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A case of atypical Posterior Reversible Encephalopathy Syndrome (PRES) in a child with snake envenomation

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Abstract

Introduction: Posterior Reversible Encephalopathy Syndrome (PRES) is a clinico-radiological syndrome characterized typically by symmetrical white matter edema affecting predominantly the vascular watershed areas of the posterior occipital and parietal lobe. However, many atypical manifestations are also reported. PRES after snake envenomation are a rare phenomenon and data in this regard is sparse in children.

Case history: An 8-year-old girl admitted with snake envenomation had right hemiparesis and aphasia which improved over a week. Magnetic Resonance Imaging of the brain revealed medium size asymmetrical diffusion restricting lesions in both frontal lobe (Left > Right) and both parieto-occipital lobe suggestive of PRES with normal MR Angiogram and MR Venogram. After a month, the neurological deficits in the child had improved but there was minimal speech articulation defect.

Discussion: Neurological symptoms and signs after snake envenomation include drowsiness, confusion, dizziness, blurred vision, loss of musclular co-ordination and convulsions, intracranial haemorrhage, ischemic strokes of different arterial territories and Acute Disseminated Encephalo Myelopathy (ADEM). A few cases of PRES have been reported in association with snake envenomation in adults. The characteristic images in PRES syndrome reveal symmetrical cortical and sub-cortical hyper intense signals on T2 weighted and Fluid-Attenuated Inversion Recovery (FLAIR) images in the parietal-occipital regions. Atypical features like asymmetry have been reported in some studies.

Conclusion: In children with snake envenomation, the development of PRES should be recognized promptly and has to be differentiated from venom induced neurological manifestations.

Keywords

Children; viper; neurotoxicity; encephalopathy; atypical; residual deficit.

Introduction

In the world annually around 500,000 cases of snake envenomation and approximately 40,000 to 100,000 deaths are reported by various authors [1]. Snake envenomation is an important cause of mortality and morbidity in rural India with about 35,000 to 50,000 deaths occurring every year and is usually common in the rainy season [2]. Three families of poisonous snakes commonly found in India are Viperidae (Vipers), Elapidae (Cobra and Krait) and Hydrophidae (Sea snakes). Viperine snake bites including Russell's viper (*Daboia russelli*) and saw scaled viper (*Echiscarinatus*) are more fatal. The local and systemic clinical manifestations of snake bites are due to the venom components that may lead to cytotoxic, hypotensive, neurotoxic, and anticoagulant or procoagulant effects [3]. Neurological manifestations are most often related to blockage of the neuromuscular transmission, causing paralysis, as venom proteins do not cross the blood–brain barrier and abnormalities in the coagulation cascade leading to cerebrovascular events [4,5]. Toxin-induced hypercoagulability, systemic hypotension, thrombotic microangiopathy, and immune-mediated vasculitis, have been proposed to explain the occurrence of cerebral infarctions in snake bite victims [5-7]. Venom-induced endothelial damage may also be the cause of ischemic strokes in these patients [8]. PRES after snake envenomation is a rare phenomenon and data in this regard is sparse in children [2].

Case Report

An 8 years old girl had a venomous snake bite in the evening hours while she was playing outside her residence after which she became unconscious within fifteen minutes. The child was taken to a local facility immediately where the fang marks were noticed over the lower third of the left leg. As the child was unconscious and had poor respiratory effort, she was intubated, ventilated, administered 20 vials of Anti Snake Venom (ASV) and shifted to our hospital. On admission the child had a Glasgow Coma Scale of 7/15 (E2 V0 M5). She had cellulitis that was progressive and had reached up to the mid-thigh of her left lower limb. There was bleeding from the site of bite and multiple petechiae over the left lower limb was also noticed. Her heart rate was 160/min, BP was 90/60 mmHg as measured in the right upper limb in supine position. There was deviation of angle of mouth to left side with right hemiparesis (Figure 1). Pupils were bilaterally equal and reacting to light. There was no ptosis and no external opthalmoplegia was noted. Cardiovascular and respiratory system examinations were normal and her urine output was adequate with no hematuria. Hence a provisional diagnosis of hematotoxic snake envenomation was made.

Blood investigations revealed Hemoglobin of 12.4 gms%, Hematocrit of 34% and a Platelet count of 28,000/cu.mm which gradually improved to 5,02,000/cu.mm over one week. Her total leucocyte count was 20,000cells/cu.mm with Polymorphs - 76%, Lymphocytes - 20% and Eosinophils - 4%. Her peripheral blood smear report showed normocytic normochromic blood picture with neutrophilic leukocytosis with severe thrombocytopenia on the day of admission. Whole blood clotting time was more than 20 min. Her renal parameters remained normal for the first two days with adequate urine output after which blood urea was 179 mg/dl, serum creatinine was 1.7 mg/dl, sodium- 144 mEq/L and potassium- 3.3 mEq/L. Liver Function Test showed total bilirubin-0.5 mg/dl, direct bilirubin- 0.2 mg/dl, AST – 66 IU/L, ALT- 14 IU/L, serum Alkaline Phosphatase - 60 IU/L, total protein- 5.1 gms%, albumin- 2.8 gms%. In the pre dialysis screen,

child tested negative for Hepatitis B Virus surface antigen, anti-Hepatitis C Virus antibody and non-reactive for Human Immunodeficiency Virus.

Magnetic Resonance Imaging of the brain on the fourth day of admission revealed medium size diffusion restricting lesion in both frontal lobe and both parieto-occipital lobe suggestive of Posterior Reversible Encephalopathy Syndrome (PRES). There was an asymmetry in involvement of cerebral hemispheres with left hemisphere having wide diffusion restriction and right hemisphere having sparse diffusion restrictions. The cortical areas of parieto-occipital region showed hyper intense signals in T2 weighted and Fluid-Attenuated Inversion Recovery (FLAIR) images. The MR Angiogram and MR Venogram were normal (Figure 2,3,4).

The child was given ventilator support and 10 vials of ASV were administered on second day of admission. Intravenous antibiotics like Ceftriaxone, Cloxacillin, Metronidazole were given for 7 days for management of cellulitis. In view of progressive cellulitis an emergency fasciotomy was done at 36 hours following snake bite. Peritoneal dialysis was done in view of Acute Kidney Injury.

As the whole blood clotting time was prolonged, 10 more vials of ASV were administered again on second day of admission. Totally 40 vials of ASV were required for the whole blood clotting time to become normal. As the child regained consciousness and started to obey commands on day 3 of admission, the child was extubated and after which the child was found to have mutism. With peritoneal dialysis for 24 hours, her renal parameters started reducing to normal values and urine output improved. Weakness of the right side and right sided Upper Motor Neuron type of facial nerve palsy improved than the previous state over a week. Her verbal output also improved with mild articulation deficit persisting. A repeat MRI brain taken after one week also showed the similar findings. The cellulitis and petechiae over the left lower limb decreased with healthy fasciotomy wound. On follow up after a month of admission, the girl had mild speech articulation defects for which she was started on speech therapy.

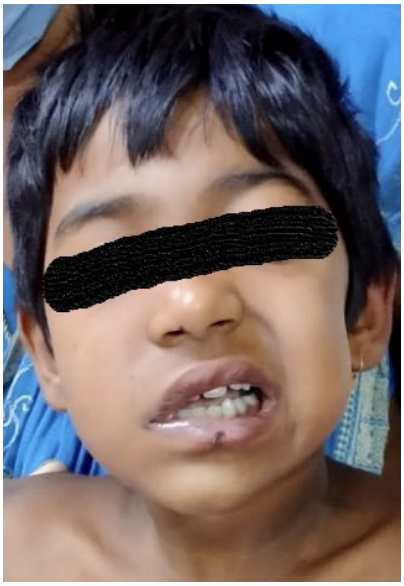

Figure 1: The girl having right UMN facial palsy

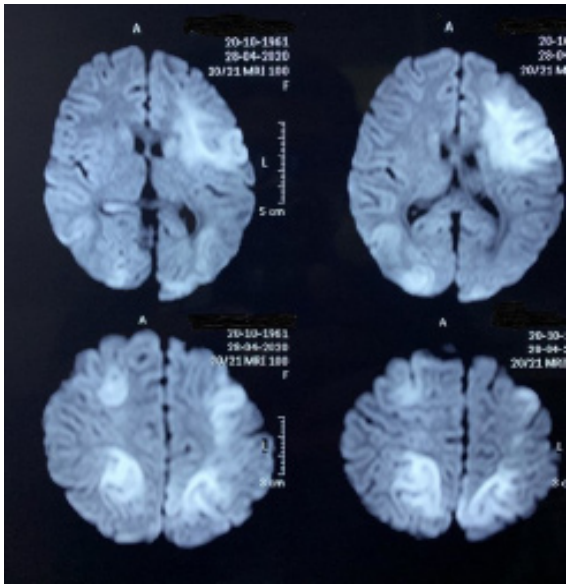

Figure 2: MRI of the brain revealed medium size diffusion restricting lesion in both frontal lobe (Left > Right) and both parieto-occipital lobe.

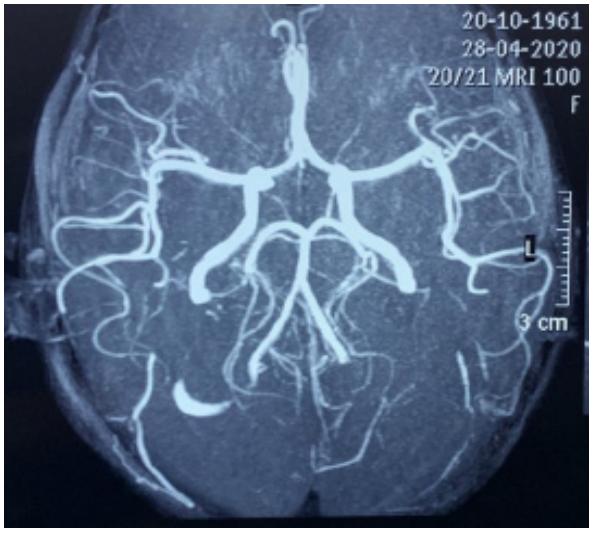

Figure 3: MR Angiogram showing normal arterial architecture.

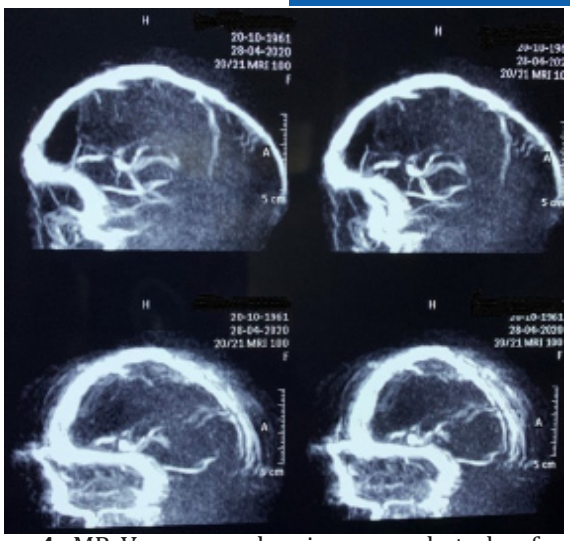

Figure 4: MR Venogram showing normal study of cerebral venous system.

Discussion

Leucoencephalopathies are a heterogeneous group of conditions characterized by developmental abnormalities or degeneration of white matter. It may be an inherited condition or acquired abnormalities of vascular, inflammatory, infection, traumatic, toxic, nutritional, neoplastic etc [9]. PRES is a reversible leucoencephalopathy and the other names of this condition are Reversible Posterior Leucoencephalopathy Syndrome, hyper-perfusion encephalopathy and brain capillary leak syndrome. The PRES is a clinico-radiological syndrome characterized by the white matter edema affecting predominantly the vascular watershed areas of the posterior occipital and parietal lobe [10,11]. But asymmetrical involvement of thalamus, basal ganglia, cerebellum, brain stem and frontal lobe has also been reported in literature. If the insult is recognized promptly and the triggering factor is withdrawn, clinical syndrome resolves within a week [11,12]. PRES is commonly reported as a result of hypertension but around 30% of patients have normal blood pressure [11-15]. Computerized Tomography images may be normal and MRI images are diagnostic.

Neurological symptoms and signs after a viperine bite include drowsiness, confusion, dizziness, blurred vision, loss of musclular co-ordination and convulsions [16]. Most common neurological manifestation in vasculo toxic snakes is intracranial haemorrhage. Ischemic strokes of different arterial territories and Acute Disseminated Encephalo Myelopathy (ADEM) have also been reported. The altered sensorium in this child immediately following snake bite can be related to direct arterial endothelial injuries by the venom itself. The venom has complex substances which are proteins, peptides, biogenic amines, lipids and polysaccharides which has cytotoxic, neurotoxic, anticoagulant and pro-coagulant activities. The highly selective venom components metalloproteinases and C- type lectin activate platelets exhibiting pro inflammatory activity known as Thrombo-inflammation. They also stimulate natural ligands of platelet adhesion receptors. The persistent thrombocytopenia in this child is due to venom induced thrombocytopenia [17]. The venom causes increased vascular permeability which in turn induces abnormal release of tumour necrosis factors and cytokines and cause systemic endothelial dysfunction. This endothelial dysfunction explains the PRES in these children [18].

The characteristic images in PRES syndrome reveal symmetrical cortical and sub-cortical hyper intense signals on T2 weighted and FLAIR images in the parietal-occipital regions. However, a large case series of PRES with 111 cases demonstrated a higher incidence of atypical features like asymmetry [19]. The MRI findings in this child are diagnostic. Repeat MRI taken a week later showed similar finding. But clinically the child had improved. Radiologic findings may take few more weeks for complete resolution [11-13]. PRES may also be a complication of anti-venom therapy [20]. But in this child this is less likely, because the clinical manifestations have started even before starting Anti snake venom. This child had mild speech articulation defects at the end of the month following discharge from the hospital. The earlier report that the PRES is associated with complete recovery was challenged by many. There are reports that the PRES is associated with some residual neurological sequelae [21].

Conclusion

In children with snake envenomation, the development of PRES should be recognized promptly and has to be differentiated from venom induced neurological manifestations. The characteristic MRI findings and a spontaneous resolution favors a diagnosis of PRES. Whereas a presentation with ptosis or external opthalmoplegia should favor the diagnosis of neurotoxic envenomation. The clinical syndrome of PRES resolves within a week in most cases. Atypical features of PRES and some residual neurological deficit have to be kept in mind.

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