Conservative management of an acute spontaneous holocord epidural hemorrhage in a hemophiliac infant

Case report

ERIN N. KIEHNA, M.D.,1 PETER E. WALDRON, M.D.,2 AND JOHN A. JANE JR., M.D.1

1Department of Neurosurgery and 2Division of Pediatric Hematology and Oncology, University of Virginia Health System, Charlottesville, Virginia

Central nervous system hemorrhages are an uncommon but severe complication of hemophilia, occurring in only 2–8% of children with hemophilia. Less than 10% of these CNS hemorrhages are intraspinal. The authors report on their care of an infant with hemophilia A who presented with irritability, meningealism, and decreased spontaneous movement. These symptoms prompted imaging studies, which revealed a spinal epidural hematoma (SEH) extending from C-1 through the cauda equina. The boy was treated with factor replacement and close monitoring. Repeat radiographic imaging 14 days later demonstrated complete resolution, and the patient had returned to his normal baseline status.

A literature review in the modern treatment era revealed 24 cases of SEH in children with hemophilia. Of these 24 cases, 11 underwent laminectomy and 13 received conservative treatment. All conservatively treated patients, 5 of whom had presented with weakness, experienced a full recovery. Of the 11 laminectomy patients, 10 presented with weakness and all but 3 experienced full neurological improvement. These 3 patients were notable for having previously undiagnosed hemophilia. An increased index of suspicion facilitates the essential management features of prompt diagnosis and correction of coagulopathies in children who present with SEHs. The authors apply a multidisciplinary approach involving a pediatric hematologist, neurosurgeon, and pediatric intensive care unit to ensure timely correction of the coagulation disorder, maintenance of adequate factor levels, and close hemodynamic and neurological monitoring. Observation with aggressive correction of coagulopathy is a reasonable treatment choice for hemophilic patients presenting with SEH and a stable neurological examination. (DOI: 10.3171/2010.4.PEDS09537)

KEY WORDS • spontaneous epidural hematoma • hemophilia • spinal cord hematoma

Abbreviation used in this paper: SEH = spinal epidural hematoma.
the examiners, but he would not interact with them. His vital signs were stable. We noted that he preferred to lie supine and was extremely distressed when placed in the seated position. No external signs of a bleed were noted, and there was no joint or bone tenderness. His abdomen was soft, nondistended, and nontender. Neurological examination revealed no focal deficits. Although he would not move spontaneously, his strength in the upper and lower extremities was full and symmetric to stimulation. His tone was normal with deep tendon reflexes of 2+/4 and upgoing toes by plantar reflex.

Imaging for an occult bleed was performed; a CT scan of the head was nondiagnostic. Magnetic resonance imaging of the spine demonstrated a large dorsal extradural mass isointense on T1-weighted images and a heterogeneous signal on T2-weighted images extending from C-1 through the cauda equina, with significant spinal cord compression at its worst at the cervicothoracic junction (Fig. 1). The cord was displaced anteriorly and slightly to the left. There was no definite intramedullary cord signal change.

Laboratory studies showed mild anemia (hemoglobin = 8.6 and hematocrit = 26.5). His admission factor VIII level was < 1%. An inhibitor screen was negative.

Treatment. The patient was admitted to the pediatric intensive care unit for neurological observation, hydration, pain relief, and factor VIII replacement therapy. He was placed on bed rest but not immobilized. Factor VIII was administered as bolus injections at 50 U/kg, which resulted in a level of 94%, and then he was maintained on a constant infusion between 4 and 12 U/kg/hr to keep his factor VIII levels between 80 and 120%. He was started on dexamethasone 0.25 mg/kg every 6 hours. He received Tylenol for pain control. An operating room was placed on standby in the event of neurological deterioration. Within 12 hours of initiating the factor VIII infusion, he was showing increased spontaneous movement of his neck, arms, and legs. On the second hospital day a central catheter line was peripherally inserted to facilitate factor VIII infusions. A steroid taper was subsequently begun, and the boy was transferred to the hospital floor. He remained on a continuous factor VIII infusion for a total of 2 weeks.

Posttreatment. Follow-up MR imaging performed 2 weeks after the patient’s admission showed complete resolution of the hematoma (Fig. 2). He was discharged home on factor VIII boluses (134 U/kg every 8 hours), determined by pharmacokinetic monitoring to maintain levels > 40%.
Conservative management of spontaneous spinal epidural hematomas

One year after admission he was maintained on prophylactic factor VIII boluses every other day. He has had no further spontaneous hemorrhages. He is walking and achieving all of his milestones appropriately.

**Discussion**

Spontaneous epidural hematomas are a serious disease entity that can have a high rate of morbidity and mortality.\(^3\) Although these hemorrhages are extremely rare, infants can be predisposed to an epidural hematoma around the age of 4–6 months as their cervical musculature begins to develop. This early cervical musculature can induce acceleration of the head mass without an ability to control deceleration, resulting in increased tension on the cervical spine.\(^3\) As a result, spontaneous epidural hematomas are most commonly seen at the cervicothoracic junction.\(^3\) Spontaneous SEHs are primarily dorsal hemorrhages. Historically these hemorrhages were believed to be venous in origin. In addition to the force produced by the mass of the head, the absence of valves in the venous plexus coursing through the epidural fat on the dorsal aspect of the cord was thought to be a factor in the dorsal localization of hemorrhages. Increased thoracic pressure, as occurs during crying, coughing, sneezing, or straining, may produce venous backflow, a sudden increase in pressure, and spontaneous rupture of these fragile vessels.\(^4\) The current belief is that venous bleeding is too low pressure to compress the spinal cord and that the spinal cord would tamponade epidural venous bleeding. Instead, bleeding is thought to originate from free epidural arteries, where the higher pressure of arterial bleeding would be sufficient to result in cord compression and the rapid symptom onset commonly seen in these patients.\(^7\)

There is heightened concern over epidural hemorrhages in children with hemophilia given that this condition was historically associated with death or quadriaparesis. The potential morbidity and death have resulted in controversies over whether maximal medical management or surgical intervention is the most appropriate treatment option. We reviewed 33 case reports of hemophilic children with SEHs between 1876 and the present to objectively analyze outcomes by treatment option.

Prior to 1958, purified coagulation factors were not available for transfusion and surgery was not considered a viable option for hemophiliacs as there was no mechanism to stop the bleeding. The first 3 case reports of pediatric intraspinal hematomas were from 1876, 1905, and 1925. Of these 3 children, 2 survived and 1 with ascending paralysis died (Table 1).\(^2,4\) As reported by Craddock et al.\(^4\) the mortality rate for general surgery in hemophiliacs was 66% in 1948. In the 1950s, there were 2 case reports of spinal epidural hematomas in teenagers.\(^28,33\) In both cases, neurological examination and myelography were used to localize the level of spinal cord impingement. A laminectomy in conjunction with whole blood and plasma transfusions was subsequently performed in both patients. Surgery did not result in any neurological improvement, and 1 of the 2 patients died.

After 1958, antihemophilic globulin (factor VIII) became available, followed by Christmas factor (factor IX). With the ability to transfuse discrete factors, the overall mortality rate for general surgery in hemophiliacs decreased to 20%.\(^33\) Two children with SEHs were treated

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age</th>
<th>Bleeding Disorder</th>
<th>Presentation</th>
<th>Level†</th>
<th>Intervention</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Priest, 1935</td>
<td>6 yrs</td>
<td>hemophilia</td>
<td>girdle pain, arms weakness, leg paralysis</td>
<td>UN</td>
<td>none</td>
<td>survival</td>
</tr>
<tr>
<td>Bulloch &amp; Fildes, 1911</td>
<td>16 yrs</td>
<td>hemophilia</td>
<td>ascending paralysis followed by death</td>
<td>UN</td>
<td>none</td>
<td>death</td>
</tr>
<tr>
<td>Janeway, 1905</td>
<td>13 yrs</td>
<td>hemophilia</td>
<td>flacidic paralysis of legs, urinary retention</td>
<td>T7–conus</td>
<td>laminectomy</td>
<td>resolution</td>
</tr>
<tr>
<td>Feissly &amp; Curchod, 1925</td>
<td>16 yrs</td>
<td>hemophilia</td>
<td>back pain, leg weakness, progressing to T-9</td>
<td>T5–L4</td>
<td>laminectomy, bovine/swine AHG</td>
<td>T-9 complete (death mos later)</td>
</tr>
<tr>
<td>Aggeler &amp; Lucia, 1944</td>
<td>16 mos</td>
<td>hemophilia B</td>
<td>paralysis</td>
<td>C6–T1</td>
<td>laminectomy, blood &amp; plasma transfusions</td>
<td>death</td>
</tr>
<tr>
<td>Douglas &amp; McAlpine, 1956</td>
<td>16 yrs</td>
<td>hemophilia A</td>
<td>paraplegia</td>
<td>C6?</td>
<td>plasma, factor IX</td>
<td>NI</td>
</tr>
<tr>
<td>MacFarlane et al., 1957</td>
<td>16 yrs</td>
<td>hemophilia A</td>
<td>quadriplegia</td>
<td>C3–7</td>
<td>PRBCs, factor VIII, laminectomy</td>
<td>foot drop</td>
</tr>
</tbody>
</table>

* Shading variations group those authors referring to or credited with the same cases. Abbreviations: AHG = antihemophilic globulin; NI = neurologically intact; PRBC = packed red blood cell; UN = unknown.
† Question marks indicate best guess based on neurological examination.

J Neurosurg: Pediatrics / Volume 6 / July 2010
with conservative therapy and factor replacement during this time period, with marked improvement in 1 case and sustained quadriparesis in the other. In both of these cases, no imaging was performed to localize the clot. In 1977 Cromwell et al. reported on an infant with a spinal epidural hemorrhage localized by myelography and drained via laminectomy resulting in significant improvement.

By 1980 CT scanners were readily available, followed by MR imaging units in 1990, which allowed non-invasive imaging of the spinal cord. In the modern era, 24

TABLE 2: Literature review of reports on pediatric spinal cord hemorrhages due to hemophilia from 1983 to the present*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age</th>
<th>Bleeding Disorder</th>
<th>Presentation</th>
<th>Level</th>
<th>Surgical Intervention</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdelaal et al., 1994</td>
<td>5 yrs</td>
<td>severe hemophilia A</td>
<td>neck pain/stiffness, progressing to quadriparesis &amp; respiratory failure; C-2 sensory level</td>
<td>C3–T1</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Cuvelier et al., 2006</td>
<td>10 mos</td>
<td>severe hemophilia A</td>
<td>irritability, torticolis progressing to loss of arm strength w/ some preservation of leg strength</td>
<td>C2–T8</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Eftekhar et al., 2003</td>
<td>9 yrs</td>
<td>hemophilia A†</td>
<td>quadriparesis progressing to L-2 incomplete sensorimotor loss</td>
<td>C5–L2 subdural?‡</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Faillace et al., 1989</td>
<td>3 mos</td>
<td>severe hemophilia A†</td>
<td>irritability, leg paralysis, sensory loss w/ a T-5 level (complete sensorimotor loss)</td>
<td>T8–L4</td>
<td>laminectomy</td>
<td>paraplegia</td>
</tr>
<tr>
<td>Heer et al., 2008</td>
<td>12 yrs</td>
<td>mild hemophilia A†</td>
<td>pain, arm/leg weakness, sensory deficits; incomplete injury progressing to complete sensorimotor loss</td>
<td>C5–6</td>
<td>laminectomy, washout</td>
<td>UE paresis, LE paralysis</td>
</tr>
<tr>
<td>Irwin &amp; Attia, 2001</td>
<td>10 mos</td>
<td>mod hemophilia A</td>
<td>irritability, refusal to stand</td>
<td>C4–L1</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Rois et al., 2009</td>
<td>13 yrs</td>
<td>mod hemophilia A</td>
<td>back pain, leg weakness</td>
<td>T5–6</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Stanley et al., 1983</td>
<td>13 yrs</td>
<td>hemophilia A†</td>
<td>back pain, leg weakness, T-8 sensory level</td>
<td>T5–8</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Travis et al., 1996</td>
<td>20 mos</td>
<td>severe hemophilia A</td>
<td>irritability, refusal to stand, constipation, decreased anal tone, neurogenic bladder</td>
<td>L4–cauda</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td>Visconti &amp; Hilgarter, 1980</td>
<td>7 yrs</td>
<td>hemophilia A &amp; B†</td>
<td>ascending paralysis, loss of bowel &amp; bladder function</td>
<td>not specified</td>
<td>laminectomy</td>
<td>paraplegia</td>
</tr>
<tr>
<td>Chrétiennot-Bara et al., 2001</td>
<td>8 yrs</td>
<td>severe hemophilia A</td>
<td>back &amp; left leg pain progressing to paralysis</td>
<td>T11–L5</td>
<td>laminectomy</td>
<td>NI</td>
</tr>
<tr>
<td></td>
<td>12 mos</td>
<td>hemophilia B</td>
<td>irritability, refusal to stand, abdominal pain/distention</td>
<td>C7–conus</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Balkan et al., 2006</td>
<td>17 yrs</td>
<td>severe hemophilia B</td>
<td>neck &amp; back pain, LE weakness, urinary retention</td>
<td>C6–T12</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Bisson et al., 2007</td>
<td>7 yrs</td>
<td>hemophilia B</td>
<td>neck &amp; arm pain</td>
<td>C2–T3</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Hamre &amp; Haller, 1992</td>
<td>9 yrs</td>
<td>hemophilia B</td>
<td>back &amp; neck pain, dorsiflexion weakness</td>
<td>C2–T6</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Hutt et al., 1996</td>
<td>11 mos</td>
<td>severe hemophilia A</td>
<td>irritability, tense abdomen</td>
<td>T9–T12</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Iwamuro et al., 2004</td>
<td>7 mos</td>
<td>mild hemophilia B†</td>
<td>irritability, tense abdomen, left leg paresis</td>
<td>T1–L1</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Jost et al., 1990</td>
<td>7 yrs</td>
<td>severe hemophilia B</td>
<td>back pain</td>
<td>thoracolumbar</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Kalina et al., 2008</td>
<td>7 mos</td>
<td>severe hemophilia A†</td>
<td>irritable, refusal to crawl</td>
<td>C2–L4</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Narawong et al., 1988</td>
<td>8 mos</td>
<td>hemophilia A</td>
<td>irritability, decreased neck mobility, refusal to crawl</td>
<td>C2–T3</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Noth et al., 1993</td>
<td>4 yrs</td>
<td>hemophilia A</td>
<td>neck pain</td>
<td>T1–T5</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>Sheikh &amp; Abildgaard, 1994</td>
<td>7 yrs</td>
<td>severe hemophilia B</td>
<td>back pain, grip &amp; dorsiflexor weakness, decreased neck mobility</td>
<td>C3–conus</td>
<td>none</td>
<td>NI</td>
</tr>
<tr>
<td>present case</td>
<td>5 mos</td>
<td>hemophilia B</td>
<td>irritability, refusal to move arms &amp; legs</td>
<td>C1–conus</td>
<td>none</td>
<td>NI</td>
</tr>
</tbody>
</table>

* LE = lower extremity; mod = moderate; UE = upper extremity.
† Hemophilia undiagnosed prior to this presentation.
‡ Represents best guess based on neurological examination.
cases of spinal cord epidural or subdural hematomas have been identified in the pediatric hemophilia population (Table 2). Eleven cases have been managed with surgical laminectomy and clot evacuation.3,12,14,18,23,25,29,30,33,40,46,48 Three patients in these cases had a complete neurological deficit preoperatively and persistent neurological deficits postoperatively;3,12,46 the remaining 8 patients recovered fully. Thirteen cases, including our own, were conservatively managed with factor replacement, blood transfusions, and close observation.3,9,22,24,26,29,30,34,35,42 Of the patients who were treated conservatively, 5 had presented with motor weakness and all recovered fully.

In only 2 of the cases we reviewed was there a known history of trauma in the form of a fall or lumbar puncture. The patients in both of these cases underwent a laminectomy with improvement in neurological function. In children it has been pointed out that “it is difficult to draw the line between mild trauma and the activities of daily living.”21 In our literature review, 40% of the patients’ hemorrages could be retrospectively attributed to normal childhood activities. In this subset of patients, all presented within 48 hours of the injury and half of them underwent laminectomies. All but 1 recovered fully; his examination revealed an unchanged status from the time of presentation.

The remaining 40% of patients had no known inciting factor. These atraumatic cases occurred in infants (5–20 months) who presented with meningeal signs and irritability, or with an otherwise normal neurological examination. These children were symptomatic for anywhere from a few hours to 9 days, signaling that a high level of clinical suspicion for bleeding even in atypical locations must be maintained in hemophilic patients, especially infants, in whom usual signs and symptoms can be missing. None of these infants had lasting deficits.

Although a surgeon’s natural instinct may be to operate when presented with a child with an epidural hematoma,4,30 our experience and the literature indicate that the pediatric hemophilia population displays remarkable resilience and may not require surgery. As Foo and Rossier20 noted in 1981, patients younger than 9 years of age with a spontaneous spinal epidural hemorrhage have a better potential for partial or complete neurological recovery than adults, especially when the sensorimotor examination in patients younger than 9 years is incomplete. Indeed, studies that reported only a 50% recovery rate included patients who had been treated before factor transfusions or modern imaging were available. Our review of 24 cases over the last 30 years demonstrated an 88% recovery rate regardless of the treatment option and 100% recovery in patients who were maximally medically treated (without surgery). These data steer us away from surgery when a patient’s condition is stable.4 Decompressive laminectomy carries a significant risk of spinal deformity—as high as 100% for children with extensive cervical laminectomies and 36% for children with lumbar deformities.39 Subsequent corrective surgery for spinal deformity is associated with a further risk of morbidity.

The 3 patients with lasting neurological deficits were unknown hemophiliacs at the time of presentation, with complete sensorimotor loss prior to surgery. This presentation underscores the importance of a full neurological examination, prompt imaging, and laboratory studies, especially a coagulation panel, when a child presents with an SEH. Any significant coagulopathies should be corrected expeditiously to limit epidural expansion. Bed rest with or without immobilization may be appropriate until the coagulopathy is reversed and/or any neurological deficits stabilize or improve. The use of steroids in the setting of acute spinal cord injury is controversial. Adverse effects were not noted with steroid administration in our case, nor historically in a case of conservative management.3

We believe in multidisciplinary treatment involving a pediatric hematologist, neurosurgeon, and pediatric intensive care unit to ensure timely correction of the coagulation disorder, maintenance of adequate factor levels, and close hemodynamic and neurological monitoring. Observation remains a safe and effective treatment choice for patients with a stable neurological examination and corrected coagulopathy.

Disclosure

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

Conception and design: Kiehna, Waldron. Acquisition of data: Kiehna. Analysis and interpretation of data: all authors. Drafting the article: Jane, Kiehna. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Study supervision: Jane.

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E. N. Kiehna, P. E. Waldron, J. A. Jane Jr.

Manuscript submitted February 1, 2010. Accepted April 1, 2010.

Address correspondence to: John A. Jane Jr., M.D., Department of Neurosurgery, University of Virginia Health System, Charlottesville, Virginia 22908. email: johnjanejr@virginia.edu.