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CASE REPORT

Rachel S. Rosen,[1] *Vernon Armbrustmacher,*[1] *M.D., and Barbara A. Sampson,*[1] *M.D., Ph.D.*

Spontaneous Cerebellar Hemorrhage in Children

ABSTRACT: Spontaneous cerebellar hemorrhages are a rare but often fatal occurrence in children. Although there are several predisposing factors such as blood dyscrasias or astrocytomas, the most common cause of cerebellar hemorrhage in an otherwise healthy child is the rupture of a vascular malformation. We reviewed the files of the Office of Chief Medical Examiner of the City of New York and found four such instances over a period of less than two years. We present these here and outline the approach the forensic pathologist should take in evaluating such deaths.

KEYWORDS: forensic science, neuropathology, pediatrics, spontaneous cerebellar hemorrhage/hematoma, arteriovenous malformation, vascular malformation

Spontaneous cerebellar hemorrhage in children is a rare and catastrophic event with a high rate of mortality, even greater than that of cerebral hemorrhage (1). The factors that predispose some children to a spontaneous cerebellar hemorrhage include vascular malformations in the posterior fossa, blood disorders such as hemophilia and acute leukemia (2), astrocytomas with abnormal vasculature (3,4) and angiomas (4,5).

The most common underlying cause of spontaneous cerebellar hemorrhage in adults is hypertension, which is rarely a factor for children or adolescents. The most common cause of such a hemorrhage in children is arteriovenous malformations, which are caused by abnormal arteriovenous shunts. The resulting increased venous flow under high pressure results in thickened, twisted and "arterialized" veins that are prone to rupture. In some instances, no predisposing factor can be found, and it is thought that a vascular malformation once was present and responsible for the bleeding, but ultimately was destroyed or obscured by the hemorrhagic event.

The symptoms of a spontaneous cerebellar hemorrhage include the acute onset of severe headache, which can be followed by nausea, vomiting, ataxia and vertigo, leaving the person in varying degrees of consciousness (6). The size of the hematoma is correlated with the severity of the initial physiological and neurological signs of rupture. Death can occur within hours or several days. Sudden death, gradual deterioration, or prolonged survival are possible (7). Certain surgical procedures, such as clot evacuation, may be performed with success depending on the size and location of the hemorrhage, as well as the speed with which it was diagnosed.

The prognosis and clinical course of the hemorrhage depends on how the mass in the posterior fossa redistributes itself to alleviate intracranial pressure. Either the mass can rupture into the fourth ventricle, thereby increasing pressure on the brain stem, or it can block the flow of cerebrospinal fluid causing hydrocephalus. In addition, herniations, transtentorially upward or downward through the foramen magnum, may occur (8). This paper describes four fatal instances of pediatric spontaneous cerebellar hemorrhage seen within two years by the Office of Chief Medical Examiner of the City of New York.

[1] Office of Chief Medical Examiner of the City of New York, 520 First Ave., New York, New York 10016.

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Case #1

A 5-year-old boy arrived home from school complaining of a headache. He went to sleep and his mother found him unresponsive soon after. He had no previous history of illness or signs of trauma.

The 1250-g brain revealed a 5.5 × 2.5 × 3.0 cm red-black coagulated hematoma in the right hemisphere of the cerebellum, extending from the superior vermis laterally through the deep white matter to the subarachnoid space at the posterior and lateral hemispheric surface (Fig. 1*a*). A 0.5 cm circular structure, consistent with a relatively large blood vessel, was embedded in the hematoma. The fourth ventricle was partially compressed and the dorsolateral surface of the pons and medulla were compressed. The surfaces of the posterior right cerebellar hemisphere, the superior cerebellar vermis and medial surface of the left cerebellar hemisphere were covered by an approximately 0.3 cm thick layer of red-black subarachnoid hemorrhage. Microscopic examination of the cerebellum showed abnormal blood vessels with thick walls and dilated lumina within the hematoma and at its margin in the leptomeninges and cerebellar parenchyma (Fig. 1*b*). The brain and spinal cord showed no other abnormalities. The remainder of the autopsy was unremarkable.

The cause of death was determined to be acute cerebellar hemorrhage due to ruptured vascular malformation.

Case #2

A 6-year-old boy complained of a headache while having dinner and went to bed when it worsened. A family member went to check on him a half-hour later and found him unresponsive and frothing from the nose and mouth. At this point, EMS was called and the

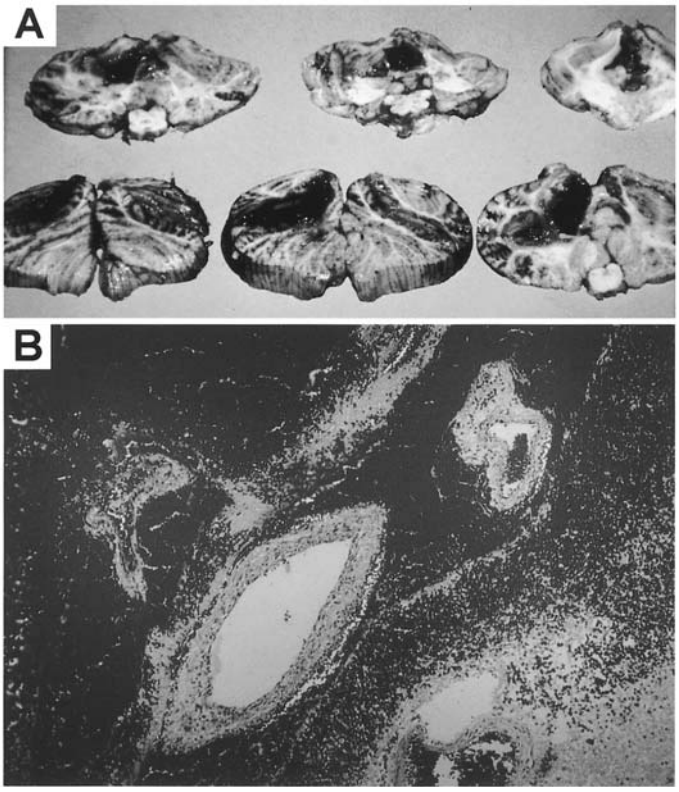

FIG. 1a—*(a) Gross photograph of cross sections of cerebellum from Case 1 showing marked cerebellar hemorrhage. (b) Photomicrograph of abnormally thick blood vessels and dilated lumina within the hematoma (trichrome stain, 25× magnification).*

child was taken to the hospital. His temperature was 96.3°F, and he had no heart beat, respirations, or blood pressure. The pupils were fixed and dilated, and there was generalized hypotonia and areflexia. He had no injury.

The brain weighed 1410 g. The dura was unremarkable. After fixation, coronal sections of the cerebrum revealed no lesions in the cortex, white matter, or deep nuclear structures. The right cerebellar hemisphere was collapsed and covered with patchy dark brown subarachnoid hemorrhage. A cavitary lesion of the white matter, measuring 3.0 × 3.0 × 3.0 cm, with peripheral dilated small vessels was focally lined by dark brown clot in the right lobe. Microscopic examination of the cerebellar tissue around the cavity showed recent hemorrhage and dilated, focally congested, thickened vessels with no distinct malformation. The frontal lobe, hippocampus, basal ganglia and medulla, as well as all other organs, were unremarkable.

The cause of death was determined to be intracerebellar hemorrhage due to a vascular malformation.

Case #3

A 9-year-old girl with no known previous medical history went to school feeling fine. When she returned home, she complained of a severe headache and went to her room. Her mother checked on her at which point the girl collapsed. EMS responded and found the girl with very labored breathing and a faint pulse. She was intubated and resuscitation was initiated and continued in the emergency room where she expired. The decedent was involved in a minor motor vehicle accident two months prior, but was uninjured.

At autopsy, no injuries were detected. The 1200 g brain was examined after fixation and a 0.2–0.3 cm thick, 3.5 cm in diameter, round layer of soft, acute, subdural blood was loosely adherent to the inner surface of the right posterior fossa dura. In addition, there was a thin acute subarachnoid hemorrhage over the posterior-inferior surface of the right cerebellar hemisphere. A prominent tentorial groove indented the left lateral surface of the superior cerebellar vermis approximately 1.5 cm from the midline. Sections exhibited a 3.5 × 3.0 × 1.5 cm hematoma cavity in the deep white matter of the right cerebellar hemisphere. The hematoma extended through the posterior-lateral folia and into the overlying subarachnoid space where a leptomeningeal defect communicated with the subdural space, producing the above noted subdural hemorrhage. The fourth ventricle was compressed and shifted slightly to the left.

Microscopic examination of the cerebellum disclosed abnormally thick, dilated blood vessels, some with elastica, in the white matter around at the margins of the hematoma. Some of these vessels were necrotic and inflamed (considered to be secondary changes). Remnants of the vascular wall were seen within the hematoma.

The spinal cord and dura were unremarkable. The lung parenchyma was spongy, with large amounts of blood tinged fluid exuding from cut surfaces. All other organs were found to be unremarkable.

The cause of death was determined to be spontaneous rupture of a cerebellar vascular malformation.

Case #4

A 9-year-old boy complained of a headache and nausea while walking to school. At school, he lay down and had a seizure and cardiopulmonary arrest. He had seemed well the morning of his death, and had eaten breakfast as usual. Two days prior, he had hit his head on a table while roller-skating but seemed to be fine afterward.

At autopsy he had a 1-in. contusion of the right frontal area with subscalpular hemorrhage. No other trauma was present. Examination of the 1590 g brain revealed acute subarachnoid hemorrhage over both cerebellar hemispheres. A hematoma, originating in the white matter of the left cerebellar hemisphere and measuring 3.0 × 1.5 cm, ruptured posteriorly in the subarachnoid space and crossed into both cerebellar hemispheres. Internally, the hematoma extended medially and downward into the vermis onto the roof of the fourth ventricle. The lateral ventricles contained a small amount of blood.

On microscopic examination of the brain, multiple, dilated and thick walled hyalinized vessels were found within the cerebellar parenchyma. The neuropil surrounding the blood vessels exhibited gliosis and focal hemosiderin deposition. The remainder of the brain was unremarkable.

Acute subarachnoid hemorrhage was present on the spinal cord around the cervical and upper thoracic areas, extending focally outside the dura through an approximately 1.0 cm linear artifact. All other organs were found to be unremarkable.

The cause of death was arteriovenous malformation of the cerebellum with acute hemorrhage.

Discussion

Cerebellar hemorrhages in children are a rare occurrence but can be fatal as demonstrated by the four instances seen by the Office of Chief Medical Examiner of New York City in less than two years. The mortality rate has been estimated to be approximately 73.5% (2). Although there has been documentation of cerebellar hemor-

rhages in the clinical literature, there is little description in the pathology literature.

To summarize, the decedents were all healthy children who experienced a sudden onset of symptoms consistent with increased intracerebral pressure, without a history of functionally important trauma. Their deaths occurred rapidly, indicating that the hemorrhaging was quite rapid. Examination of the brains showed abnormally thickened vessels in all cases, providing a pathologic basis for the hemorrhages. Extensive sectioning of the cerebellum was performed in order to detect these abnormal blood vessels. In Case 2, the abnormal vessels were subtle, seen only in one section, and easily could have been missed. In the fourth case, many more distorted vessels were visible histologically. In the first and third cases, thickened vessels were found within the hematoma, as well as in other sections. These vessels could not definitively be called arteriovenous malformations and were labeled simply as vascular malformations.

When the forensic pathologist is presented with a pediatric death due to a cerebellar hemorrhage, there are several steps to take in the investigation. The medical history may reveal other medical problems, such as blood dyscrasias or tumor, that may predispose to hemorrhage. A history of trauma should be sought, and a complete autopsy, including radiographs, should be performed. At autopsy, if an intraparenchymal hemorrhage is suspected, a complete neuropathological exam should be performed. The brain should be fixed in formalin and then sectioned. Comprehensive gross photos of the specimen should be taken. Extensive histological sampling of the cerebellum is essential to seek abnormal blood vessels which may be obscured or destroyed by the hemorrhage. In addition, toxicological screening should be carried out to test for the presence of cocaine or other drugs, which could have caused or contributed to the hemorrhage.

Ruling out child abuse in these cases is an important consideration. Subdural hemorrhage, in particular, is usually a result of trauma, but it is possible for an intraparenchymal hemorrhage to rupture into the subarachnoid and subdural spaces. With cerebellar hemorrhages, it is particularly important to rule out fractures of the posterior cranial fossa. None of our cases raised any suspicion of abuse or functionally important trauma.

Cerebellar hemorrhage can result from vascular malformations which do not necessarily fulfill the criteria for arteriovenous malformations. It is important that the forensic pathologist be aware of this rare condition and to know the steps to take to determine the origin of the bleeding. An extensive neuropathologic exam is the key to finding this subtle cause of bleeding in an otherwise healthy child.

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Additional information and reprint requests:
Barbara A. Sampson
Office of Chief Medical Examiner of the City of New York
520 First Ave.
New York, New York 10016
E-mail: basampson@yahoo.com