Neurogenic Pulmonary Edema Following Status Epilepticus: An Unusual Case

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Abstract

Neurogenic pulmonary edema (NPE) is rare in pediatric patients. It occurs most commonly following brain malignancies, traumatic brain injuries, infections and seizures. It has rarely been reported in pediatric patients following status epilepticus. The pathophysiology is not completely understood and is proposed to be due to overactivity of sympathetic nervous system leading to pulmonary interstitial and alveolar edema. Here we present a case of a 4-year-old healthy boy with status epilepticus secondary to febrile seizures who developed NPE. Treatment is supportive ventilator management with low tidal volume and high positive end-expiratory pressure. Timely diagnosis and management is the key for good recovery. It should be considered in the differential diagnosis of patients with rapidly developing pulmonary edema in the setting of normal cardiac function.

Keywords: Neurogenic pulmonary edema; Status epilepticus; Respiratory distress

Introduction

Neurogenic pulmonary edema (NPE) could be a contributing factor to morbidity after status epilepticus. It was first described by Shanahan et al in 1908 following epileptic seizures but still after hundred years, it remains an under-appreciated entity in the clinical world [1]. It manifests as an acute onset of pulmonary edema in the setting of central nervous system injury without previous cardiac pathology [2]. In adults, NPE could result from various central nervous system disorders such as subarachnoid hemorrhage following aneurysm rupture, brain malignancies, traumatic brain injuries, infections and prolonged seizure activity [3]. It develops within minutes, hours or days following injury and significantly complicates the overall clinical status of the patient [4]. In pediatric patients, respiratory compromise has been attributed to anticonvulsant drug use and only isolated case reports exist for its association with recurrent seizures [3, 5, 6]. This is likely due to lack of awareness about the diagnosis and unpredictable nature of its etiology [2, 3]. The diagnosis of NPE requires high index of clinical suspicion. We report a case of a 4-year-old healthy boy, known case of simple febrile seizures, who presented with febrile status epilepticus and developed NPE. Strong clinical suspicion and prompt management resulted in complete recovery with no morbidity. The case highlights the importance of considering NPE in the differential diagnosis of respiratory compromise after pediatric status epilepticus.

Case Report

A 4-year-old developmentally normal boy was a known case of simple febrile seizure, with onset at age of 1.5 years. He presented with history of fever of 1 day duration followed by history of generalized tonic-clonic seizures for 2 h. Detailed clinical examination revealed temperature of 101 °F, tachycardia with heart rates in range of 140 - 160/min, and tachypnea with subcostal retractions. However, rest of the systemic examination was within normal limits. Initially, child was managed as per protocol for status epilepticus, i.e. supportive management for airway, breathing and circulation and given intravenous lorazepam as antiepileptic agent. Seizure was not controlled with two doses of lorazepam, so child was loaded with intravenous valproic acid was given and the child was intubated for airway protection. Finally seizure responded to IV midazolam infusion. After seizure control, child developed features of respiratory distress in form of tachypnea, tachycardia and low SpO2 (86-88%). Child was immediately put on mechanical ventilation. The lung compliance was poor and child required high pressures and high positive end-expiratory pressure to expand the lungs. There was clinical suspicion of acute respiratory distress syndrome. Routine chest X-ray revealed diffuse bilateral...
that is due to an acute central nervous system injury and usually develops rapidly after the injury [2]. Sympathetic overactivity in hypothalamus, medulla, nuclei of solitary tract and area postrema after central nervous system injury is a proposed mechanism [7]. The downstream effects of sympathetic overactivity vary depending on the cardiopulmonary effects [2]. Pathologically, there is marked pulmonary vascular congestion with perivascular edema, extravasation and intra-alveolar accumulation of protein rich edema fluid and intra-alveolar hemorrhage [8]. The clinical pathology overlaps with acute lung injury and acute respiratory distress syndrome [6]. It is largely underdiagnosed and poorly reported because of the poor clinical status of the patient caused by the primary central nervous system injury and it necessitates rapid and often complex treatment [9].

Clinically, NPE presents in minutes to hours (immediate onset) of a severe central nervous system insult such as subarachnoid hemorrhage, traumatic brain injury or status epilepticus. However, presentation in hours to days (delayed onset) has been described [2]. The signs of NPE are quite non-specific [3,9]. It presents subjectively with a sudden onset of dyspnea, chest pain, worsening of expectoration, vomiting and weakness. Clinical examination reveals tachyypnea, tachycardia, basal bilateral pulmonary crackles, respiratory distress or failure, expectoration of sanguinolent sputum or even hemoptysis, hypoxemia and increased systemic blood pressure [10]. The most relevant imaging method is the chest X-ray examination that reveals a normal size heart with diffuse bilateral alveolar opacities [9]. Although the levels of some substances such as brain natriuretic peptide, blood C-reactive protein and IL-6 are increased, unfortunately none of these can be used as a marker specific for NPE [10]. It is proposed that measurement of serum catecholamines might be helpful and treatment with α-adrenergic blocking agent, such as phentolamine, may be considered [2].

In this case, a 4-year-old child developed respiratory distress following status epilepticus and clinical evaluation revealed features of pulmonary edema. The differential diagnosis included acute respiratory distress syndrome due to aspiration and cardiogenic pulmonary edema. The cardiac examination and workup was within normal limits. There was no evidence of acute infection and the blood cultures for bacteria, fungus and viruses were also negative. Cardiac functions were also within normal range. In pulmonary edema of unknown origin, associated with neurological condition, the possibility of NPE should be considered.

The case was reported to highlight the importance of strong clinical suspicion and prompt management of the condition. So far, there is only one reported case of NPE following febrile status epilepticus in this age group [3]. Such cases are rare and need constant vigilant management for speedy recovery. Our patient received timely supportive treatment and recovered completely from the event. Early recognition and appropriate use of positive end-expiratory pressure and judicious fluid management are important in the management of such patients.

Discussion

NPE is an increase in pulmonary interstitial and alveolar fluid that is due to an acute central nervous system injury and usually occurs within 72 h. The child is on follow-up and with no cardio-vascular/neurogenic sequelae.

Conclusion

NPE should be considered in the differential diagnosis for the rapid progression of respiratory failure following an acute neu-
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Conflict of Interest

None.

References