Disseminated intravascular coagulation as a complication of ventricular catheter placement

Case report

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A child who developed generalized bleeding immediately after placement of a ventriculoperitoneal shunt was found to have evidence of disseminated intravascular coagulation (DIC). Infusion of fresh frozen plasma was followed promptly by improvement in laboratory values and cessation of bleeding. Complications of the acute bleeding episode included intraventricular hemorrhage, loss of 50% of the red cell volume into subcutaneous tissues, and transient peritoneal irritation. Defibrination syndrome (that is, DIC) due to release of tissue thromboplastin is a recognized complication of trauma, particularly with brain injury. Defibrination can be induced in experimental systems with administration of very small amounts of thromboplastin, which is present in high concentration in brain tissue. This has not been described previously with minor neurosurgical procedures. The diagnosis of DIC should be considered if excessive bleeding occurs after any brain insult, since early recognition and restoration of normal hemostasis by replacement of clotting factors should prevent major complications due to ongoing hemorrhage.

KEY WORDS • disseminated intravascular coagulation • coagulopathy • hemorrhage • intraventricular bleeding • clotting factor • thromboplastin • ventricular catheter

Disseminated intravascular coagulation (DIC) is a recognized complication of major injuries, especially severe head trauma. It is thought to be due to the release of tissue thromboplastin, with consumption of clotting factors, and tends to be associated with a poor prognosis and a usually fatal outcome. We report here a patient in whom apparent DIC developed immediately after an otherwise uncomplicated procedure for placement of a ventriculoperitoneal shunt. The transient hemorrhagic diathesis resulted in intraventricular hemorrhage that fully resolved without apparent adverse effect on the patient. The abnormalities in laboratory clotting values responded immediately to a single dose of replacement therapy with fresh frozen plasma, and did not recur.

We wish to call attention to the possible development of this uncommon complication of an invasive neurosurgical procedure, because early recognition and appropriate therapy should prevent serious complications and neurological deficit.

Case Report

This 15-year-old boy was diagnosed in infancy as having severe mental retardation. A 17-year-old brother was similarly affected. Both parents and two older siblings (one female and one male) were normal neurologically. Both affected boys had been extensively investigated without a specific diagnosis of the etiology of the mental retardation.

Examination. Peripheral blood chromosomes, blood and urine amino acid profiles, metabolic and lysosomal enzyme screen, and electroencephalograms were all normal. The child had bilateral epicanthal folds, anti-mongoloid slant, bilateral coxa valga, thoracic scoliosis, and growth retardation. He was responsive to social interaction, and comprehended some speech, but could not talk. The right pupil was miotic but reactive to light. There was ptosis and limited upward gaze of the right eye, and a persistent twitch of the right side of the face. Motor examination and tone were intact, except for generally
DIC complicating catheter placement
diminished muscle bulk. He could walk with support.
Sensory examination and tendon reflexes were normal, and there were no abnormal reflexes. Height and weight were well below the 3rd percentile (50th percentile for age 8 years), but the head circumference was above the 98th percentile (59.5 cm). The bone age was 10 years, and there was generalized demineralization of all the bones. Skull films showed widening of the coronal sutures.

Neither the patient nor his family members had any history of an excessive bleeding tendency. The older sibling had a ventriculocav al shunt placed at the age of 9 months and removed because of infection 1 month later, without hemorrhagic complications. The patient and his brother had both sustained several bone fractures, and had fallen on many occasions without major hematomas.

A computerized tomography (CT) scan was performed because of the macrocephaly and widening of the sutures. The lateral ventricles were markedly enlarged, the left more than the right, whereas the third and fourth ventricles were normal (Fig. 1). Blood studies performed before ventriculography were entirely within normal limits, including a platelet estimate. The prothrombin time (PT) and partial thromboplastin time (PTT) were not measured. A ventriculogram was performed with a ventricular needle through a stab incision in the right coronal suture. The cerebrospinal fluid (CSF) pressure was 140 mm H2O. Instillation of contrast material demonstrated marked hydrocephalus of the lateral ventricles above the foramen of Monro, with small third and fourth ventricles.

Operation. A ventriculoperitoneal shunt was placed through a burr hole into the right lateral ventricle. The patient tolerated the procedure without incident, but as he was being removed from the operating table, he was noted to be oozing from all incisions and to have marked swelling along the shunt tubing. Clotting studies drawn at that time showed a PT of 30.2 seconds (control, 12.5 seconds), and a PTT greater than 150 seconds (control, 25.6 seconds). Fibrinogen determined immunologically by a Hyland Laboratories kit* was 76 mg/dl (normal, 170 to 400 mg/dl). Fibrinogen-related antigens (FRA) determined by a Thrombo-Wellcost kit were greater than 40 μg/ml. Platelet count was 120,000/μl.

Postoperative Course. Two units (17 ml/kg) of fresh frozen plasma and 5 mg vitamin K1 were administered in the recovery room. The oozing stopped within 30 minutes. A repeat PT was 13.7 seconds (control, 12.4 seconds), the PTT was 23.2 seconds (control, 21.5 seconds), and the platelet count was 217,000/μl. Twenty-four hours later, plasma fibrinogen was 196 mg/dl, and the FRA were greater than 40 μg/ml.

Repeated coagulation studies over the next 3 weeks were always within normal limits. One week after the shunt placement, FRA were not detectable.

Eighteen hours after surgery, the hematocrit had dropped from 34% to 17%, and the hemoglobin from 11.7 to 5.8 gm/dl. Two units of packed red cells were given. The hematocrit stabilized at 35%, hemoglobin was 11.8 gm/dl, and did not fall again during the next 4 weeks. A CT scan showed blood in the ventricles and around the shunt track in the frontal lobe (Fig. 2).

*Hyland Laboratories kit manufactured by Hyland Diagnostics Division, Travenol Laboratories, Inc., Costa Mesa, California.
FIG. 3. Computerized tomography scan 2 weeks after shunt placement. The blood in the ventricles has completely disappeared. Both lateral ventricles remain enlarged, but are smaller than before the shunt placement, and the asymmetry between the right and left lateral ventricles is no longer evident.

There was marked discoloration along the subcutaneous shunt track. The patient was obtunded and his respirations were irregular, but he improved over the ensuing 48 hours. Approximately 1 week postoperatively, he developed signs of peritoneal irritation, thought to be due to blood that had traversed the shunt tubing from the ventricles into the peritoneal space. Cerebrospinal fluid obtained from the shunt reservoir was sterile at the time of peritoneal inflammation. The peritoneal signs were not accompanied by fever or fall in hemoglobin. Two weeks postoperatively, the ventricles were smaller, with no blood noted in the ventricles on CT scan (Fig. 3), and the CSF pressure was 80 mm H2O. The skin overlying the shunt tubing, which had sustained considerable ecchymosis, broke down, and the subcutaneous portion of the shunt tubing in the chest wall was revised. The neurological condition of the patient at the time of discharge was unchanged from his admission status.

Discussion

Immediately after placement of a ventriculoperitoneal shunt, the patient had evidence of DIC characterized by excessive bleeding, hypofibrinogenemia, mild thrombocytopenia, and the presence in serum of FRA (fibrin split products). Prompt laboratory and clinical recovery followed the transfusion of fresh frozen plasma.

Release of tissue thromboplastin into the circulation activates the intrinsic clotting mechanism, and results in consumption of clotting factors. With low levels of clotting factors, hemostasis is impaired, and clinical bleeding may occur, particularly with trauma or surgery. One hundred years ago, it was shown that large intravascular clots did not develop if the rate of infusion was sufficiently slow, but the blood became incoagulable. By 60 years ago, it was known that this incoagulability was due to the absence of fibrinogen. Subsequent work confirmed the causal relationship between administration of thromboplastic material and defibrination.

Brain is very rich in tissue thromboplastin, as is suggested by the early experiments. The vascular tissues of the choroid plexus and meninges are rich in substances that induce fibrinolysis, such as plasminogen activators. Several patients have been described with DIC after extensive cerebral injury. The coagulopathy has been reversible by administration of fresh frozen plasma, but, in most of the reported cases, the extensive cerebral injuries have been fatal.

We are not aware of previous reports of consumptive coagulopathy induced by the relatively minor cerebral trauma that accompanies insertion of a ventricular cannula for shunt placement. Other tissues are also damaged during the procedure, and multiple sources of tissue thromboplastin may increase the likelihood of development of DIC. Thousands of similar procedures have been performed at this institution without precipitating clinical bleeding diatheses, although laboratory evidence of DIC has not been sought in the absence of excessive bleeding.

There are many situations in which DIC is commonly seen, among them overwhelming bacterial, viral, or fungal infections, malignancy, amniotic fluid embolism, premature separation of the placenta, and transfusion of incompatible blood. Much controversy surrounds the therapeutic aspects of the disorder, but treatment of the underlying condition is clearly necessary for control of the bleeding diathesis. In an acute insult, the consumptive process should not be ongoing, since activated clotting factors are rapidly removed from the circulation by the liver. Thus, the major concern is limiting the extent of damage due to bleeding when the clotting factors are depleted. In this patient, there was extensive intraventricular bleeding at the time of surgery, but no further bleeding occurred after correction of the clotting disorder by administration of plasma.

Precise characterization of the extent to which individual factors are depleted is not possible in an emergency situation such as is posed by a patient with DIC and intracranial bleeding. Some authors have suggested the administration of prothrombin concentrates in this circumstance. Commercial preparations of prothrombin concentrates contain Factors II, VII, IX, and X. Many preparations also contain activated clotting factors as contaminants. These factors aggravate intravascular coagulation, and are contraindicated in DIC. Furthermore, because the concentrates are prepared from pooled plasma, they carry
DIC complicating catheter placement

high risk of transmitting hepatitis. The absence of fibrinogen and of other clotting factors that may be deficient in this situation are potential drawbacks to the effectiveness of prothrombin concentrates for replacement therapy. The broader coverage provided by fresh or frozen plasma, the absence of activated clotting factors, and the lower risk of hepatitis with single donors make this the indicated therapy for restoration of hemostatic competence in DIC induced by minor or major trauma. Platelet transfusions might also be required if significant thrombocytopenia should accompany depletion of clotting factors. However, drugs that stabilize clots, such as epsilon-amino-caproic acid, are contraindicated in DIC because they aggravate the effects of intravascular thrombosis.

Early recognition of the nature of the coagulopathy and institution of appropriate therapy should minimize the long-term adverse effects of this rare complication of surgery.

References

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