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Temporal arteritis presenting as hand motor cortex infarction: A report from India

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Abstract

This is the first Indian case report of a biopsy proven temporal arteritis that presented as acute ischemic stroke. The 60 year old woman presented with an isolated pure motor flaccid fractional weakness of the left distal hand, as a rare stroke chameleon due to isolated infarction of the 'hand motor cortex' area. The hand motor cortex infarction masquerades as 'pseudoperipheral palsy'.

Keywords: Stroke, Temporal arteritis, hand motor cortex, pseudoperipheral palsy, monoparesis, fractional weakness, temporal artery biopsy, magnetic resonance imaging, diffusion weighted imaging, cerebrovascular accident, ischemic infarction, stroke chameleon

INTRODUCTION

Temporal arteritis is an immune mediated chronic granulomatous vasculitis, mainly involving large and medium-sized vessels almost exclusively affects patients older than 50 years. Its prototype presentation is a typical vascular headache syndrome with neuro-ophthalmologic signs and symptoms. The diagnosis is supported by the American College of Rheumatology (ACR) criteria and the temporal artery ultrasonographic finding of the 'halo sign'; while the temporal artery biopsy is the gold-standard.[1,2]

Cerebrovascular accidents have rarely been reported in the published literature as presenting symptoms of temporal arteritis, only 3%-4% during the course of the disease.[3] To the best of our knowledge there are no previous reports of temporal arteritis associated stroke from India.[4] Isolated hand palsy is reported to occur in less than 1% of all ischemic strokes.[5] It is often mistaken for peripheral lesion, due to the strategic involvement of hand motor cortex (HMC), otherwise known as the cortical "hand knob" in the precentral gyrus.[6-8] We report here a temporal arteritis patient whose presentation is an isolated hand weakness from infarct of hand motor cortex. Informed patient consent was obtained for the publication of this case report and the accompanying images.

CASE REPORT

A 60-year-old woman presented with one day history of difficulty in moving the thumb, index, middle finger and the wrist of her left hand without sensory disturbances upon waking up in the morning after an uneventful night. She was on adequate and compliant treatment for Stage II primary hypertension and LDL hyperlipidemia for one with an otherwise unremarkable medical past history. With medical treatment, she achieved target blood pressure and LDL levels. However she did report a 2 months history of a non disabling mild left sided frontotemporal new onset vascular type headache with scalp allodynia.

Neurological examination revealed a moderate flaccid paresis of the left hand without wasting of muscles of the hand and wrist. The muscle weakness was worst in those innervated by the median nerve, followed by radial nerve and ulnar nerve, in a decreasing order. (Figure 1a, b,c) She had a 'ape thumb' deformity, a positive 'pen test', pincer grip weakness due to involvement of adductor pollicis and the first dorsal interossei, wrist drop and partial finger drop, pathological "circle sign" due to inability to oppose the tips of digits I and II due to weakness of flexor pollicis longus muscle and the flexor digitorum profundus muscle of the index finger. General physical examination was unremarkable. The neurologic

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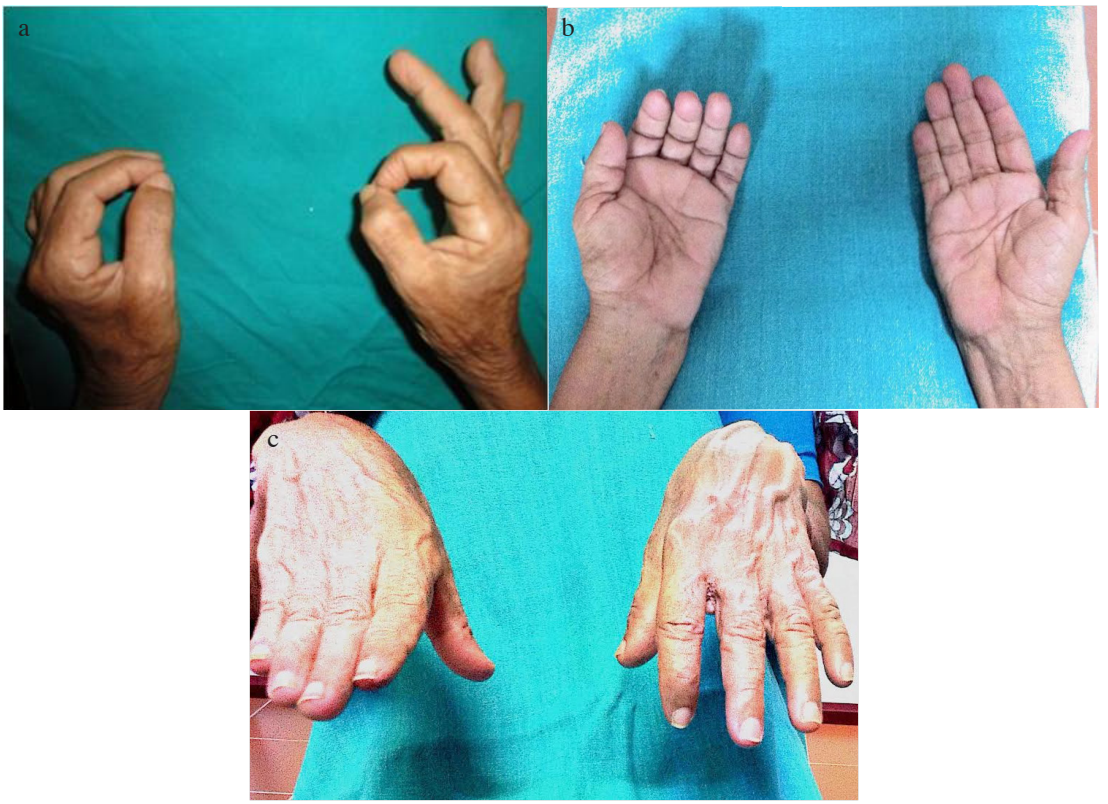

Figure 1. (a) The patient's left hand manifested with a high-grade flaccid paresis involving muscles of the hand and wrist innervated by the median nerve, followed by radial nerve and ulnar nerve, in a decreasing order of differential clinical involvement with the characteristic pathological "circle sign" due to inability to oppose the tips of digits I and II due to weakness of flexor pollicis longus muscle and the flexor digitorumprofundus muscle of the index finger; (b) demonstrating an ape thumb deformity and (c) a wrist drop.

examination did not reveal any other signs suggesting a central or upper motor neuron type of abnormality. The nerve conduction study done immediately upon admission was unremarkable. MRI Brain with DWI and MRA demonstrated a small diffusion restriction in a selective part of the right precentral gyrus that represents the hand motor cortex with the characteristic "omega sign" (Figure 2). The extracranial and intracranial MRA was unremarkable.

In retrospect, she also report a new onset right-side vascular headache over the preceding two months accompanied by right-sided scalp allodynia, and she scored three on the Wong-Baker Faces Rating Scale and 4 out of 10 in the Numerical Pain Rating Scale. In view of the association of the new onset left sided vascular headache and acute cerebrovascular accident, she was investigated for the usual stroke risk factors, including transthoracic echocardiogram, transesopahegal echocardiogram and a 24

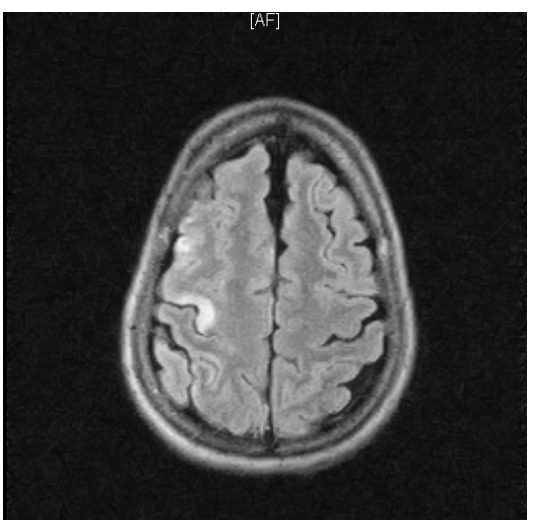

Figure 2. FLAIR magnetic resonance imaging (MRI) showing a well defined circumscribed acute infarction of the right precentral "hand motor cortex" area with the "omega sign"

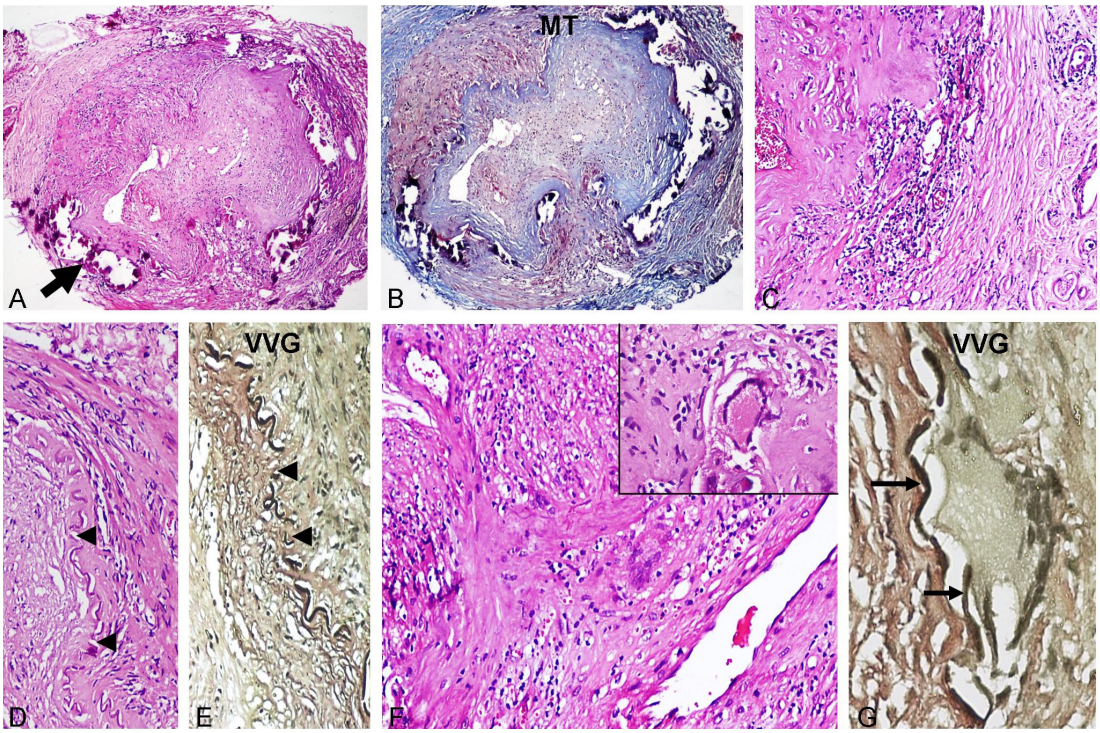

Figure 3 (a-g). Transverse section of temporal artery [A: H&E x40] shows organized luminal thrombi, recanalisation, subendothelial fibrosis, uneven thinning and thickening of tunica media [B: Masson's Trichrome (MT) x40] and medial calcification (thick arrow) [A: H&E x40]. Transmural lymphocytic infiltrate [C, F: H&E x100], multinucleated giant cells [F: H&E x100; inset H&E x400], fragmented internal elastic lamina (arrow heads) [D: H&E x100; E: Verhoff-Van Geison (VVG) x100] and attempted phagocytosis of internal elastic lamina by multinucleated giant cell [G: VVG x400] is seen.

hours Holter ECG monitoring. Since multiple measurements of blood pressure and other cardiovascular risk factors proved unremarkable, we considered the possibility of temporal arteritis as the cause of her isolated hand motor cortex ischemic stroke. Both superficial temporal arteries were not thickened. They were mildly tender and pulsations felt, with the opthalmoscopy and temporal artery Doppler to be unremarkable. All investigations proved unremarkable except for erythrocyte sedimentation rate (ESR) of45 mm/1st hour, elevated C-reactive protein (12 mg/L) with a thrombocytosis of 870 000 cells/mm.[3] She underwent a left temporal artery biopsy on the second day of admission that was consistent with temporal arteritis. (Figure 3) She was started on 1gm of pulse methylprednisolone for five days, followed by oral 1mg/kg/day oral prednisolone with osteoporosis protection, and triple therapy (aspirin 75mg, clopidogrel 75mg, atorvastatin 80mg stat, followed by 40g/day, and ramipril 10 mg per day).

There was slight improvement in her left hand motor weakness, and she was discharged after two weeks of hospital admission. Since then, she has been followed up for a year and a half and has made almost complete recovery of her pseudoperipheral weakness, with her being steroids tapered off. No new neurologic symptoms appeared during her follow-up.

DISCUSSION

We report here a patient with biopsy-proven temporal arteritis. The case was unusual in the occurrence of ischemic stroke presenting with hand weakness from involvement of the hand motor cortex. We believe that the ischemic infarct is related to the temporal arteritis. This was because there was no underlying cardioembolic source of stroke, as the transthoracic and transesophageal echocardiography, as well as 24 hours Holter monitoring were normal. The MRA was also normal. The patient had no further stroke after treatment with steroid. However, the patient did have atherosclerotic risk factors, which were elderly age, treated hypertension and hyperlipidemia. Nevertheless, we do acknowledge

that the vascular inflammation triggered by temporal arteritis could synergize with other cardiovascular risk factors to induce ischemic stroke. In a recent population based study, among the 57 biopsy-proven patients with temporal arteritis, 4 (7.0%) did experience a temporal arteritis-related stroke. Three were men and all had ≥2 vascular risk factors and were ≥80 years.[9] We thus felt justified to add antiplatelets and atorvastatin to her treatment, since both agents not only exerts antiplatelet effects, but also suppresses the various inflammatory mechanisms believed to cause damage to the arterial wall in temporal arteritis. This is in addition to ameliorating the atherosclerotic risk factors of advanced age, primary hypertension and hyperlipidemia.

This case is also unusual as temporal arteritis related stroke has not been hitherto reported from India. A study from India in 2015 documented only a total of 72 patients with temporal arteritis, none did present with a stroke phenotype.[4] Though the association of temporal arteritis and stroke is definite, it should be noted that overall transient ischemic stroke and stroke are uncommon in temporal arteritis, especially as the presenting symptom.[9-11]

The other unusual feature of our patient is the 'pseudoperipheral palsy' from selective HMC infarction. Indeed, to the best of our knowledge, an isolated ischemic infarction in the hand motor cortex in association with a biopsy proven temporal arteritis has not been previously reported in literature. Isolated weakness of the hand in either a pseudo-radial, pseudo-ulnar, pseudo-median, isolated index finger paresis, or as isolated thumb flexion weakness occur as an atypical, rather under recognized stroke phenotype, due to selective involvement of the hand motor cortex which has a well-recognized somatotopic arrangement in the primary motor cortex.[12-15] An accurate clinical history and familiarity with not only the typical but also the atypical clinical and radiographic findings of temporal arteritis can assist in establishing an early diagnosis, which enables prompt initiation of disease specific treatment.

Although the incidence of strokes related to temporal arteritis is exceedingly low, and the most common cause of strokes in the elderly population is atherosclerosis of the extracranial and intracranial arteries, we reiterate physicians and neurologists should be aware that temporal arteritis may be a cause of not only strokes, but also multi-infarct state with vascular dementia. In conclusion, we present here a 60 year old woman with biopsy-proven temporal arteritis presenting with ischemic stroke. The stroke manifested as an isolated hand weakness, from selective HMC infarction.

DISCLOSURE

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Conflict of interest: None

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