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Case Report

# Bilateral Vertebrobasilar Insufficiency Secondary to a Biopsy-Proven GCA: A Case Report with Literature Review

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## Abstract

Giant Cell Arteritis (GCA) is the commonest form of systemic vasculitis in elderly patients of North American and European descent. Stroke in GCA is uncommon and under-recognized. We report a case of a 70-yearold male who had posterior circulation stroke and active Polymyalgia Rheumatica (PMR) at the time of GCA diagnosis. He presented with acute hemiparesis, ataxia, slurred speech and blurred vision associated with a few weeks history of hemicranial headache, myalgia, and abnormal acute phase response. Neuroimaging revealed multi-territory infarcts involving midbrain and both occipital cortices. CT angiography showed bilateral stenosis of vertebral, basilar and posterior cerebral arteries. Temporal artery biopsy confirmed classic changes of GCA. 14 days into high dose aspirin and glucocorticoids, the patient developed another posterior stroke. He survived with moderate cognitive, physical and functional dependence. This is one of the few available reports in literature denoting intracranial arterial involvement in GCA.

## Background

Giant Cell Arteritis (GCA) is an immune-mediated vasculitis affecting large and medium-sized vessels with a tendency to involve vascular beds of the external carotid branches (e.g., temporal and occipital arteries), the vertebral, ophthalmic, distal subclavian, axillary arteries, the thoracic aorta, and typically spare intracranial vessels [1,2]. It almost exclusively affects people over 50-year of age with a peak incidence between 70-80 years, particularly females [3]. The end result is usually ischemia due to luminal narrowing ± occlusion or aneurysm formation secondary to vascular wall inflammation. There is typically no associated autoantibody production and it is currently regarded as a T-cell mediated disease [4].

GCA commonly presents with headaches, scalp tenderness, ophthalmic manifestations or Polymyalgia Rheumatica (PMR) with elevated acute phase reactants [5]. GCA - related acute stroke presentation is uncommon and tends to be overlooked, as it tends to be associated with a lower acute phase response. Timely institution of high dose systemic glucocorticoid therapy is imperative to prevent irreversible ischaemic complications, most commonly sight loss. Atherosclerosis and hypertension remain the most common causes of acute ischemic stroke in elderly patients, which is also a risk factor for GCA, which contributes to the difficulty in discriminating such cases appropriately during emergency presentations. Theoretically, instituting fibrinolytic therapy in acute ischemic stroke related to inflamed, fragile and possibly aneurysmal vessels can have hazardous consequences with unfavourable outcome. In this report, we present a case of a biopsy-proven GCA masquerading as bilateral posterior circulation stroke shortly following sub acute presentation of PMR.

## Case Presentation

A 70-year old man with good pre-morbid physical function and no added cardiovascular risks attended our emergency department with a 3-hour history of acute right-sided upper and lower limb pyramidal weakness. This was accompanied by unsteadiness, dizziness and visual blurring to his right eye. He denied jaw claudication or diplopia.

On arrival, his triage observations reported Blood Pressure (BP) of 170/110, sinus rhythm of 81 beats per minute, normal temperature and glucose level. The neurological examination confirmed the MRC muscle power scale of 2/5 to his right upper and lower limbs together with a toxic gait and right superior temporal quadrantanopia [6]. Speech and language were intact. There were no ophthalmoplegia, bulbar muscle weakness, and extra pyramidal or dorsal tract signs. His right Temporal Artery (TA) was pulsatile and tender. The rest of the systemic examination was otherwise unremarkable. On further questioning, he admitted ongoing headaches, fatigue, early morning stiffness, shoulder and hip girdle myalgia for the last 6 weeks. He also reported right-sided mild to moderate headaches going back to a similar period and not feeling generally well. His past medical history included duodenal ulcer and open-angle glaucoma to his right eye. Initial labs showed elevated acute phase reactants (Table 1).

Computed Tomography (CT) of the brain and angiogram of major extracranial arteries on arrival excluded intracerebral, sub-dural or sub-arachnoid bleed or proximal thrombus. Acute fibrinolytic therapy was not considered as his pyramidal weakness was improving spontaneously to MRC power 3-4/5and was started on aspirin 300mg

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with high dose prednisolone 60mg/daily. Color doppler ultrasound of temporal arteries showed circumferential hypoechoic wall edema with vascular wall thickening (Halo sign) suggestive of temporal arteritis (Figure 1). Vascular duplex ultrasound showed < 50% stenosis to both carotids. Brain Magnetic Resonance Imaging (MRI) 3 days later revealed multi-focal infarcts involving the territory supplied by right posterior cerebral and left Posterior Inferior Cerebellar Arteries (PICA) (Figure 2). TA biopsy identified medium-sized arterial segment with extensive plasma cells and lymphocytes infiltrate to the media with multi-nucleated giant cells, associated fibrinoid necrosis and coexistent intimal fibrosis (Figure 3). The ophthalmologic assessment showed no evidence of ischemic optic neuropathy. The patient was making good progress until day 14 day of admission when he developed short-lived episodes of slurred speech which shortly became persistent and established. Repeat neuroimaging confirmed new interval right occipital infarction (previously left). Nocardio-embolic cause was revealed on subsequent work-up.

| Test | Result | Normal reference |
|---|---|---|
| C reactive protein | 91 mg/L | 0-5 |
| Erythrocyte sedimentation rate | 65 mm/hr | 0-25 |
| White cell count | 11.2^9/l | 3.5-11 |
| Hemoglobin | 15.2 g/dl | 13-17 |
| Mean cell volume | 84 fl | 80-100 |
| Platelets | 369^9/l | 150-400 |
| Urea | 6.0 mmol/L | 2.5-7.8 |
| Creatinine | 74 umol/L | 59-104 |
| Calcium (adjusted) | 2.36 mmol/L | 2.2-2.6 |
| Liver function tests | Normal | -- |
| Total cholesterol | 3.8 mmol/L | ESC guidelines* |
| Triglyceride | 1.5 mmol/L | ESC guidelines* |
| HDL cholesterol | 1.3 mmol/L | ESC guidelines* |
| LDL cholesterol (calculated) | 1.8 mmol/L | ESC guidelines* |
| HbA1c | 46 mmol/mol | 20-42 |
| Immunoglobulins (IgG, IgA, IgM) | Normal | -- |
| Serum protein electrophoresis | No monoclonal bands | -- |
| Anti-nuclear antibody | Negative | -- |
| Rheumatoid factor | Negative | -- |
| Antineutrophil cytoplasmic antibodies | Negative | -- |
| Anti-phospholipids antibodies | Negative | -- |

**Table 1:** Patient's laboratory profile.

*European Society of Cardiology guidelines on managing dyslipidaemia. DOI:10.1093/eurheartj/ehw272

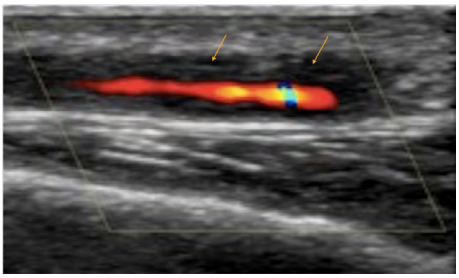

**Figure 1:** Colour Doppler longitudinal US image shows a circumferential non-compressible hypoechoic wall oedema (arrowhead) of the superficial temporal artery, known as "Halo" sign.

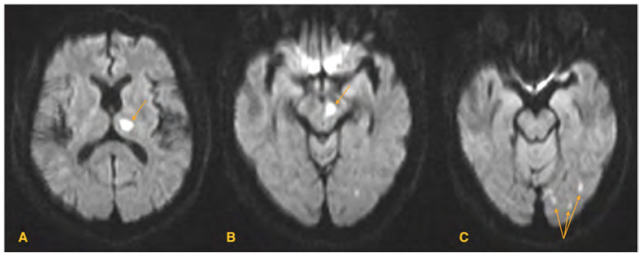


**Figure 2:** Diffusion-Weighted Magnetic Resonance Imaging (DWI-MR) of the brain shows infarcts (arrowheads) involving left thalamus (A), left midbrain (B) and left occipital lobe (C).

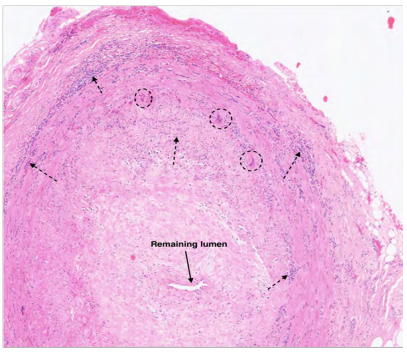


**Figure 3:** TA biopsy shows luminal narrowing, infiltrate with plasma cells and lymphocytes (broken arrowheads) and some of the multinucleated giant cells (circled).

## Investigations

Full biochemistry, hematology and immunology panel are listed in table 1. The most striking abnormalities were the bilateral tortious narrowing of the vertebral, posterior cerebral and basilar arteries (Figure 4) with raised inflammatory markers without clinical, radiological or microbiological evidence of sepsis. Echocardiography and Holter cardiac rhythm monitoring didn't reveal any thrombus or atrial fibrillation with good cardiac function. The inflammatory markers normalized on high dose glucocorticoid therapy (Figure 5). CT angiogram of the common, internal & external carotids, aorta and its major branches in thorax and abdomen did not reveal other vascular territory involvement.

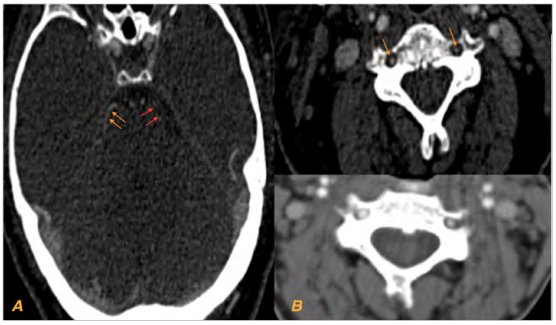


**Figure 4:** CT angiogram (A) showing narrowed right posterior cerebral artery (arrowheads) and non-visible left posterior cerebral (red arrowheads), CT angiogram (B) showing the patient's bilateral stenosed vertebral arteries at C4 level (Top) compared to normal looking arteries at similar level (bottom).

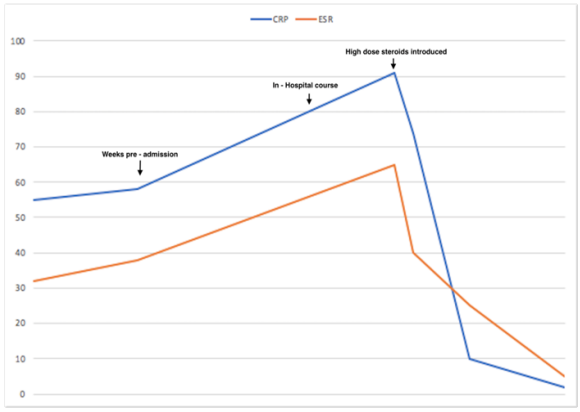


**Figure 5:** The course of acute phase reactants before, during and after treatment.

## Differential Diagnosis

The commonest culprit for such presentation is in situ thrombosis due to arteriosclerotic vascular disease or embolic phenomenon secondary to atrial fibrillation, however, they both don't explain the abnormalities of his multi-territorial vascular luminal stenosis. Cerebral amyloid angiopathy and primary systemic amyloidosis are also mimicking. Granulomatosis with polyangiitis and polyarteritis nodosa can occasionally involve temporal arteries [7,8].

## Treatment

The patient was maintained on high dose aspirin of 300 mg for 2 weeks followed by 75mg and prednisolone 60 mg/day for the following 4 weeks with gastric and bone protection followed by gradual glucocorticoid tapering. Clopidogrel 75mg was, arguably, added on following developing the new interval infarct. BP was controlled with 5mg Amlodipine. The patient received standard of care within the stroke unit including multidisciplinary team input and discharged then to the rehab unit.

## Outcome and Follow-Up

3 months later, the patient remained dysarthric with subtle receptive aphasia. He walks with one stick and requiring assistance to most of his daily living activities. His inflammatory markers remained normal. He was on 20 mg prednisolone andis continuing to down titrate. He is currently being considered for Ustekinumab as a steroid-sparing agent.

## Discussion

Stroke as atypical presentation or sequelae of GCA was frequently reported, however, population-based studies on this issue are lacking and therefore little information is currently available on the actual incidence of stroke in biopsy-proven GCA. The largest published report (Spain) on stroke at the time of GCA diagnosis had reported stroke in 8 out of 287 cases (2.8%) with biopsy-proven GCA (vertebrobasilar stroke = 7, carotid stroke = 1) [9]. The same study reported female gender, anemia and headache to be protective factors at the time of diagnosis while smoking increased the risk by 5 folds. Another study (France) on 97 biopsy-proven GCA cases reported stroke in 8 cases (8.2%) at the time of GCA diagnosis, with 100% involvement of vertebrobasilar territory [10]. Reports of 284 cases (Sweden)and 90 cases (England) with biopsy-proven GCA reported significantly increased the risk of vascular death one year after diagnosis, compared to age and gender-matched population, mainly due to cerebrovascular disease especially during the first 6 weeks to 4 months [11,12].

Stroke in GCA has a strong predilection to involve vertebrobasilar territory compared to carotid circulation [9]. A comprehensive but not systematic review of the literature revealed 100% involvement of vertebral arteries in all the examined reported cases [13-24]. In a post-mortem report, vertebral arteries were found to be affected throughout its length up to a point of 5mm above dura matter penetration, where the involvement ceases abruptly [25]. The same report revealed a strong correlation between the extent and severity of arterial involvement and the amount of elastic tissue in the media and adventitia of the arterial wall, a fact which could explain the relative sparing of the intracranial (intradural) arteries which have little or no elastic tissues in their outer two coats.

PICA is the largest and first branch of the vertebral artery within the skull, usually arises from the intradural segment [26]. It is postulated that their involvement in GCA is either through a direct extension of thrombus from vertebral arteries or direct embolization from diseased thrombosed extradural vertebral segments [25]. A fact which could explain the finding in our case. The intracranial arterial involvement in GCA although very uncommon but had been reported [10]. Our case was found to have involvement of basilar and bilateral symmetrical posterior cerebrals as shown on vascular imaging, a finding which is extremely uncommon.

In all the reported cases in the literature, the majority of strokes took place during the period of active disease as reflected by raised inflammatory markers, unexplained pyrexia, and the presence of other constitutional symptoms or by classic GCA symptoms [13-24]. This was considered the index period for instituting high dose systemic glucocorticoid therapy. Even with timely administration, deterioration and even fatal outcomes were frequently reported, either due to the extent of disease or to the fact that arterial inflammation is already well established by the time it is clinically manifest. Whilst glucocorticoids may control or prevent inflammation, they have a limited role during the thrombo-occlusive phase of the disease [27-30]. This is also the case regarding potential sight loss associated with GCA; when instituting immediate high dose glucocorticoid therapy the primary focus is to protect the contralateral eye rather than saving the affected one.

## Learning Points/Take Home Messages

- Stroke in GCA is uncommon, yet serious and potentially fatal complication and tends to involve vertebral arteries almost invariably
- Vertebrobasilar territory stroke needs careful assessment for other potential atypical aetiologies especially in the context of raised inflammatory markers
- Timely administration of systemic glucocorticoids can potentially result in a favorable outcome, yet irreversible ischemic complications or even fatal outcomes can take place in some cases

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