Case Report

A case of post-operative posterior reversible encephalopathy syndrome in children: A preventable neurological catastrophe

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinic-radiological syndrome that is generally reversible and may lead to permanent neurological damage if left untreated. PRES has been commonly linked with hypertension along with associated vasogenic oedema. Children are more susceptible to these perturbations due to the narrow range of cerebral autoregulation. PRES must be considered in differentials of any neurological dysfunction which is associated with hypertension in the immediate post-operative period. Inadequate pain control in the post-operative period may cause hypertension that may lead to subsequent PRES. We report a case of postoperative PRES in a 12-year-old previously normotensive child posted for splenectomy with an acute rise in blood pressure in the post-operative period.

Key words: Posterior, post-operative, reversible encephalopathy, syndrome

INTRODUCTION

A reversible, predominantly posterior reversible encephalopathy syndrome (PRES) was first reported in 1996 by Hinchey *et al*.[1] PRES often manifests as headache, blurring of vision, seizures, altered consciousness, transient motor deficits or cortical blindness.[2] Permanent neurological sequelae may follow if left untreated on time. PRES can be easily diagnosed on computed tomography scan (CT), magnetic resonance imaging (MRI) or diffusion-weighted images. Although PRES is a well-known entity in adults, very few cases are reported in the perioperative period especially in the paediatric population. We wish to report a child suffering from thalassemia major with immune thrombocytopenia who developed PRES after splenectomy.

CASE REPORT

A 12-year-old male child weighing 20 kg suffering from thalassemia major and immune thrombocytopenia presented with features of pancytopenia. His bone marrow examination showed red cell aplasia. The child was given steroid therapy in the form of oral prednisolone 1 mg/kg/day for 4 weeks.

Child's haemoglobin was 9 g/dl and his platelet count was 30,000/mm^3 despite steroid therapy. Ultrasound of the abdomen showed hepatosplenomegaly. The child had pancytopenia, hypersplenism and was posted for splenectomy. Splenectomy was done under general anaesthesia with endotracheal intubation (ETT). Stress dose of steroid hydrocortisone 25 mg was given

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perioperatively. Before extubation, ultrasound-guided bilateral single shot erector spinae block with 15 ml of 0.2% ropivacaine was given at T-8 level. The trachea was successfully extubated and the child was shifted to the post anaesthesia care unit (PACU).

The child was pain-free in the immediate postoperative period with a visual analog score (VAS) score of 1/10. The patient was discharged from PACU. On the second post-operative night, the child complained of severe pain at the surgical site with a VAS score of 8/10. Intravenous paracetamol 15 mg/kg was given for pain control but the pain was not relieved. The nonsteroidal anti-inflammatory drug was not given due to the risk of platelet dysfunction. The child had a heart rate of 120 beats/min and non-invasive blood pressure of 170/94 mmHg. The patient had continued pain and at midnight and he complained of a bilateral diffuse occipital headache followed by the blurring of vision and visual hallucinations. The symptoms worsened rapidly. Two hours later, the child had a generalised tonic-clonic seizure lasting for 2 min. Seizure episode was managed by intravenous (IV) midazolam 4 mg. A loading dose of IV phenytoin 20 mg/kg was given by slow infusion followed by 100 mg intravenous per day. The seizure was followed by postictal confusion. During this episode, blood pressure increased up to 180/96 mmHg, and the child was shifted to the intensive care unit (ICU).

On admission to the ICU, his Glasgow Coma Scale was 9/15 (motor 4, eyes 2, verbal 3). His blood pressure (bp) was 180/100 mmHg, heart rate 106 beats/min, body temperature 36.7°C and pupils were 2.5 mm in diameter, equal and reactive. The fundus examination was unremarkable. Blood glucose, arterial blood gases, serum osmolality and serum electrolytes were normal. The child maintained oxygen saturation throughout the whole event.

The patient had a recurrence of seizure after ICU admission, following which levetiracetam 20 mg/kg intravenous infusion was given. MRI brain showed T2-weighted-fluid-attenuated inversion recovery (T2 FLAIR) hyperintensities in the bilateral parieto-occipital region and bilateral cerebellar hemispheres with underlying cortical swelling [Figures 1a and b]. Magnetic resonance venography (MRV) was also done to rule out venous sinus thrombosis. All clinical and radiological features were consistent with PRES.

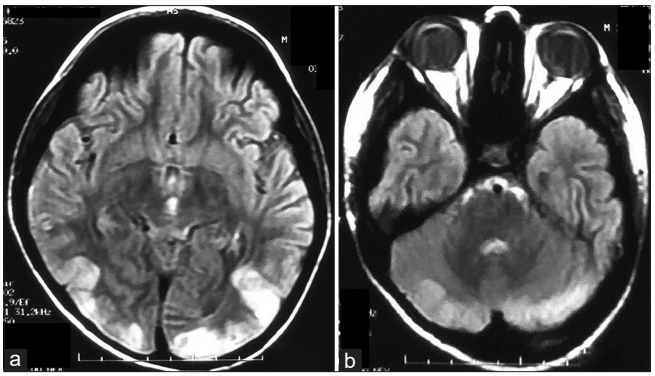

Figure 1: Magnetic resonance imaging (MRI) of the patient showing FLAIR hyperintensities in the bilateral parieto-occipital region (a) and bilateral cerebellar hemispheres (b)

Blood-pressure monitoring showed wide fluctuations in reading hence left radial intra-arterial blood pressure monitoring was started in ICU. The systolic blood pressure of child ranged between 120–190 mm of Hg, whereas diastolic blood pressure ranged between 70–100 mm of Hg. The hypertension was managed with intravenous labetalol 0.4–1 mg/kg/hr intravenous infusion. Within 24 h, the blood pressure fluctuations resolved and oral prazosin tablet 0.5 mg once daily was started for maintenance. Oral phenytoin 150 mg and levetiracetam 400 mg were continued once daily. For analgesia paracetamol, 15 mg/kg was given intravenously every 6 h and intravenous fentanyl 1 μ/kg bolus as per requirement. The dose of steroid was tapered to prednisolone 6 mg once daily. Headache and visual symptoms resolved gradually over 2 days. He did not have any recurrence of seizure and phenytoin was stopped while oral levetiracetam was continued. The patient was discharged on day 15.

DISCUSSION

PRES is generally a benign and self-limiting condition but permanent neurological damage can occur if left untreated. Pathophysiology of PRES is explained by two theories, namely, cytotoxic and vasogenic. The cytotoxic theory implies vasoconstriction and hypoperfusion as causative factors of brain ischaemia and subsequent vasogenic oedema. The other theory states that weak cerebral autoregulation and endothelial dysfunction causes the breakdown of the blood-brain barrier and vasogenic oedema.[3] Most commonly, there is vasogenic oedema within the occipital and parietal regions (~95% of cases), relating to the posterior cerebral artery supply. The oedema is usually symmetrical. Despite being termed posterior, PRES can be found in the non-posterior

distribution, mainly in watershed areas, frontal, inferior temporal, cerebellar and brainstem regions. Both cortical and subcortical locations are affected. Preferential involvement of posterior circulation in PRES is due to sparse sympathetic innervation of vertibro-basilar circulation, predisposing them to impaired autoregulation during hypertensive episodes.[4] Since children have a narrower range of cerebral autoregulation, they are more prone to PRES than adults under conditions of high systemic blood pressures.[3] PRES is manifested as seizures associated with headache, visual hallucinations, blurring of vision, gaze deviation, cortical blindness, impaired consciousness and sometimes paresis or focal neurological deficits.

In 70–80% of patients moderate-to-severe hypertension is found to be the precipitating factor of PRES. However, in 20-30% of patients, blood pressure is within the normal range.[5] Cerebral venous sinus thrombosis, encephalitis, cerebrovascular accident, hypoxic encephalopathy and dyselectrolytaemia usually have the same presentation as PRES; hence neuroimaging is imperative to differentiate PRES from the former. An early neuroimaging is also vital since the line of management of all these conditions are entirely different.

Our patient had an acute rise in blood pressure at the time of the start of symptoms. Hypertension was never elicited during pre-operative evaluation and blood pressure returned to normal within a few days following adequate medical treatment and pain control, suggesting the transient nature of hypertension. Pain is an important cause of transient hypertension.[6] Severe pain is known to cause an acute rise in blood pressure in the post-operative period, especially in children.

Hypertension is known to occur with corticosteroid therapy. This possibly due to the effect of corticosteroids on mineralocorticoid receptors. This steroid-induced hypertension is responsible for the development of PRES.

An even brief duration of steroid therapy can frequently precede PRES; this was evaluated by Parikh *et al*. in their retrospective review of radiology reports between 2008–2014 in 99 cases.[7] The patient was on steroid therapy and steroids have been well-reported to precipitate PRES. However, in our case, hypertension was sudden and time was correlated with the effect of erector spinae block wearing off time. Hence it might be possible that steroid therapy might have added to the development of PRES in this case The peri-operative pain was well-managed with ultrasound-guided single-shot erector spinae block. However, as the effect of the block weaned off, the patient suffered severe pain. Although the pain was partially managed with paracetamol, it did not provide complete analgesia. Severe pain could have precipitated an acute rise in blood pressure, which lead to symptoms of PRES a few hours later. The temporal association of pain with hypertension and neurological symptoms supports our hypothesis that pain may lead to a rise in blood pressure causing neurological damage.

This case underlines the importance of pain management and blood pressure in a post-operative paediatric patient, especially who is on pre-operative steroid therapy. While immediate post-operative pain analgesia is addressed by the anaesthesiologists and PACU team, it is often neglected once the patient is discharged from the PACU. Common causes of improper pain management during this period include failed anticipation, delayed recognition and inadequate treatment.

It has been shown that a single-shot erector spinae block provides effective pain relief for approximately 16 h postoperatively.[8] Therefore, it is imperative for physicians to anticipate escalation in pain score and empirically step-up the appropriate analgesic supplementation.

In present time, peripheral regional nerve blocks and fascial plane blocks are changing the way perioperative pain management was looked upon. One must also take care of the wearing off time the effect of these blocks and intervene with appropriate analgesics by communication between ward staff and acute pain service personnel. This is particularly important in patient groups who could have difficulty conveying pain, as in children.

Multidisciplinary care involving a team of the intensivist, anaesthesiologist, pain physicians, and neurologist is crucial for optimum management. Delay in management could lead to permanent neurological sequelae.[9] Early diagnoses with MRI and immediate blood pressure control form the cornerstone of management of PRES.[10]

CONCLUSION

Inadequate pain management in the post-operative period can precipitate PRES in children.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms. In the form the Parent/s has/have given his/her/their consent for his/her/their child's images and other clinical information to be reported in the journal. The parents understand that their child's names and initials will not be published and due efforts will be made to conceal their child's identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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