Case Report

Neurogenic pulmonary edema in an infant with ventriculoperitoneal shunt malfunction

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Received 23 June 2012

Revised 6 December 2012

Accepted 15 December 2012

Abstract. Neurogenic pulmonary edema (NPE) and neurogenic stunned myocardium are known complications of acute neuro-pathology, but these conditions have only rarely been reported with ventriculoperitoneal shunt malfunction. Furthermore, shunt malfunction presenting with NPE has not been described in a pediatric patient. A case of acute shunt malfunction and NPE followed by a complicated cardiopulmonary clinical course is reviewed.

Keywords: Cerebrospinal fluid shunting, hydrocephalus, neurogenic pulmonary edema, neurogenic stunned myocardium, pediatric neurosurgery

1. Introduction

Neurogenic pulmonary edema (NPE) and neurogenic stunned myocardium (NSM) are well-recognized complications of subarachnoid hemorrhage and other acute neurological states. Myocardial stunning and pulmonary edema frequently occur simultaneously. In these cases, pulmonary edema may stem from a primary neurological or secondary cardiac event. Massive sympathetic over-activity during the acute neurological episode likely contributes to both pathophysiological mechanisms [1–4]. Patients who survive the initial insult usually have a rapid cardiopulmonary recovery without late onset morbidity [5,6]. NPE and NSM are rare in children but have been reported with trauma [7–11], hemorrhage [12–16], seizure [17], tumor-related obstructive hydrocephalus

[4,18], viral encephalitis [19–22], ketamine toxicity [23], and following decompression of Chiari malformations [24]. Johnson et al. [4] reported two cases of NSM and pulmonary edema secondary to acute hydrocephalus. A 3-year-old girl with a posterior fossa medulloblastoma and a 13-year-old girl with a tectal glioma who failed endoscopic third ventriculostomy developed NSM leading to pulmonary edema at presentation.

One published case of NPE has been attributed to acute ventriculoperitoneal shunt (VPS) malfunction in a 29-year-old myelomeningocele patient [25]. Another case was described in a 21-year-old patient during an endoscopic shunt revision that required extensive irrigation for intraprocedural hemorrhage [26]. Shunt malfunction-related NPE has not been previously described in the pediatric population, but the pathophysiological process of NPE secondary to acute hydrocephalus predicts this possibility. We describe our experience treating a 1-year-old male who developed NPE and possibly NSM related to VPS malfunction.

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2. Case report

The patient presented on his first birthday to the emergency department with a 2 wk history of nausea and vomiting that worsened 3 d prior to admission. The patient was born at 32 wk gestation. Pregnancy was complicated by pregnancy-induced hypertension and disseminated intravascular coagulation. He was diagnosed with grade 3 intraventricular hemorrhage and had a permanent shunt placed at 1 month of age. The patient did well following surgery and was discharged home. His developmental progress was reassuring at subsequent clinic visits [27].

On admission, computed tomography (CT) of the head revealed enlarged ventricles (Fig. 1A). Chest radiograph demonstrated a normal cardiac silhouette and was unremarkable for pulmonary pathology (Fig. 2A). The patient was taken to the operating room the following morning for shunt revision. During the procedure, the proximal catheter had sluggish cerebrospinal fluid (CSF) flow, which significantly improved after replacement. There were no complications with the procedure, and postoperative CT demonstrated acceptable position of the ventricular catheter. The patient improved quickly, becoming more active and tolerating a diet, and he was discharged from the hospital on postoperative day one.

Several hours after discharge, the patient returned to the emergency room with episodes of vomiting. CT of the head revealed an increase in ventricle size compared to postoperative imaging (Figs. 1B and 1C). Early shunt failures are frequently caused by occlusion

with postoperative intraventricular debris, but the exact cause of failure in this case could not be determined. While preparing to take him to the operating room for shunt revision, the patient had periods of apnea and altered mental status. During uncomplicated intubation, copious frothy secretions were noted at the level of the vocal cords and filling the endotracheal tube (ETT). After the airway was secured, an external ventricular drain was emergently placed. Postintubation chest radiograph revealed diffuse airspace opacities without evidence of an enlarged cardiac silhouette (Fig. 2B).

The patient continued to have blood-tinged frothy secretions, which required rapid escalation of ventilatory support. Several hours after initial intubation, the patient had acute desaturation with concerns of ETT obstruction. ETT replacement was complicated by hemodynamic instability and cardiac arrest. With cardiopulmonary resuscitation and relief of bilateral pneumothoraces, spontaneous circulation returned. Despite high inotropic and ventilatory support, the patient continued to be hemodynamically unstable and difficult to ventilate and oxygenate. The patient was cannulated to veno-arterial extracorporeal membrane oxygenation (ECMO). Initial post-cannulation echocardiogram was significant for left ventricular enlargement and decreased ventricular wall motion.

On postoperative day two of VPS revision and day one of ECMO, the patient had an episode of sustained wide-complex ventricular tachycardia and asystole. After defibrillation, chest compressions, and increased ECMO flow, sinus rhythm returned. ECHO following

Fig. 1. Chronological CT scans at the level of maximum lateral diameter of the third ventricle. (A) Preoperatively. (B) Postoperatively. (C) At the time of second presentation prior to treatment. The two images during shunt failure are very similar, but the progression of failure at the second presentation was more rapid.

Fig. 2. Chest radiographs. (A) Before first shunt revision. (B) After intubation during second shunt malfunction.

this dysrhythmia while on ECMO demonstrated a large left ventricular mass consistent with a thrombus. Repeat ECHO performed several hours later revealed resolution of clot, which was concerning for dislodgement into the coronaries and/or cerebral vasculature. CT scan of the head after repeat ECHO was unremarkable for signs of acute ischemia. Serial troponins were followed, with peak of 28 ng/mL following the event. During the ECMO course, the ventricular drain was noted to be intermittently functioning. The EVD was not replaced due to concerns of life-threatening hemorrhage, as the patient was fully anticoagulated on ECMO. On ECMO day 13, repeat CT scan of the head was obtained due to the patient's decreased movement and level of responsiveness, revealing diffuse cerebral infarction with extensive bilateral subdural hematomas. A repeat ECHO on the same day showed normal ventricular sizes and wall motion. The patient was successfully decannulated and continued to have clinical signs consistent with diffuse neurological injury. Due to the patient's grim neurological prognosis, mechanical support was withdrawn and comfort care was provided. The patient died within 48 h of withdrawal of support. The family declined the request for autopsy.

3. Discussion

Morbidity and mortality of CSF shunting in pediatric patients remains high [28], and cardiopulmonary failure is a frequent terminal event [29,30]. The true incidence of NPE and NSM is unknown, but these diagnoses should be considered in cases of acute cardiopulmonary clinical decline from shunt failure. CSF obstruction must

be relieved rapidly and followed by aggressive cardiopulmonary support. Our patient likely suffered a cerebrovascular event secondary to inadequate cerebral perfusion from a low cardiac output state and/or dislodgement of an intracardiac thrombus. The patient's subdural hematomas can be explained as a known complication of anticoagulation related to ECMO [31]. Unfortunately, full anticoagulation precluded adequate ventricular drain revision. Though his cardiopulmonary function was eventually restored, his neurological status never recovered.

The patient's clinical course was characteristic for NPE at presentation with later findings of cardiac dysfunction. All chest radiographs prior to and during the patient's clinical decline demonstrated a normal or narrowed cardiac silhouette, which would be inconsistent with cardiac dysfunction as a primary cause of pulmonary edema. On presentation, the patient rapidly went into florid respiratory failure with profuse blood-tinged frothy secretions consistent with pulmonary edema. The first chest radiograph after his second shunt failure showed diffuse opacities. In addition to the sympathetic surge related to increased intracranial pressure, general anesthesia and mechanical ventilation used during the patient's initial revision may have resulted in vulnerability of the cardiopulmonary system to neurogenic insult.

Cardiac dysfunction may have been neurogenic, but the first echocardiograms were performed under the strain of bilateral pneumothoraces and immediately after high-frequency oscillatory ventilation on ECMO. Echocardiographic abnormalities preceded the separate cardiac event, which occurred 2 d after the initial clinical decline. Like other reports of NSM, the echocardiographic abnormalities present in our patient after shunt failure resolved completely. We remain uncertain

whether the patient's cardiac dysfunction was secondary to NSM, but based on the temporal relationship of the pulmonary and cardiac events, the argument for NPE is convincing.

Neurogenic etiologies of myocardial stunning and pulmonary edema involve acute increase in intracranial pressure followed by massive sympathetic discharge. Shunted patients with low cerebrospinal fluid reabsorptive reserve can be at risk for rapid intracranial pressure elevation. Generally, shunt failure episodes in a single patient are clinically similar. In the presented case, however, the patient's initial shunt malfunction was caused by incomplete proximal catheter obstruction, so we hypothesize that his dramatic second presentation involved complete occlusion of the new shunt system and rapid increase of intracranial pressure.

In conclusions, we report a case of NPE and possible NSM secondary to shunt malfunction in a pediatric patient. NPE and NSM present challenging critical care problems acutely, but may not cause long-term morbidity if the brain can be adequately protected during the phase of cardiopulmonary compromise. These diagnoses should be considered in cases of rapid-onset shunt failure so that supportive management can be initiated early.

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