Unusual presentation of Takayasu's arteritis as posterior reversible encephalopathy syndrome

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Abstract

Takayasu's arteritis is a chronic, idiopathic, inflammatory disease primarily affecting aorta and its branches. It mainly affects young females in the age group of 10-30 years. Various atypical presentations of Takayasu's arteritis have been described in children. These atypical presentations can cause delayed diagnosis resulting in increased morbidity and mortality. Posterior reversible encephalopathy syndrome (PRES) is a neuroradiologic condition associated with headache, seizures, altered sensorium, visual disturbances, and characteristic lesions on neuroimaging. We report a child with Takayasu's arteritis who presented a posterior reversible encephalopathy syndrome. He also had associated abdominal tuberculosis for which anti-tuberculous treatment was started. PRES was diagnosed by magnetic resonance imaging with fluid-attenuated inversion recovery sequences. The child was started on nifedipine and propranolol. The child regained his consciousness within 48 h of admission. Prompt treatment of hypertension led to rapid reversal of neurological symptoms. In view of hypertension a computed tomography aortogram was done, which showed features suggestive of high grade (>75%) focal proximal left renal artery stenosis. EULAR (European League Against Rheumatism)/PReS (Paediatric Rheumatology European Society) consensus criteria was used for the diagnosis of Takayasu's arteritis in our patient. Percutaneous transluminal balloon angioplasty of the stenotic left renal artery was performed. Post-angioplasty, nifedipine was gradually omitted and oral propranolol was continued.

Key Words

Posterior reversible encephalopathy syndrome, takayasu arteritis, tuberculosis

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Introduction

Takayasu's arteritis (TA) is a chronic, autoimmune, and potentially progressive inflammatory disease of large vessels with a preference for the aorta and its major branches.[1] Various atypical presentations of TA as cerebral aneurysms, uveitis, abdominal pain, and cardiac failure have been described in children.[2-5] Due to these atypical presentations, it is not uncommon for TA in children to get diagnosed late or even remain unrecognised resulting in increased morbidity and mortality. We herein describe an unusual presentation of TA in a child as posterior reversible encephalopathy syndrome (PRES).

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Case Report

In July 2008, a 9-year-old Indian boy was admitted with complaints of fever since six days and headache with vomiting since three days. He had three episodes of convulsion on the day of admission. The convulsions were generalized tonic clonic, each lasting for about five minutes. In between the convulsions the child remained unconscious. There was no history of blurring of vision, drug intake, head injury, or Koch's contact. His past, development, and family history was normal. On admission, the child was unconscious with a Pediatric Glasgow coma scale score of 8/15 (E2 V2 M4). He was afebrile with a heart rate of 106/min, respiratory rate of 22/min, and blood pressure of 140/110 mmHg in right upper arm (>95th percentile for age and sex). Both his weight (21 kg) and height (128 cm) were below 5th percentile for his age and sex. All his peripheral pulses were felt normally. There were no neurocutaneous markers or skin rash on examination. His pupils were equal and reactive to light and fundoscopy was normal. There were no meningeal signs, focal neurological deficit, or cerebellar signs. Deep tendon jerks were depressed and plantars were extensors. Other systemic examination was normal. Laboratory studies revealed that hemoglobin was 10.9 gm/dL, white blood count 15,600/cumm (neutrophils

70%, lymphocytes 30%), and platelet count 5.5 lac/cumm. Erythrocyte sedimentation rate was 104 mm at 1 h. His liver and renal function tests, serum electrolytes, and serum calcium were within normal limits. Peripheral blood smear examination for malarial parasites was negative. His urine microscopy, cerebrospinal fluid examination, and chest x-ray were normal. Antinuclear antibody titers and anti-double stranded DNA titers were negative. Mantoux test was strongly positive (24 mm). Chest radiograph was normal. Gastric lavage fluid examination for acid fast bacilli for three consecutive days was negative. Ultrasonographic examination of the abdomen revealed multiple necrotic mesenteric and retro-peritoneal lymph nodes with the largest being 2.7 × 2.1 cm.

The child was started on nifedipine (0.3 mg/kg/dose, 6 hourly) and propranolol (0.5 mg/kg/dose, 6 hourly). Intravenous valproic acid (10 mg/kg/day) and anti-cerebral edema measures (i.e., intravenous mannitol and dexamethasone) were also started. In view of the unexplained multiple seizures, severe hypertension, and persistent altered sensorium a magnetic resonance (MR) imaging with fluid-attenuated inversion recovery (FLAIR) sequence was done. It showed hyperintense lesions in bilateral frontal and parieto-occipital parenchyma on FLAIR and T2 weighted images suggestive of "posterior reversible encephalopathy syndrome" [Figures 1 and 2]. The blood pressure was controlled and he regained full consciousness within 48 h of admission. In view of persistent hypertension a computed tomography aortogram was done, which showed features suggestive of high grade (>75%) focal proximal left renal artery stenosis [Figure 3]. Color Doppler of neck vessels and bilateral subclavian arteries showed normal flow. As per EULAR (European League Against Rheumatism)/ PReS (Paediatric Rheumatology European Society) consensus criteria the diagnosis of Takayasu's arteritis was confirmed as the child had angiographic abnormalities and hypertension.[6] He was started on oral prednisolone (2 mg/kg/day), which was gradually tapered over the next six weeks. In view of the strongly positive Mantoux test, elevated ESR and abdominal lymphadenopathy, a diagnosis of abdominal tuberculosis was made and the child was started on anti-tuberculosis treatment. Percutaneous transluminal balloon angioplasty of the stenotic left renal artery was performed. Post-angioplasty, nifedipine was gradually omitted and oral propranolol was continued. On follow-up after 6 months, the child was asymptomatic with his blood pressure well controlled on propranolol.

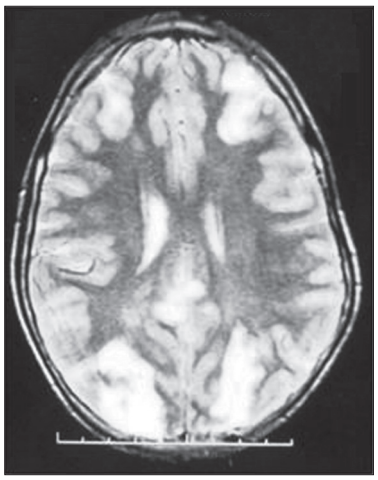

Figure 1: T2 weighted images showing more or less symmetric lesions in bilateral frontal and parietooccipital parenchyma

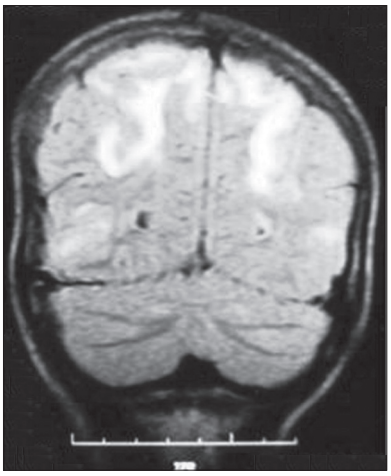

Figure 2: MRI study with FLAIR sequence shows bilateral symmetric hyperintense lesions in parietooccipital parenchyma

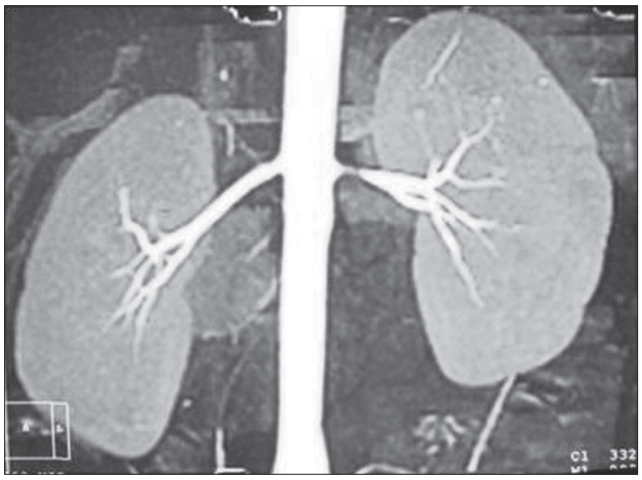

Figure 3: Computed tomography aortogram showing left artery renal stenosis

Discussion

Posterior reversible encephalopathy syndrome (PRES), first reported in 1996 by Hinchey *et al.*, is a rare neuroradiologic condition associated with headache, vomiting, seizures, altered sensorium, visual disturbances, and occasionally focal neurological deficit.[7,8] Most cases of PRES are described in conditions associated with an abrupt increase in blood pressure.[9-11] The seizures are usually of generalized tonic-clonic type and status epilepticus occurs in more than half of the cases.[9,10] They may be preceded by visual auras and visual hallucinations, consistent with occipital lobe seizures.[10] TA is the third most common cause of childhood vasculitis in the world. Though the precise etiology is still unknown, there appears to be some association with an underlying tuberculosis infection. Autoimmune nature of this disease has also been suspected because of its association with various autoimmune disorders.[12] There are two commonly used

criteria for the diagnosis of TA, i.e., American College of Rheumatology criteria (ACR) and European League Against Rheumatism (EULAR)/PRES (Paediatric Rheumatology European Society) consensus criteria. The ACR criteria were primarily designed for adults; however, the more recent EULAR/Pres criteria are specific for the diagnosis of TA in children. According to EULAR/PReS consensus criteria, diagnosis of TA requires angiographic abnormalities (conventional, CT, or MRI/A) of the aorta or one of its major branches plus one or more of the following: (1) claudication or decreased peripheral artery pulses, (2) blood pressure difference >10 mm Hg, (3) bruits of the aorta or its major branches, (4) hypertension.[6] Our patient had angiographic abnormalities and hypertension and was diagnosed as TA as per EULAR/Pres consensus criteria.

Although the most common presenting feature of TA is hypertension, few cases can present acutely as hypertensive encephalopathy and lead to PRES.[10,12,13] An extensive literature search revealed only a single case report of TA presenting as "posterior reversible encephalopathy syndrome".[10] The hypothesis of "hyperperfusion injury" has been put forward to explain the pathophysiology of PRES in hypertensive encephalopathy. A rapid rise of blood pressure overcomes the normal autoregulation of cerebral blood flow and causes dilatation of the cerebral arterioles, resulting in brain hyperperfusion. This increased perfusion pressure can overcome the blood-brain barrier, resulting in opening up of endothelial tight junctions, and leakage of plasma and red cells into the extracellular space, leading to cerebral edema (vasogenic edema).[11,14] The relative paucity of sympathetic innervation in the posterior brain results in increased susceptibility to hyperperfusion and vasogenic edema during acute blood pressure elevations.[9,10] The lesions of PRES are best visualized with MR imaging with FLAIR sequence. T2 weighted MR images, at the height of symptoms, characteristically show diffuse hyperintensity selectively involving the parietooccipital white matter. Occasionally the lesions also involve the grey matter.[9-11]

The management of PRES includes early recognition of the condition, adequate control of hypertension and appropriate anticonvulsant treatment.[9,10,14] As PRES is a reversible condition, long-term antiepileptic therapy is usually not recommended.[10] Irreversible cytotoxic edema, ischemia, and infarction in brain can occur due to delay in the diagnosis and treatment. This can result in long term sequelae including neurodevelopmental delay, persistent vision abnormalities, neurological deficits, chronic epilepsy, or even death.[9,10,14] The treatment of TA begins with control of the acute arteritis with immunosuppressive drugs inorder to induce clinical remission. Corticosteroids (prednisolone, 60-80 mg/day or 0.7-1 mg/kg/day) are the commonest immunosuppressive drugs used in the active phase of the disease.[15,16] The steroids are continued for 1-3 months in patients showing good response to therapy. Once remission is obtained, steroids are tapered to 10-20 mg/day at 3-6 months f/b 5-10 mg/day by 12 months. Response rates of 20-100% have been reported with subsequent resolution of symptoms and stabilization.[15,16] This may relate to the stage of disease at which treatment is introduced in addition to disease extent. Patients who show poor or no response to steroids can be started on either cyclophosphamide or methotrexate or azathioprine.[15,16]

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