www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 83.27
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i8.173

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Giant Cell Arteritis, a Headache that Blinds -Case report

Authors
Navaneeth G[1], Deepak Rajendran[2], Madhu.G.Kandathil[3], Arathi.N[4], Suresh Raghavan[5], Shameem K Ummerali[6]
[1,2]Junior Resident, [3,4]Lecturer, [5]Professor, [6]Associate Professor
Department of General Medicine, Govt.T.D Medical College, Alappuzha, Kerala, India
*Correspondence Author
Navaneeth G
Junior Resident, Dept of General Medicine, Govt. T.D Medical College, Alappuzha, Kerala, India-688005
Email: *navaneethg45@gmail.com*

ABSTRACT
Giant cell arteritis is a rare systemic granulomatousvasculitis affecting large- and medium-sized arteries.. Here we present a case of GCA with history of seizures, recurrent cerebrovascular accidents, ischemic heart disease and recent complaints of severe persistent headache and visual loss. This case highlights the importance of assessing the possibility of giant cell arteritis through appropriate clinical history, estimation of acute phase reactants and the judicious use of superficial temporal artery biopsy, to clinch the diagnosis.

INTRODUCTION

Vasculitis is a clinico-pathological process that is characterized by inflammation and damage to the blood vessels.The vessel lumen will be compromised and this will be associated with ischemia of the tissues supplied by the blood vessel. Vasculitis can be a primary vasculitis syndrome or due to a secondary cause. Giant cell arteritis is one such primary vasculitis syndrome affecting the large sized blood vessels with well-developed wall layers and adventitial vasa vasorum. The vascular beds that are usually affected include the external carotid branches (eg., temporal and occipital arteries), the ophthalmic, vertebral, distal subclavian and axillary arteries and the thoracic aorta. Giant Cell Arteritis occurs only in older adults, mainly those over age 50, may occur together with polymyalgia rheumatica. The most dreaded complication is ischemic optic neuropathy which causes vision loss in 10-15% of patients

CASE REPORT

A 58 year old male, manual labourer,ex-smoker,occassional alcoholic with past history of Type 2 DM, Hypertension, coronary artery disease, seizure and recurrent ischemic strokes. The first stroke occurred 5 years back causing left hemiparesis- Right MCA territory infarct and the second stroke 2 years back involving the same vascular territory. The patient presented with history of right sided headache, throbbing in

nature associated with numbness of right half of head andblurring of vision of both eyes for the past two weeks. Head ache was persistent and was not subsiding with the usual dose of analgesics. No history of fever, vomiting, photophobia, lacrimation or trauma. There is only minimal residual weakness because of the old CVA. He also gives history of severe body pain, malaise and loss of appetite.

On examination, his vitals were normal. General physical examination showed bilateral mature senile cortical cataract. There was no evidence of any neuro cutaneous markers or cutaneous angiomatosis.

Nervous System examination showed normal Higher Mental functions. Cranial Nerve Examination- On examining the 2nd Cranial nerve, hisvisual acuity was counting finger close to face in the Right eye and perception of only hand movements in the left eye. Other Cranial Nerves were within normal limits. There was no evidence of Horner's syndrome. Motor system examination showed left hemiparesis. There was no sensory deficit .But cerebellar signs were positive on the left side. There was no neck stiffness or other signs of meningeal irritation. Skull and Spine were normal

Examination of other systems was normal.

Clinically we suspected the possibility of a vasculitis in view of recurrent attacks of stroke, seizure and a short duration of severe headache followed by vision loss. However other possibilities like Cerebrovascular disease due to atherosclerosis aggravated by Diabetes, Hypertension, Smoking and Age affecting the Internal Carotid Arteryis also considered.

His routineblood examination revealed an ESR value of 70. FBS and PPBS were 140 and 190 respectively. LFT, RFT, lipid profile, TFT, serum electrolytes, urine routine were normal.HbA1C was 7. Peripheral Smear showed normocytic normochromic anemia. C - Reactive protein was 20.HIV, HBsAg, HCV, VDRL were negative. Coagulation parameters were also normal. Serum Electrophoresis, p-ANCA,c-ANCA, ANA profile, serum ACE were also normal. RA factor, Anti CCP were negative.

His Chest X-ray was normal and ECG showed evidence of old inferolateral ischemia. USG Abdomen showed fatty liver changes and renal artery Doppler was normal. An ECHO Cardiogram was done and it showed global LV hypokinesia with moderate systolic and diastolic dysfunction.

Upon further investigations, CT scan showed an acute infarct in cerebral cortex in multiple areas.MRI brain showed acute infarcts in the cerebral cortex with old infarct in the left cerebellum. In MR Venogram, there was no evidence of acute dural venous sinus thrombosis. MR angiogram showed azygous Anterior Cerebral Artery with hypoplastic Right Vertebral Artery with no obvious aneurysms.

MRI of the orbit and Paranasal sinuses showed normal optic nerve and sinusitis.

A CSF study was done and there was no evidence of acute or chronic meningitis.

Ophthalmoscopic evaluation showed evidence of retinal ischemic changes.

The diagnosis was confirmed by taking a Right Temporal Artery Biopsy which was consistent with giant cell arteritis.

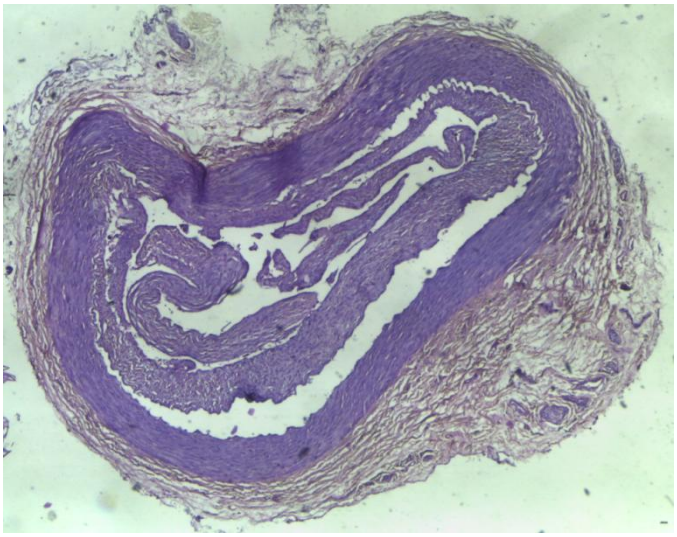

Figure 1: Temporal artery biopsy-Section shows blood vessel wall with focal infiltration by lymphocytes, few macrophages and occasional giant cells. Vascular lumen shows cholesterol clefts, consistent with giant cell arteritis.

DISCUSSION

The first clinical description of GCA was made by Hutchison in 1890. The typical pathology in the temporal arteries was described by Horton *et al.* in 1930. GCA occurs primarily in patients over the age of 50.Thisincidence increases with agewith GCA being almost 10 times more common among patients in their 80s than in patients aged 50-60.GCA is twice more common among women than men. Siblings of a patient with GCA are at increased risk (10 fold) of getting the disease. GCA has been most commonly reported in whites of Northern European descent. Epidemiological studies suggest that the incidence of GCA in Blacks, Hispanics and Asians is not as rare as once thought.

Most patients will present with one of four presentations:-

1) 20% cranial symptoms with superficial headache, scalp tenderness, jaw and tongue claudication and rarely scalp necrosis, diplopia or blindness
2) 40% polymyalgia rheumatic (PMR) with pain and stiffness of proximal muscle groups such as neck, shoulders, hips and thighs. Muscle symptoms are usually symmetric
3) 20 % both cranial and PMR symptoms
4) 15 % fever and systemic symptoms without any localized symptoms. Patients can present with a fever of unknown origin

5 % other symptoms like cough, claudication (upper > lower extremity) or synovitis

Onset of symptoms may be acute or insidious. Most patients have fever (40%), weight loss (50%), fatigue, malaise, as nonspecific symptoms

Visual loss occurs in 15% of patients. It can be an early symptom and is most commonly due to Ischemic Optic Neuritis (posterior ciliary branches of Ophthalmic Artery).Condition is abrupt and painless. Retinal artery and Ophthalmic Artery thrombosis also lead to blindness. Other eye manifestations are blurring of vision, amaurosis-fugax, iritis, conjunctivitis, photophobia and ophthalmoplegia from ischemia of extra ocular muscles.

Neurological complications of GCA are involvement of Internal Carotid and Vertebral artery, leading to strokes, seizure, acute hearing loss, vertigo, cerebral dysfunction and depression. Involvement of Intracranial arteries is unusual because they lack internal elastic lamina.

Even though it is classically a Large vessel vasculitis, involvement of medium sized vessels can also occur. Involvement of Carotid Artery can lead to stroke, seizure, Ophthalmic artery leads to visual loss, Subclavian artery lead to upper limb claudication, Renal artery lead to Hypertension, Mesentric Artery lead to intestinal ischemia, Iliac artery lead to Limb claudication, Coronary Artery to Ischemic heart diseases. But involvement of Pulmonary Artery is extremely rare

The dendritic cells residing within the vessel wall initiate the inflammatory cascade that recruit T cells and macrophages to form granulomatous infiltrates. Inflammatory cellular infiltrates are usually concentrated around inner half of media. Whereas in Takayasu arteritis tends to localize adventitia and outer parts of media, including vasa vasorum

When ESR is normal, an elevated CRP is useful in detecting an acute phase response. Diagnosis is confirmed by taking Temporal Artery Biopsy. In general biopsy should be obtained within 7 days of starting corticosteroid therapy whenever possible.

Sudden blindness and other stroke like events have occasionally being reversed in patients by institution of high dose corticosteroid therapy (1 g IV methyl prednisolone daily for 3 days) if started within 24 hours. Corticosteroids are dramatically effective in suppressing systemic symptoms of GCA within 72 hours after initiation of therapy. Localised manifestation of arthritis, headache, scalp tenderness jaw or tongue claudication steadily improve over a longer period of time. Patients typically die of vasculitis complications like Stroke/MI. The risk of death from GCA appears to be increased within 1st 4 months of

therapy. After 4 months the mortality is similar to that of age matched general population except there is an increased prevalence (17 times) of thoracic aortic aneurysm and aortic dissection. Patients with GCA should be followed for development of new AR murmur. If one develops, further investigation of aneurysm should be undertaken. Surgery is considered when aneurysm enlarge to >5 cm or dissect.

CONCLUSIONS

In this patient with previous 2 episodes of stroke, IHD, seizure, vision loss it is suggestive of a multiple arteritis- possibly due to Giant Cell Arteritis. The very short history with high ESR and sudden loss of vision well fits with Giant Cell Arteritis. Atherosclerotic occlusive artery disease producing a visual loss and severe head ache is extremely rare. But the atypical feature in this case is that the classical description of a tender nodular non pulsatile temporal artery is absent here.