Disseminated intravascular coagulation associated with ventriculoperitoneal shunt surgery

Case report

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Disseminated intravascular coagulation (DIC) as a complication of surgery for ventriculoperitoneal (VP) shunts is extremely rare, and only one case has been documented in the literature. The authors present the case of a 9-year-old girl with shunted hydrocephalus who presented with a 3-day history of headaches and vomiting. A head CT showed enlarged ventricles compared with baseline. An emergent VP shunt revision was performed, during which an obstructed proximal catheter was found. Immediately after extubation, the patient became apneic and progressed to cardiopulmonary arrest. A breathing tube was reinserted followed by resuscitation attempts that led to extracorporeal membrane oxygenation. Soon after reintubation, bloody drainage was noted in the endotracheal tube, and subsequent laboratory studies were consistent with DIC. The patient died on postoperative Day 1, and autopsy findings confirmed DIC. Note that DIC is a recognized complication of trauma, particularly with brain injury, but it is rare with neurosurgical procedures. Disseminated intravascular coagulation should be considered if excessive bleeding occurs after any brain insult. (DOI: 10.3171/2009.10.PEDS0974)

Key Words • disseminated intravascular coagulation • shunt • brain

Disseminated intravascular coagulation is a disorder characterized by the widespread activation of coagulation, which results in the intravascular formation of fibrin and thrombotic occlusion of small and midsize vessels. This cascade may lead to multiple organ failure as well as excessive bleeding due to the depletion of platelets and coagulation proteins. Traumatic head injury is a known risk factor for DIC, which has been demonstrated to occur after some intracranial neurosurgical procedures. There has been only one documented report of DIC as a complication of VP shunt surgery. We present the second reported case of DIC occurring after VP shunt surgery in a 9-year-old girl, who presented with a proximal obstruction.

Abbreviations used in this paper: CPS = cardiopulmonary support; DIC = disseminated intravascular coagulation; ET = endotracheal tube; PT = prothrombin time; PTT = partial thromboplastin time; VP = ventriculoperitoneal.

Case Report

History and Presentation. This 9-year-old girl had a history of prematurity (24 weeks), myelomeningocele, Chiari malformation Type II, retinopathy of prematurity, chronic lung disease of prematurity, neurogenic bladder, and shunt-dependent hydrocephalus. She was transferred to our institution from another hospital after presenting with 3 days of headaches and vomiting. She was alert and exhibited her baseline neurological status at the time of presentation. A head CT study performed at the other hospital revealed enlarged ventricles in comparison with baseline (Fig. 1). The patient was taken to the operating room emergently for a VP shunt revision. She was alert and oriented prior to the induction of anesthesia. She had taken no medications at home at the onset of her symptoms. She had a history of the intermittent use of albuterol for the chronic lung disease of prematurity, but she had last used an inhaler more than a month prior to sur-
Ventriculoperitoneal shunt and intravascular coagulation disorder

**Fig. 1.** Left: Baseline head CT showing the ventricular catheter in the right frontal horn. Right: Head CT obtained on presentation with headaches and vomiting, demonstrating enlarged lateral ventricles compared with their appearance on baseline head CT.

surgery. Her preoperative coagulation studies, including PT, activated PTT, and international normalized ratio, were within the normal ranges 30 minutes before surgery.

**Operation.** She was brought to the operating room at 1850 hours. Routine monitors were placed, and anesthesia was intravenously induced with propofol 50 mg, lidocaine 20 mg, and fentanyl 30 µg (patient weight 15 kg). After easy mask ventilation was demonstrated, she received 1.5 mg of vecuronium intravenously, and the trachea was intubatedatraumatically with one attempt of direct laryngoscopy. She inhaled a mixture of isoflurane, air, and O₂ for the maintenance of general anesthesia. She received 1.5 mg of intravenous ondansetron for postoperative nausea and/or vomiting prophylaxis and 375 µg of intravenous cefazolin. During the operation, a proximal obstruction was noted. A new proximal catheter was inserted, and brisk CSF outflow under high pressure was achieved. A small amount of intraventricular hemorrhage was noted but immediately cleared after irrigation with normal saline through the proximal catheter. She was hemodynamically stable throughout the operation. She was given neostigmine and glycopyrrolate for reversal of the neuromuscular blockade, with train-of-four stimulation showing 4 strong twitches. She was extubated at 2025 hours without any difficulty after exhibiting signs of awakening from the anesthesia, including spontaneous respirations and movement of her head and limbs.

**Postoperative Course.** Upon transfer from the operating table to the stretcher for transport to the postanesthesia care unit, she suddenly became apneic. A jaw thrust prompted a few spontaneous breaths. A nasopharyngeal airway was placed into the left nares, and pink fluid was noted from the airway. She no longer generated spontaneous respirations, and bag-mask ventilation was undertaken with evidence of good chest excursion, but she was nonresponsive. Intravenous atropine and muscle relaxant were administered, and she was reintubated for her inability to maintain spontaneous ventilation. Coarse breath sounds were noted bilaterally, with O₂ saturation of 92–93%. On reintubation, she immediately displayed decreased lung compliance with copious fresh, bloody drainage coming from the ET. She progressively had decreases in O₂ saturation and heart rate. Subsequently, she went into cardiopulmonary arrest, and resuscitation was initiated with chest compressions (100 beats/minute) and intravenous epinephrine for the severe bradycardia and asystole noted on electrocardiography. Epinephrine (200 µg) was given intravenously every 3 minutes per the pediatric advanced life support (PALS) protocol for asystole. End-tidal carbon dioxide was not measurable because of a copious amount of bloody drainage coming out of the ET and filling the length of the anesthetic circuit, requiring frequent disconnection and drainage of material.

The pediatric surgical service was urgently called to perform cannulation of the carotid artery and internal jugular vein for the initiation of rapid CPS. Cardiopulmonary resuscitation was performed during the event and during cannulation for a total of 1 hour and 40 minutes. The patient received 3 units of packed red blood cells and 1500 ml of normal saline in the operating room during the resuscitation and cannulation for CPS. An infusion of 1 mU/kg/min of vasopressin and 0.2 µg/kg/min of epinephrine was started when she was placed on CPS. The vasopressin infusion was increased to 2 mU/kg/min, and the epinephrine was increased to 0.3 µg/kg/min. She was transferred to the pediatric intensive care unit in critical condition, and she was switched from CPS to extracorporeal membrane oxygenation. Laboratory studies, which were performed on arrival to the pediatric intensive care unit 2 hours after surgery, revealed prolongation of PT (22.2 seconds) and PTT (> 200 seconds), thrombocytopenia (101,000 mm³⁻¹), and elevation of D-dimer (6.56), which were all consistent with DIC. Factor VII, cryoprecipitate, and fresh frozen plasma were administered, but the patient continued to have bleeding from the ET tube. On postoperative Day 1, she had no evidence of brainstem reflexes perhaps because of hypoxia, and the family decided to withdraw care. An autopsy revealed diffuse alveolar hemorrhages of both lungs, diffuse subarachnoid hemorrhage, intraventricular hemorrhage, and diffuse petechial hemorrhages of the liver, stomach, and rectum. In addition, fibrin thrombi were observed in the right ventricle of the heart and liver, along with glomerular thrombotic microangiopathy (Fig. 2). There was no evidence of an occipital lobe infarction.

**Discussion**

Disseminated intravascular coagulation is an abnormal activation of the coagulation cascade in which there is consumption of the components of the coagulation system and microvascular fibrin deposition. Laboratory studies used to diagnose this condition include the prolongation of PT and PTT, thrombocytopenia, and elevated D-dimer. This condition leads to severe bleeding and can occur in various clinical scenarios, including trauma, sepsis, and obstetrical conditions. Tissue thromboplastin, an activator of the coagulation cascade, is found throughout the body and has been found in high concentrations in the brain. Some authors have hypothesized that injury to the brain causes the release of thromboplastin into the systemic circulation with the potential development of DIC. Thromboplastin has been detected at higher lev-
els in the CSF of patients who have sustained damage to the CNS.3,4 In one prospective study, 340 neurosurgical patients with intracranial hematomas due to head trauma were studied, and those with significant brain tissue injury had laboratory values consistent with DIC.5 Subsequently, Pattisapu et al.10 demonstrated in an animal model that DIC was reproducible with the intravascular injection of CNS-derived thromboplastin.

In a recent retrospective study, Pasternak et al.8 evaluated the incidence of DIC after craniotomy procedures at their institution. These authors found the incidence of DIC within 72 hours of primary craniotomy to be between 13 and 44 cases per 10,000 patients utilizing criteria based on laboratory values. Although the incidence was low, the data revealed an associated mortality rate of 43 to 75% and that traumatic brain injury was a significant risk factor for the occurrence of DIC.

In 1981 Shurin and Rekate12 provided the first case report of DIC as a complication of VP shunt placement in a child. Immediately after surgery generalized bleeding occurred, which included intraventricular hemorrhage, transient peritoneal irritation, and loss of 50% of the red blood cell volume into the subcutaneous tissues. Our case represents the second reported instance of DIC associated with VP shunt surgery, although our patient presented with a proximal shunt malfunction. She had severe bleeding immediately postoperatively, as was evident by copious hemorrhage into the ET tube. Disseminated intravascular coagulation is associated with cardiopulmonary arrest and the use of extracorporeal membrane oxygenation,13,14 but the copious bloody drainage found in the ET tube preceded both of these events in our case. As mentioned, higher levels of thromboplastin have been detected in the CSF of patients with CNS damage, and the time that elapsed between reconnecting the revised VP shunt and extubation in our patient was ~ 30 minutes. This time interval would allow an adequate amount of CSF with elevated levels of thromboplastin to flow into the peritoneum through the shunt and to be resorbed into the systemic circulation. This sequence of events might have led to DIC with ensuing respiratory distress and reintubation, which occurred ~ 15 minutes after extubation. The team initially considered whether the patient’s bloody ET secretions were a result of pulmonary edema, that is, either neurogenic pulmonary edema due to elevated intracranial pressure or postobstructive pulmonary edema following extubation. Neither of these possibilities was consistent with the clinical picture, however. The child was under close observation throughout, and there were no periods of airway obstruction. In addition, the liquid that was coming from the ET was not pink, thin, and frothy as one would expect with pulmonary edema, but rather was the consistency of a pulmonary hemorrhage—that is, red and thick, and it continuously poured from her airway. We also considered the possibility of a traumatic intubation or some type of disruption of a major vessel, but such hypotheses were ruled out on autopsy. While we do not have repeat coagulation labs until after the prolonged resuscitation, the nature of the bleeding from her ET was most clinically consistent with a child with coagulopathy, not pulmonary edema.

Regarding the patient’s brain, autopsy studies revealed diffuse subarachnoid and intraventricular hemorrhage. Although the etiology of DIC in this case is difficult to determine specifically, the patient may have been predisposed to this condition with high intracranial pressure from the shunt malfunction and minor trauma to the brain from revision of the proximal catheter. These insults may have led to the release of thromboplastin from the brain—which is known to contain high concentrations—into the systemic circulation, causing the development of DIC in this child.

Conclusions

Excessive bleeding that occurs in the setting of any brain insult should lead one to consider DIC as a diagnosis. Although rarely associated with neurosurgical procedures such as VP shunt surgery, DIC can nonetheless occur. Early recognition should encourage the prompt administration of clotting factors in an attempt to prevent major complications of excessive hemorrhage.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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