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Case Report

A story of thundeclap headache and stroke

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ABSTRACT

Giant cell arteritis causes large and medium vessel vasculitis that can involve the aorta and great vessels. We report a case of a 59-year-old female with no known comorbidity who presented with complaints of sudden onset, sharp, severe right-sided headache for 15 days which was gradually progressive and not responding to any painkillers followed by acute onset left-sided weakness. On examination, she had reduced power in the left upper and lower limb, right temporal, parietal scalp tenderness, and low-volume temporal artery pulsation. MRI and MRA of the brain revealed a subcortical infarct in the right frontal subcortical region with normal cerebrospinal fluid findings. His ESR and CRP levels were raised and her hemoglobin level was low (vasculitis markers and other stroke workups were non-contributory). VEP showed moderate to severe retino-optic pathway dysfunction predominantly demyelinating type. We started aspirin, atorvastatin, and prednisolone as we suspected Giant cell arteritis (GCA). GCA is an unusual cause of ischaemic stroke (stroke prevalence is around 1.5% - 11% in GCA cases). In our case, the patient with no history of headache or stroke presented with ischaemic stroke in her very first presentation and was diagnosed with Giant Cell Arteritis. The headache was acute at onset and the stroke was in the anterior circulation, which is relatively uncommon. We need to consider the possibility of GCA, especially in this age group, and, after ruling out other possibilities, should not delay starting corticosteroids in these patients to avoid catastrophic consequences, including blindness.

1. Introduction

Giant Cell Arteritis (GCA, also known as Horton's disease, cranial arteritis, and temporal arteritis) is a type of vasculitis of large and medium-sized vessels because it can involve the aorta and great vessels.[1] GCA usually presents with symptoms such as fever, fatigue, weight loss, headache, jaw claudication, visual symptoms; particularly transient monocular visual loss (TMVL or Amaurosis Fugax), diplopia and symptoms of polymyalgia rheumatica (PMR). GCA appears almost only after the age of 50 years and its incidence increases constantly with advancing age, reaching a peak between 70 and 80 years. It requires a high level of clinical suspicion to diagnose, where classical signs and symptoms are not present.

2. Case Presentation

59 years old female patient presented with complaints of sudden onset, sharp, severe right sided headache with a pain score of 9/10 from 15 days prior to presentation. The headache was gradually progressive, was localised over the right temporal parietal and occipital region, sharp and was not responding to paracetamol or any other NSAIDS. It was not associated with photophobia, phonophobia, nausea, vomiting, visual disturbances, tearing from eyes, or loss of consciousness. There was a history of recent low-grade intermittent fever a few days before the onset of headache.

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The patient also had sudden onset left upper limb and lower limb weakness, along with generalised fatigue seven days after the onset of headache. She did not have any comorbidities and was not on any regular medication. She denied any history of joint pain, rashes, any symptom of headache or similar illness in past. On examination, she had the right temporal-parietal scalp tenderness, temporal artery pulsation had low volume. Power in left upper limb and lower limb was 4/5 along with weak left-hand grip. Visual field examination appeared to be normal and other systemic examination were non-contributory. Patient underwent MRI brain which showed sub-acute infarct in the right frontal subcortical white matter region along with chronic small vessel ischaemic changes (the MR Angiogram with vessel wall imaging was within normal limit). Echocardiography and 24 hours Holter monitoring were non-contributory. We did a carotid doppler study which showed normal carotid and vertebral arteries flow. Blood investigations showed low hemoglobin levels (8gm/dl), ESR of 59, CRP 9.6, other blood parameters related to stroke evaluation was normal (evaluation for diabetes, dyslipidaemia, thyroid disorder, hyperhomocysteinemia, liver and renal function tests were within normal limits). Her low hemoglobin levels were investigated, which showed low iron levels along with raised reticulocyte count. Hemoglobin electrophoresis suggested HB-E heterozygous state. Possibility of systemic infection was ruled out (with relevant investigations including cultures). The detail cerebrospinal fluid study was non-contributory. Temporal artery doppler study was done, which was within normal limits. ANA, ANCA were all negative. Patient was started on antiplatelet (aspirin 75 mg) and atorvastatin 80 mg, however there was no relief to headache. She underwent VEP (Visual Evoked Potential), which showed moderate to severe retino-optic pathway dysfunction, predominantly demyelinating type. We suspected Giant Cell Arteritis (GCA) and started her on prednisolone 60mg. She showed remarkable improvement: headache subsided, fever did not return and she never felt fatigued (symptoms started improving from two days of treatment).

Table 1: American college of rheumatology criteria for giant cell arteritis

S.No.	Criterion	Definition
1	Age at onset >50 years	
2	New headache	New onset of/new type of pain in the head
3	Temporal artery abnormality	Temporal artery tenderness on palpation/ decreased pulsation, unrelated to cervical atherosclerosis
4	Increased ESR	ESR > 50 mm/hr (Westergren method)
5	Abnormal artery biopsy	Biopsy showing vasculitis characterised by predominant mononuclear infiltrate or granulomatous inflammation

3. Discussion

GCA is an unusual cause of ischemic stroke. In a population-based stroke registry made by Wiszniewska M, Devuyst G and Bogousslavsky, it was found that only 0.15 percent of 4086 cases of ischemic stroke satisfied criteria for the diagnosis of GCA.[1]

Strokes because of GCA can occur in the distribution of both the internal carotid and vertebrobasilar arteries, however they are more common in the latter location - more than one-half of strokes can be because of GCA occurring in the vertebrobasilar system.[2] Involvement of the vertebral arteries can cause patients presenting with vertigo, ataxia, dysarthria, homonymous hemianopsia, or bilateral cortical blindness.[3] In bilateral vertebral artery involvement, a rapidly progressive brainstem and/or cerebellar neurologic deficits with high mortality, suggests GCA.[4] Very few studies have reported the involvement of intracranial vessels in patients with GCA. In a study done with 463 patients having a clinical diagnosis of central nervous system vasculitis at the Mayo Clinic, only 2 patients had firm evidence of intracranial GCA.[5] In a subsequent case series of 185 patients with GCA evaluated at the Mayo Clinic, only 9 cases of GCA with intracranial involvement were identified.[6]

In a series of 2000 consecutive stroke patients from Spanish hospital, temporal arteritis was an infrequent diagnosis among ischemic stroke of unusual cause (5.7% of cases, 4 of 70 patients). The major findings were a high incidence of neurologic deficits and poor prognosis despite treatment. Other clinical cases of intracranial involvement in the setting of GCA are described,[7] and 3 Tesla (3T) magnetic resonance imaging (MRI) has shown vessel wall enhancement of intradural arteries in a few GCA related strokes.[8] In our case, we can see that our patient had no history of any underlying comorbidities, which would increase the risk of atherosclerosis and ischaemic CVA, nor could we find out any other stroke mechanism.

She presented with unilateral headache and right sided weakness in her very first presentation. MRI of the brain revealed subacute frontal infarct, we started her with antiplatelet and statin. However, while evaluating for the cause of stroke, we could not delineate any specific reason for the findings of increased ESR and CRP along with sharp right sided headache which raised our doubts regarding the possibility of underlying vasculitis. Keeping Giant Cell arteritis in mind, we performed a doppler study of the temporal artery, which however did not show the classical halo sign (sensitivity of 67% and specificity of 95% in GCA).[9]

Temporal artery biopsy is considered the gold-standard investigation for the diagnosis of GCA, revealing necrotizing arteritis with a predominance of mononuclear cell infiltrations or granulomatosis with multinucleated giant cells. However, in all cases, we might not get a positive temporal artery biopsy report or it may not be workable to perform a biopsy.

So, the classification of GCA developed by the American College of Rheumatology in 1990 includes five criteria: Age of 50 years or older, new-onset headache, temporal artery tenderness or decreased pulsation, erythrocyte sedimentation rate of at least 50 and abnormal temporal artery biopsy results. If at least three five criteria are present in a single case, the diagnosis can be made with a sensitivity of 93.5% and a specificity of 91.2%. And in our case, 4 out of 5 criteria[4] were satisfied, so we made the diagnosis of GCA.

The pathophysiology of ischaemic CVA in cases of GCA can result because of arterial inflammation, which can lead to intimal thickening, luminal irregularities, stenosis, and ultimately leading to occlusion, which may cause infarction or hypoperfusion in watershed territories. Distal occlusion or embolization may also occur because of thrombosis secondary to dissection of artery. Temporal arteritis, other types of inflammatory vasculitis resulting in cortical and watershed infarcts may be other types of strokes.[10]

We started her with corticosteroids in view of "Temporal Arteritis" being a high possibility. She showed marked improvement after receiving corticosteroids and there was even a considerable amount of improvement in her right sided weakness. Steroids are the mainstay treatment option for giant cell arteritis; however, there are no definite guideline which suggest their role in treating the ischaemic cerebrovascular events secondary to giant cell arteritis. Our patient came with ischaemic CVA in her very first presentation and was diagnosed with giant cell arteritis while we were evaluating for the cause of her underlying vascular event. Another important thing to note is that the patient had acute onset headache and an anterior circulation stroke, which is a relatively rare finding in GCA associated strokes (stroke prevalence is around 1.5% - 11% in GCA cases).

4. Conclusion

It is important to do a thorough stroke work up, and in all cases where risk factors are not present, it is imperative to rule out the various rare causes of CVA (like vasculitis including GCA when a new onset unilateral headache is in an association of stroke). In this age group (more than 50 years) possibility of giant cell arteritis must be considered and thoroughly evaluated and there should be no delay in initiating corticosteroids as it may progress to catastrophic consequences, including blindness (our patient already had changes in VEP). On follow up after six months, she is doing extremely well, and we have decreased dose of steroid and added methotrexate.

5. Source of Funding

None.

6. Conflict of Interest

None.

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