Surgical Treatment of Arteriovenous Anomaly in a Hemophiliac Patient

Case Report

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The purpose of this paper is to report problems of management in a case of intracerebral hemorrhage from an arteriovenous malformation in a child with hemophilia.

Hemophilia is an inherited disorder of hemostasis transmitted as a sex-linked, recessive Mendelian trait by the female and affecting the male. The abnormality is a deficiency of Factor VIII (antihemophilic globulin, AHG) which results in a prolonged coagulation time and a life-long tendency to excessive bleeding. Hemorrhage is usually associated with trauma but may be spontaneous, and typically is slow persistent oozing out of proportion to the injury. Subcutaneous, intramuscular, and gastrointestinal hemorrhages are common as are bruising, hematuria, and hemarthrosis. Bleeding into the central nervous system is the most serious complication of hemophilia. Intracranial bleeding occurs in 6% of hemophiliacs and carries a mortality of 70%. Subdural and epidural hemorrhages are the most common forms of intracranial bleeding. Intracerebral hemorrhage comprises one-third of cases of intracranial bleeding.

Case Report

A 2-year-old boy with known severe Factor VIII deficiency (<1%) was admitted on May 15, 1967, in a semicomatose condition with a 7-hour history of nausea, vomiting, and headache. He had had 10 previous admissions for hemorrhage into soft tissues, joints, and scrotum. His family history was negative for bleeding disorders.

Two weeks previous to this admission he had been admitted because of low-grade fever, lethargy, nausea, vomiting, headache, and tonic-clonic left-sided seizures. There was no history of trauma or exposure to infectious disease. He had left hemiparesis and left hyperreflexia. His neck was supple. A brain scan was normal. An EEG showed a mild right temporal lobe focal slow wave abnormality. After 5 days of treatment with glycine precipitate AHG, he had become asymptomatic and was discharged on diphenylhydantoin.

Examination. The patient was a pale, semicomatose child with blood pressure 160/120, pulse 62, respiratory rate 30, and temperature 37.8°C. He had mild papilledema, left hemiparesis with left hyperreflexia, clonus, and positive Babinski response. He appeared to be deteriorating rapidly. Skull x-ray films were normal. A brain scan (Fig. 1) showed increased uptake in the right temporal lobe. Preoperative angiography was not done.

Immediately preoperatively the patient received 50 units per kilogram, or 750 units, of glycine precipitate AHG calculated to provide him with a Factor VIII level of 100%. His subsequent AHG replacement and partial thromboplastin times (PTT) are shown in Fig. 2. During his hospital stay the patient received both glycine precipitated antihemophilic factor and cryoprecipitate concentrate of Factor VIII.

Operation. On May 15, 1967, a few hours after admission a right temporal craniectomy was done under general endotracheal anesthesia. The exposed temporal lobe was dark and hemorrhagic in appearance. A 60 cc semi-solid hematoma was removed through a transcortical incision. A small tangle of vessels was then removed from the anterior depths of the intratemporal lobe defect. The pathological diagnosis was arteriovenous malformation (Fig. 3). Estimated blood loss was 150 cc.

Postoperative Course. On the fourth postoperative day the patient's temperature...
spiked to 39°C. A spinal tap revealed a red blood count of 14,265, a white blood count of 135, 54 mg% glucose, and 130 mg% protein. Spinal fluid cultures were negative.

Three days later, after his fever had subsided, a left femoral cervicocephalic angiogram was done. There was no residual arteriovenous malformation. There was no vessel shift but the right middle cerebral artery was slightly elevated due to residual cerebral edema. Nine days postoperatively he developed a wound hematoma that gradually resolved with local ice packs and correction of his PTT. The patient was discharged 17 days after admission alert and neurologically intact.

Three weeks postangiography, he developed a groin hematoma at the site of a previous catheter insertion. This also resolved with local ice packs and AHG therapy.

He has had three subsequent admissions for possible intracranial hemorrhage, but this condition has never been proven. Tests for anti-AHG antibodies done during a recent admission 4 months postcraniectomy show inhibitors positive in 1:16 dilution, whereas at the time of surgery tests for inhibitors were negative.

One year postcraniectomy he has retarded speech but otherwise is neurologically intact. The operative site is completely normal.

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**Fig. 1.** Anteroposterior (left) and lateral (right) Tc-99m brain scans showing an abnormal accumulation of radioactive material in the right temporal lobe.

**Fig. 2.** Summary of patient’s partial thromboplastin time and antihemophilic globulin therapy.

**Fig. 3.** Arteriovenous malformation showing general features of blood vessels. H. & E., × 400.