

An Uncommon Presentation of Takayasu Arteritis: A Diagnostic Challenge with Posterior Reversible Encephalopathy Syndrome

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Abstract

Takayasu arteritis (TA), a large-vessel vasculitis affecting young females more, may present with diverse clinical manifestations. This case report describes an 11-year-old girl who presented with headache, vomiting, bilateral ocular deviation, and altered sensorium. Clinical examination revealed hypertensive emergency and grade IV papilledema. Brain imaging demonstrated white-matter edema in the occipital and parietal lobes, suggestive of Posterior reversible encephalopathy syndrome (PRES). Color Doppler of abdominal vessels and computed tomography angiography revealed left renal artery stenosis, leading to the diagnosis of TA as per European League Against Rheumatism/ Pediatric Rheumatology International Trial Organization consensus criteria. This case highlights the rare association between TA and PRES, emphasizing the importance of considering underlying vasculitis in children presenting with hypertensive emergencies and neurological symptoms. The patient was managed with antihypertensive and immunosuppressive agents, including corticosteroids and mycophenolate mofetil, resulting in symptomatic improvement. Surgical intervention was done for definitive management. This report underscores the significance of early recognition and treatment of PRES in the context of TA to prevent potentially catastrophic consequences and establish causality.

Keywords: Immunosuppressive agent, posterior reversible encephalopathy syndrome, takayasu arteritis, vasculitis

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Introduction

Takayasu arteritis (TA) is a rare chronic idiopathic inflammation of large blood vessels and their branches that is characterized by granulomatous panarteritis consisting of thickening, inflammatory infiltrates, and hyperplasia of the arterial wall.¹ Takayasu arteritis may present as posterior reversible encephalopathy syndrome (PRES) with symptoms of headache, vomiting, seizure, focal neurological deficit, visual disturbance, and altered sensorium. Pregnancy, hypertension, renal disease, rheumatological disorders, and immunosuppression are the risk factors for PRES.² We report an unusual presentation of TA in a child in the form of PRES.

Case Presentation

An 11-year-old girl presented with a complaint of headache and vomiting for 1 month, nasal deviation of both eyeballs for 10 days, and loss of consciousness for 1 day. She had no history of seizures, fever, tubercular contact, gait disturbance, loss of vision, double vision, drooping of eyelids, facial deviation, drooling of saliva, nasal regurgitation, tingling and numbness, bladder or bowel involvement, head trauma, ear discharge, yellowish coloration of eyes, bleeding from any site, rash, joint pain, and passage of high-colored urine. Past, birth, and development history were normal.

On examination, the child was afebrile. Her pulse rate was 102/minute, regular, normovolemic, with no radio-radial and radio-femoral delay, and all peripheral pulses were palpable. Her respiratory rate was 24 breaths/minute, her BP was 160/100 mm of Hg (>95th centile + 12 mm Hg for age, sex, and height), and she had no significant difference in blood pressure of all 4 limbs. Renal bruit was not heard. The fundus showed grade IV papilledema, and other general examinations were normal. On central nervous system examination, the child's Glasgow Coma Scale was E4V4M6. She appeared sick-looking and irritable, with bilateral sixth cranial nerve palsy. In motor function, bulk and tone were normal, power was IV/V in both upper and lower limbs,

and plantar reflexes were bilaterally down-going with deep tendon reflexes +1. Meningeal signs, sensory system, cerebellar system, and autonomic system were unremarkable. Other systemic examinations were normal.

The provisional diagnosis of hypertensive emergency was made and managed as per hospital protocol. The possible etiology of the intracranial space-occupying lesion, tubercular meningitis, and acute demyelinating encephalomyelitis were considered. Laboratory

investigation showed hemoglobin 15.4 g/dL, total leukocyte count $6.9 \times 10^3/L$, platelet count $21 \times 10^9/L$, C-reactive protein 26 mg/L, and erythrocyte sedimentation rate 36 mm/h. Her serum electrolyte, renal function, liver function test, serum calcium, and urine microscopy were within normal limits. Cerebrospinal fluid (CSF) was deferred due to papilledema. The tubercular workup, including tuberculin test and gastric aspirate for cartridge-based nucleic acid amplification test, was negative. Further investigation to rule out autoimmune

conditions and vasculitis (C3, C4, c-ANCA, p-ANCA, ANA, anti-dsDNA, VDRL, ASO titer) were also normal. Echocardiography showed mild concentric left ventricular hypertrophy with trivial aortic incompetence with a left ventricular ejection fraction of 70%.

Magnetic resonance imaging of the brain showed a hyperintense lesion in the bilateral parieto-occipital parenchyma on FLAIR and a T2-weighted image suggestive of PRES (Figure 1). Further, the color Doppler of renal

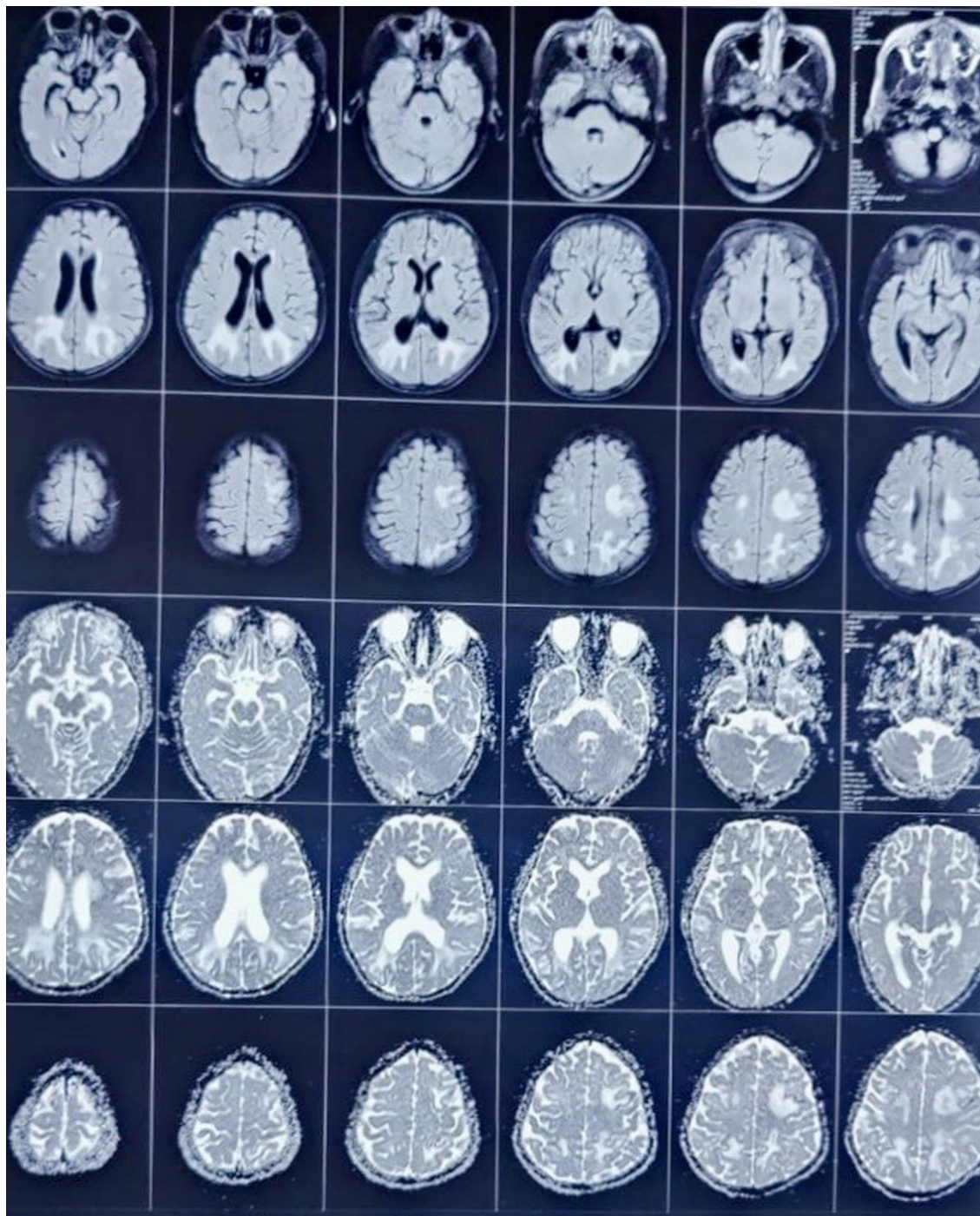


Figure 1. FLAIR and T2-weighted image showing hyperintensities in the occipito-parietal region.



Figure 2. CT angiography showed circumferential thickening of the left main and accessory renal arteries with severe narrowing at the origin and diffuse circumferential mural thickening of the abdominal aorta from the level of origin of the superior mesenteric artery extending up to the origin of the bilateral renal artery, causing luminal narrowing.

vasculature suggested left renal artery stenosis. Later, computed tomography (CT) angiography of the thorax and abdomen showed left renal artery stenosis with renal infarction due to aorto-arteritis and diffuse circumferential thickening of the abdominal aorta causing mild luminal narrowing, which was suggestive of TA type IV (Figure 2).

As per the European League Against Rheumatism/ Pediatric Rheumatology International Trial Organization/ Pediatric Rheumatology European Society consensus criteria (angiographic abnormality with hypertension and raised erythrocyte sedimentation rate [ESR and/or C-reactive protein [CRP]], the diagnosis of TA type IV with hypertensive emergency with PRES with bilateral sixth cranial nerve palsy was made.

Initially, supportive management for a hypertensive emergency involved the use of injectable antihypertensive medication, later shifted to oral antihypertensive medication. For the definitive treatment of TA, the child was induced with corticosteroids, and the disease activity was stable over 2 weeks, maintained with immunosuppressive therapy (corticosteroids and mycophenolate mofetil) and a planned slow de-escalation of immunosuppression over 2 years. At the 6-month follow-up, the child was assessed using the Indian Takayasu Clinical Activity Score 2010. The patient's score at baseline was 8, suggesting substantial disease activity. The score reduced to 2 during the follow-up following the commencement of therapy.

Informed consent was obtained from the father for publication of this case report and

any accompanying images. The father was aware of the purpose of the report, its potential publication in the European Journal of Rheumatology, and the potential readership. All necessary measures were taken to protect the patient's confidentiality and anonymity. The patient had the right to refuse participation or withdraw consent at any time.

Discussion

Takayasu arteritis is a granulomatous large vessel vasculitis that predominantly involves the aorta and its major branches. The annual incidence rate of TA is 0.4-3.4 per million individuals.³ It affects mainly younger females (10-30 years).⁴ Clinical manifestations are variable as per territory of vascular involvement and include hypertension (73%), dyspnea (32%), fever (29%), headache (24%), weight loss (19%), and pain abdomen (14%).⁵ Literature review till now has seen 13 additional instances where PRES was linked to TA. Similarities in all cases involve the left renal artery. Magnetic resonance angiography or CT angiography is used for establishing the diagnosis of TA. EULAR/PRINTO/PRES criteria were used for the diagnosis of TA. It includes 1 mandatory criterion (angiographic abnormalities of the aorta or its main branches) and minor criteria: at least 1 of the following criteria, decreased peripheral artery pulses and/or claudication of extremities, blood pressure difference between arms or legs >10 mm Hg, bruits over the aorta and/or its major branches, hypertension, and elevated acute phase reactants.

Posterior reversible encephalopathy syndrome is a disorder of reversible subcortical vasogenic

brain edema in patients with acute neurological symptoms in the setting of renal failure, blood pressure fluctuations, cytotoxic drugs, autoimmune disorders, and pre-eclampsia or eclampsia. The postulated hypothesis of PRES is a sudden increase in blood pressure or cytokine-induced endothelial injury, which results in breakdown of the blood-brain barrier and cerebral edema. Posterior reversible encephalopathy syndrome presents with clinical features of encephalopathy, seizure, headache, visual abnormalities, nausea, vomiting, focal neurological signs, and acute hypertension. The diagnosis of PRES is crucial, and treatment includes symptomatic therapy (antihypertensive, anticonvulsant) and correction or removal of the underlying cause, as seen in our case, to prevent catastrophic events such as hemorrhages, ischemia, massive infarction, and even death.⁶

The management of TA involves a multidisciplinary approach that includes medication, renovascular procedures, lifestyle changes, and regular monitoring. Corticosteroids are the main drugs used to reduce inflammation, along with immunosuppressive drugs like methotrexate, azathioprine, Mycophenolate mofetil (MMF).⁷ In non-responsive patients, we can use biological agents such as infliximab or tocilizumab and revascularization procedure.⁸ Antiplatelet or anticoagulant medications help to prevent blood clots. To monitor response, CRP and ESR levels should be measured every three months, with an annual MRI angiography.⁹

Takayasu arteritis in children poses diagnostic and therapeutic challenges due to its rarity and nonspecific presentation, like PRES. The outcome of PRES in children with TA is usually favorable; nevertheless, it is necessary to promptly diagnose and quickly treat the underlying condition to prevent further progression of the disease.

Data Availability Statement: XXX

Informed Consent: Informed consent was obtained from the patient's parent who agreed to take part in the study.

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