Posterior Reversible Encephalopathy Syndrome: An Atypical Presentation of Takayasu Arteritis

Dear Editor,

Takayasu arteritis (TA) is a systemic vasculitis causing granulomatous inflammation of the large- and medium-sized vessels with a predilection for the aorta and its major branches.[1] Various atypical presentations of TA have been described in the literature. Posterior reversible encephalopathy syndrome (PRES) as a presenting manifestation of TA is rarely reported in the literature.

A 15-year-old boy was admitted with severe headache and vomiting of 3 weeks duration. He also had three episodes of discrete generalized tonic-clonic convulsions and blurred vision from the last 2 weeks. His birth and development history were normal with unremarkable past and family history. On admission, he was conscious and oriented. He was afebrile with a regular pulse rate of 106/min and blood pressure of 180/110 mmHg in the right upper arm and 110/70 in the left upper arm. His peripheral pulse was asymmetrically reduced in the left radial, brachial, and carotid artery compared to the right side. There were no meningeal signs or focal neurological deficit. His visual acuity was reduced bilaterally with pupils equal and reactive to light. The fundus examination showed features of grade 4 hypertensive retinopathy bilaterally. The laboratory studies revealed normal blood counts, serum electrolytes, liver, and renal function tests. The erythrocyte sedimentation rate (ESR) was 22 mm in the 1st hour and his C reactive protein (CRP) was raised (59 mg/L). A non-contrast computed tomography (NCCT) of the brain was done initially which showed hypodense areas in the bilateral occipital lobes. He was managed with intravenous labetalol, amlodipine, levetiracetam, and mannitol. The blood pressure was controlled with improvement in headache and vision with no reoccurrence of seizure.

On further evaluation, the magnetic resonance imaging (MRI) of the brain showed a patchy hyperintense signal in the bilateral frontal, parietal, occipital, and left temporal white matter on T2 and fluid-attenuated inversion recovery (FLAIR) sequences [Figure 1a-d]. The echocardiography and electroencephalography were normal. The ultrasonographic examination of the abdomen further showed increased echoes in the bilateral kidneys with the small size of the left kidney. The computed tomography angiography of the aorta and renal arteries showed low attenuation concentric mural thickening of the proximal left common carotid artery [Figure 2a-b], left subclavian artery [Figure 2a-c], and left renal artery [Figure 3a-c], resulting in significant stenosis. There was also mild involvement of the infrarenal aorta [Figure 3d] with small kidney on left side [Figure 3e]. The child fulfilled the diagnostic criteria for TA as per EULAR (European League Against Rheumatism) and PReS (Pediatric Rheumatology European Society) consensus criteria.[2] He was started on oral glucocorticoids and methotrexate with a resolution of posterior reversible encephalopathy syndrome (PRES) finding on the MRI of the brain at 1 month after discharge.

PRES is a rare clinico-radiologic syndrome that usually manifests as an acute to subacute headache, confusion, visual disturbances, seizure, and encephalopathy with typical findings on neuroimaging.[3] PRES is often associated with

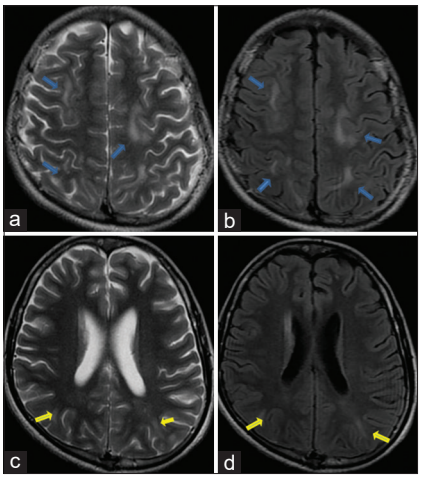

Figure 1: T2 and FLAIR MRI axial sections demonstrates hyperintense signal in bilateral frontal and parietal white matter in watershed zone (A and B, blue arrow) and bilateral occipital region (C and D, yellow arrow)

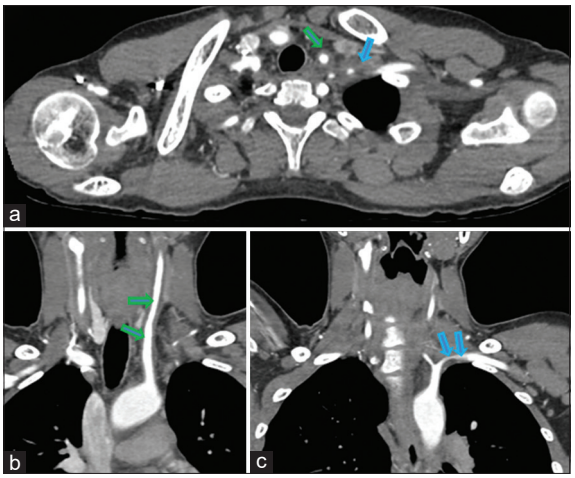

Figure 2: Axial post-contrast CT images showing concentric hypodense thickening with narrowed lumen of proximal left common carotid artery (A, green arrow) and subclavian artery (A, blue arrow). Curved planar reformation illustrates the narrowed lumen of left common carotid (B, green arrow) and subclavian artery (C, blue arrow)

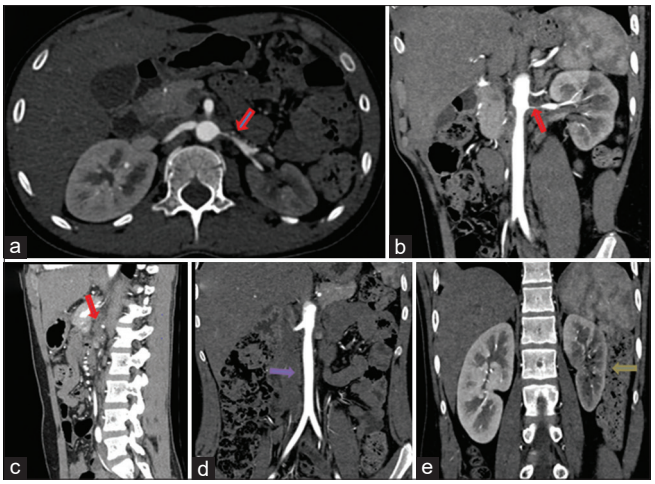

Figure 3: Axial post-contrast CT and curved planar reformatted images showing concentric soft tissue thickening of proximal left renal artery with significant narrowing (A, B &C, red arrow). Coronal reformatted images showing mild concentric smooth thickening with narrowing of infra renal abdominal aorta (D, purple arrow) and small left kidney with reduced cortical enhancement (E, brown arrow)

fluctuations in blood pressure, renal failure, eclampsia, vasculitis, and cytotoxic drugs. TA is a large vessel vasculitis causing panarteritis of the aorta and its branches. It is common in younger women of Asian descent.[4] The clinical presentation is usually nonspecific constitutional symptoms leading to delay in diagnosis.[4] Hypertension with asymmetrical pulses is a common presentation of TA, though it rarely presents as PRES.[5-9] Transiently impaired cerebral autoregulation explains the pathophysiology of PRES. The abrupt rise of blood pressure causes the failure of autoregulation of cerebral blood flow leading to dilatation of the cerebral arterioles and hyperperfusion. The differential vascular sympathetic innervation in the anterior versus posterior circulation causes increased susceptibility of posterior circulation for hyperperfusion and vasogenic edema during an acute rise in the blood pressure.[3] Endothelial injury, secondary to vasculitis or inflammation, disrupts the blood–brain barrier leads to vasogenic edema.[6] TA is associated with hypertension and endothelial dysfunction and both features lead to PRES in TA.

The 2006 EULAR/PReS consensus criteria for the diagnosis of TA requires angiographic abnormalities of the aorta or one of its major branches plus one or more of claudication or decreased peripheral artery pulses; blood pressure difference >10 mm Hg; bruits of the aorta or its major branches or hypertension.[2] Our patient had angiographic abnormalities in the form of subclavian artery and renal artery stenosis with the presence of hypertension, decreased peripheral pulsation, and blood pressure difference >10 mmHg.

The typical findings of PRES on the MRI of the brain are best appreciated on T2 and FLAIR sequences that show symmetric posterior hyperintense signal in the parieto-occipital region at the cortical-subcortical junctions irrespective of the underlying etiology. The frontal lobe, temporal lobe, cerebellum, basal ganglia, and brainstem are the other sites involved. The MRI lesions represent vasogenic edema, which is usually reversible. Various MRI patterns have been described. The most common being the parietooccipital pattern, holohemispheric watershed pattern, and superior frontal sulcus pattern.[3]

PRES is a benign reversible condition, but a delay in the diagnosis and treatment can lead to irreversible damage of the brain, that is, ischemia or infarction, which leads to chronic complications like permanent vision abnormalities, focal neurological deficit, and chronic epilepsy. The management of PRES in TA includes early and adequate control of blood pressure along with immunosuppressive agents in the form of high-dose oral daily glucocorticoids with the addition of non-glucocorticoids in the form of methotrexate, azathioprine, or tumor necrosis factor inhibitor.[6] The anti-seizure drugs and anti-edema measures are used as supportive management in PRES. Methotrexate is the preferred immunosuppressive agent in children.[10] The indications for surgical intervention in TA are very limited and usually indicated in cases of vascular changes in the new territory with active disease on imaging and progressive ischemia.

This case highlights the importance of early recognition and management of PRES and its underlying etiology, like, TA which results in a better outcome for both PRES as well as TA and prevents permanent damage of the brain and its sequel.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published

and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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