery stenosis) in patients with giant cell arteritis: A population-based study over 50 years. Arthritis & Rheumatism. 2003;48(12):3522-31.

15. Wilkinson IMS, Russell RWR. Arteries of the Head and Neck in Giant Cell Arteritis: A Pathological Study to Show the Pattern of Arterial Involvement. Archives of Neurology. 1972;27(5):378-91.

16. Bogousslavsky J, Deruaz JP, Regli F. Bilateral obstruction of internal carotid artery from giant-cell arteritis and massive infarction limited to the vertebrobasilar area. European neurology. 1985;24(1):57-61.

17. Siemonsen S, Brekenfeld C, Holst B, Kaufmann-Buehler AK, Fiehler J, Bley TA. 3T MRI reveals extra- and intracranial involvement in giant cell arteritis. AJNR American journal of neuroradiology. 2015;36(1):91-7.

18. Parra J, Domingues J, Sargento-Freitas J, Santana I. Extensive intracranial involvement with multiple dissections in a case of giant cell arteritis. BMJ case reports. 2014;2014.

19. Bajkó Z, Bălaşa R, Moţăţăianu A, Bărcuţean L, Stoian A, Stirbu N, et al. Malignant Middle Cerebral Artery Infarction Secondary to Traumatic Bilateral Internal Carotid Artery Dissection. A Case Report. Journal of critical care medicine (Universitatea de Medicina si Farmacie din Targu-Mures). 2016;2(3):135-41.

20. Filep RC, Bajko Z, Simu IP, Stoian A. Pseudo-dissection of the internal carotid artery in acute ischemic stroke. Acta neurologica Belgica. 2020;120(2):469-72.

21. Bajkó Z, Maier S, Moţăţăianu A, Bălaşa R, Vasiu S, Stoian A, et al. Stroke Secondary to Traumatic Carotid Artery Injury - A Case Report. Journal of critical care medicine (Universitatea de Medicina si Farmacie din Targu-Mures). 2018;4(1):23-8.

22. Proft F, Czihal M, Rémi J, Fesl G, Woischke C, Koops H. Fatal Spontaneous Bilateral Vertebral Artery Dissection in Giant Cell Arteritis (GCA). Journal of Vasculitis. 2017;03.

23. Zheng X, Wang Y, Chen G, Ma C, Yan W, Chen M. Diagnosis of ischemic optic neuropathy caused by dissection of the internal carotid artery: A case report. Medicine. 2020;99(33):e20034-e.

24. Bienvenu B, Ly KH, Lambert M, Agard C, André M, Benhamou Y, et al. Management of giant cell arteritis: Recommendations of the French Study Group for Large Vessel Vasculitis (GEFA). La Revue de

medecine interne. 2016;37(3):154-65.

Figures

Figure 1

Diffusion weighted brain magnetic resonance imaging showing watershed infarct in right hemisphere.

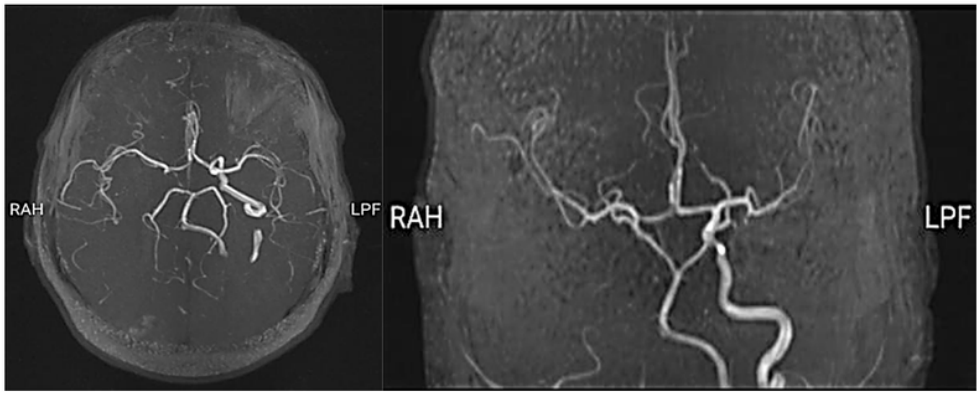

Figure 2

Magnetic resonance angiography showing occluded right proximal internal carotid artery.

Figure 3

Right carotid artery dissection on contrast-enhanced MR angiography (CE-MRA) (a) and CTA (b & c). Carotid artery dissection is characterized by a narrowed eccentric flow void, which is surrounded by a crescent-shaped, hyperintense mural hematoma expanding the outer vessel diameter

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