Large-vessel Giant Cell Arteritis: A Rare Cause of Stroke

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A 61-year-old man presented to the emergency with sudden-onset blurring of vision and dizziness for 6 h. The patient did not have conventional risk factors for stroke. His medical and family histories were insignificant. He had a history of bilateral shoulder and hip girdle pain with a significant duration of morning stiffness for 3 months. Shoulder and hip X-rays showed mild osteoarthritis. Neurological, peripheral vascular, and systemic examinations were unremarkable. Blood pressure was 160/80 mmHg. Electrocardiogram showed sinus rhythm, and routine blood tests were normal other than raised inflammatory markers (erythrocyte sedimentation rate [ESR] 60 mm/h, C-reactive protein [CRP] 68 mg/L). HbA1c, lipid profile, chest radiograph, and computed tomography (CT) of the brain were normal. His symptoms resolved spontaneously within 8 h. A diagnosis of transient ischemic attack was made, following which daily aspirin was started and follow-up arranged in the stroke clinic for further workup.

Two weeks later, he again presented with a left-sided hemiparesis. Higher mental functions and cranial nerve examination were normal. Power was 3+/5 in left upper and lower limb with hyperreflexia and extensor plantar responses. Scalp tenderness was absent, and temporal arteries were normally palpable. Cardiac auscultation was normal without peripheral vascular bruits or asymmetry of upper limb blood pressure. Repeat laboratory tests showed persistently raised ESR and CRP. Serology for ANA and ANCA was negative. Repeat CT brain showed a new lacunar infarct in the right corona radiata. Echocardiography and 24 h Holter electrocardiographic monitoring were normal. Carotid Doppler ultrasound showed moderately calcified atheroma in proximal internal carotid arteries with normal flow. Magnetic resonance imaging (MRI) brain with contrast showed multifocal deep white matter infarcts of various ages in the right corona radiata and left middle cerebellar peduncle [Figure 1]. CT angiogram of large vessels raised a possibility of vasculitis. The patient was started on warfarin and referred to the rheumatology clinic in view of possible large vessel vasculitis (LVV) on CT angiogram.

In the rheumatology clinic, a diagnosis of polymyalgia rheumatic (PMR) was established in view of the on-going symptoms of shoulder and hip girdle stiffness with

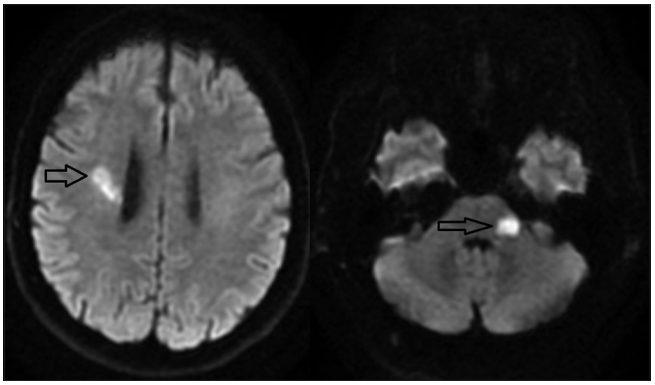

Figure 1: Right corona radiata infarct and left cerebellar peduncle infarct on MRI brain

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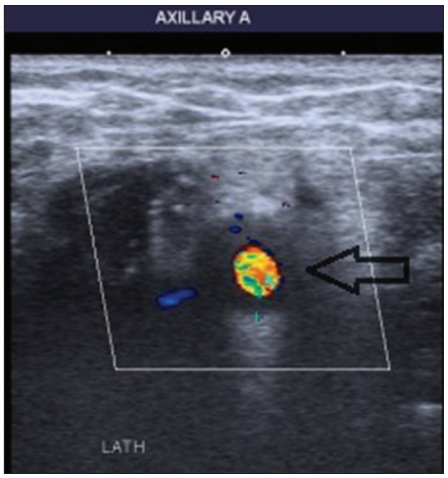

Figure 2: Halo sign on axillary artery ultrasound

raised inflammatory markers. Ultrasound of the axillary arteries showed bilateral "halos" [Figure 2], consistent with an active inflammatory process. To confirm the possibility of LVV, fluorodeoxyglucose-positron emission tomography (FDG-PET) scan was done which demonstrated increased uptake in the thoracic aorta, subclavian, axillary, and proximal common carotid arteries (bilateral), suggesting metabolically active vasculitis [Figure 3a-c]. Oral prednisolone 1 mg/kg/day with osteoporosis protection was commenced, and warfarin was changed to aspirin. Within a week, he reported a dramatic improvement in general well-being and resolution of constitutional and PMR symptoms with normalization of CRP and ESR. A tapering course of oral prednisolone was continued. He was asymptomatic at 3 months follow-up with the resolution of hemiparesis.

Age, hyperlipidemia, hypertension, atrial fibrillation, congestive heart failure, smoking, and diabetes are common risk factors for stroke.[1] Previous reports have shown an association between ischemic stroke and LV-GCA.[2,3] Although GCA characteristically involves temporal and ophthalmic arteries, large vessel involvement has been reported in 29%–83% of GCA patients in prospective studies depending on radiographic modality used for diagnosis. The predominant large arteries involved are ascending aorta, aortic arch, descending aorta, and subclavian and axillary arteries.[4,5]

The 1990 ACR classification criteria for GCA are inadequate for the diagnosis of LV-GCA. In the absence of typical cranial symptoms or temporal artery examination findings, temporal artery biopsy is less useful in diagnosis. Color Doppler ultrasound, CT/MRI angiography, and FDG-PET are useful for the diagnosis of LV-GCA.[4,5] FDG-PET has a sensitivity and specificity of 80% and 89%, respectively,

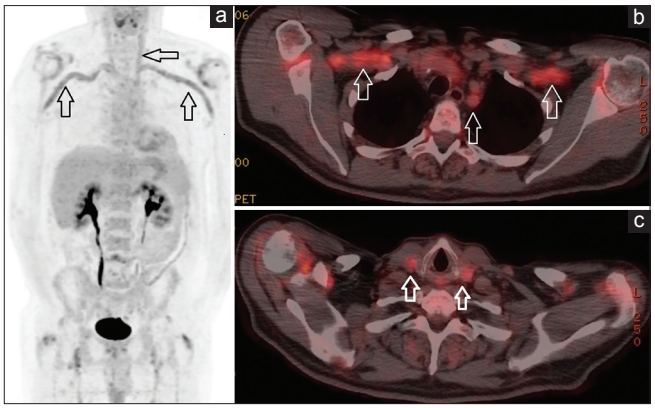

Figure 3: Fluorodeoxyglucose-positron emission tomography: Increased uptake in thoracic (a), subclavian, axillary (b), and carotid arteries (c)

for extracranial GCA and has emerged as an important tool for diagnosis.[6] High-dose oral corticosteroids (1 mg/kg/day) with dose tapering after resolution of clinical and laboratory abnormalities remain the mainstay of treatment in LV-GCA. Steroid-sparing therapies including methotrexate or azathioprine may be required for resistant cases.[4] Our case highlights the importance of detailed history and review of appropriate investigations for preventing rare causes of stroke being overlooked, especially in a population otherwise susceptible to stroke such as the elderly.

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Conflicts of interest

There are no conflicts of interest.

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