Intraventricular hemorrhage in a term neonate secondary to a third ventricular AVM

Case report

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An initially healthy infant born of an uncomplicated full-term gestation was brought for evaluation of the acute onset of vomiting, irritability, lethargy, and opisthotonus at 14 days of age. Computerized tomography demonstrated an intraventricular hemorrhage. Arteriography defined an angioma on the roof of the third ventricle which was successfully removed via the transcallosal interfornicial approach on the 34th day of life. Other than an easily controlled seizure disorder, the postoperative course was uneventful. At 8 months of age the child is developing normally. Arteriovenous malformations should be considered in the differential diagnosis of intraventricular hemorrhage in full-term neonates without predisposing trauma or bleeding diathesis. High-speed digital subtraction arteriography may be used to screen for this diagnosis. The transcallosal interfornicial exposure offers a satisfactory approach for excising third ventricular angiomas in young infants.

KEY WORDS • intraventricular hemorrhage • arteriovenous malformation • digital angiography • neonate

Intraventricular hemorrhage (IVH) in the full-term neonate is an uncommon event without predisposing factors such as bleeding diatheses, hypoxic-ischemic insults, respiratory distress, intra- or postpartum trauma, or tumor.4-6,12,13,15-17 The origin of the bleeding in these cases is generally considered to be either the choroid plexus or a vestigial germinal matrix, except in the rare case of tumor or angioma.5,11,16,18 There are several reported cases of IVH in full-term neonates with no predisposing factors.4,6,15,17 In general, these neonates did well clinically and rarely underwent angiography. Schum, et al.,18 reported the case of a patient similar to the one reported here who had an angiographically demonstrated third ventricular arteriovenous malformation (AVM); however, operative intervention was not mentioned and the follow-up period was brief.

We are reporting the case of a full-term infant who suffered an IVH on the 14th day of life. Digital subtraction arteriography demonstrated an angioma on the roof of the third ventricle. She underwent a successful transcallosal interfornicial excision of the lesion and is developing normally at 8 months of age.

Case Report

A full-term baby girl was born via an uncomplicated spontaneous vaginal delivery after a gestation complicated only by maternal sinusitis during the second trimester. Her 31-year-old mother had had six previous pregnancies, three of which came to term. The mother had received a course of ampicillin and a cephalosporin for the sinusitis. The child's head circumference was 35.5 cm at birth and the neurological examination was normal. From the time of discharge on the 2nd day post partum until the age of 2 weeks, she was vigorous, fed well, and appeared healthy. On the 14th day of life, she became acutely irritable and began vomiting. Shortly thereafter, lethargy and opisthotonic posturing ensued. At another hospital she was noted to have intermittent opisthotonus and symmetrical hyperreflexia. Her pulse was 140 to 160/min and her systolic
blood pressure was 120 to 130 mm Hg. She had no respiratory distress. Laboratory studies included normal prothrombin and partial thromboplastin times, a platelet count of 745,000/cu mm, a hematocrit of 42%, and normal electrolyte levels. A lumbar puncture demonstrated bloody cerebrospinal fluid with 29,000 red blood cells/cu mm and 110 white blood cells/cu mm. A computerized tomography (CT) scan of the head revealed blood in all four ventricles, with slight ventriculomegaly but no intraparenchymal hemorrhages. There was no history of trauma and skull films were normal. Her three siblings, aged 3, 6, and 8 years, were healthy, and there was no family history of bleeding diathesis. She remained clinically stable except for a brief apneic episode which responded to tactile stimulation. She was transferred to the Yale-New Haven Hospital Pediatric Intensive Care Unit the following day.

Examination. Pulse ranged from 90 to 100/min and systolic blood pressure was 130 to 140 mm Hg. Respirations were somewhat irregular but she had no respiratory distress. She weighed 4.2 kg and was afebrile. The head circumference was 37 cm, no trauma was evident, and the anterior fontanel was full, although soft. Lethargy, opisthotonus, and intermittent conjugate tonic downward gaze were apparent. She intermittently fixed her gaze and followed movement, and she had spontaneous eye opening with conjugate eye movements. The pupils were equally reactive to light, corneal reflexes were intact, facial movements were symmetrical, and the gag reflex was intact. She spontaneously moved all extremities equally well, but had increased passive tone and symmetrical hyperreflexia with bilateral ankle clonus and extensor plantar responses.

A repeat CT scan revealed no change in ventricular size nor any evident bleeding source. Electroencephalography did not demonstrate seizure activity. The patient progressively improved neurologically. Serial lumbar punctures were required to control a moderate degree of communicating hydrocephalus. Opening pressures ranged from 130 to 210 mm H2O. On the 16th day of life, she underwent digital subtraction cerebral arteriography (6 images/sec) which demonstrated early filling of the internal cerebral veins and the possibility of a small vascular malformation in the roof of the third ventricle. To further define the lesion, the study was repeated at 4 weeks of age. This second study was performed with both digital subtraction arteriography (6 images/sec) and conventional film-screen arteriography imaging (3 images/sec). Both demonstrated a small angioma on the roof of the third ventricle fed by the posterior choroidal arteries. Early drainage through the internal cerebral veins was again noted (Fig. 1).

Operation. Preoperatively, the patient was begun on a course of phenobarbital. She underwent surgery at 5 weeks of age. The angioma was excised through a right frontal craniotomy via an interhemispheric transcalsal interfornicial approach. Histological examination of the specimen revealed hemosiderin deposits and abnormal vascular structures suggesting an AVM.

Postoperative Course. The postoperative course was uneventful except for intermittent seizures on the first 2 days, which required additional loading with phenobarbital (10 mg/kg loading dose plus 5 mg/kg/day maintenance dose). Later, because of persistent intermittent seizure activity, diphenylhydantoin was added (15 mg/kg loading dose and 5 mg/kg/day maintenance dose). The patient experienced no further seizure activity and
diphenylhydantoin was tapered off prior to her discharge on the 10th postoperative day. Follow-up digital cerebral arteriography before discharge demonstrated no residual AVM (Fig. 2). Serial postoperative CT scans have shown a gradually resolving right frontoparietal subdural hygroma. Follow-up neurological examination of the patient at age 4 and 6 months revealed a normal, vigorous, socially active infant. Her pupils fixed and followed movement well, her muscles had normal passive and active tone, and she was able to sit with minimal support.

At 8 months of age, the patient underwent developmental evaluation by a pediatric psychologist who was not familiar with the patient's perinatal history. Bayley's Scales of Infant Development were used. The Bayley development indexes are distributed in the same manner as the intelligence quotient, with a mean of 100 and a standard deviation of 16 in the general population. On the Bayley Scales, the patient's performance would have been typical of a normal child of her age group with a mental development index of 98 and a motor development index of 106.

**Discussion**

In contradistinction to intraventricular hemorrhage (IVH) in the preterm infant, IVH in the full-term infant is an unusual occurrence. A review of the literature reveals a total of 95 reported cases of IVH in full-term neonates. Clinical data for comparison were lacking in 15 of these cases. Of the remaining 80 cases, 67 had predisposing factors such as respiratory distress syndrome, severe congenital heart defects, significant intra- or postpartum trauma, asphyxia, or bleeding diathesis. Necropsy data available in 45 of these cases implicated either the choroid plexus or vestigial remnants of the germinal matrix as the source of the hemorrhage in the majority of these patients. These findings presumably represent part of the spectrum of IVH commonly seen in premature infants. No angiomas were reported in this group. A frontal glioblastoma multiforme was responsible for an IVH in one case. The literature review revealed 13 cases which had no evident predisposing factors such as those listed above. Of these, cerebral angiography was performed in only three patients. The angiogram was unrevealing in the case reported by Mitchell and O'Tuama. The case reported by Schum et al. involved a full-term infant who was delivered by Caesarean section due to dystocia, but was otherwise unstressed. This infant bled on the 4th day post partum, and cerebral arteriography demonstrated an AVM in the region of the third ventricle, fed by the posterior choroidal arteries and draining directly into the vein of Galen. A follow-up CT scan at 3 weeks of age revealed resolving IVH and dilation of the vein of Galen. The infant was alert, appropriately responsive, and gaining weight. No operative intervention or further follow-up was mentioned. Maki and Shirai reported the angiographic findings of four full-term neonates suffering from IVH. Three had peripartum asphyxia and one had undergone a breech delivery but was otherwise unstressed. The angiograms of these four children demonstrated hydrocephalus, stasis of contrast medium in the choroid plexus of the lateral ventricles, and early filling of the deep venous system.

A review of the larger series of childhood AVM's suggests that patients who present with bleeding have at least a 5% per annum chance of rebleeding, and that total extirpation of operable angiomas would be the treatment of choice. Although there are no such series of AVM's in early infancy, one would expect the risk of rebleeding at some time in the child's life to be quite high. Even though this type of lesion must be extremely rare, a full-term neonate suffering an IVH who has no significant predisposing factors for neonatal IVH should be investigated with angiography. Digital subtraction arteriography should demonstrate even small rapid-flow angiomas because it allows high-speed imaging with very low amounts of contrast material. The latter is an important factor in the young infant. If an operable AVM is found, it should be excised.

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The safety and efficacy of the interhemispheric transcalsosal interfornicial approach to lesions of the third ventricle have been demonstrated. Apuzzo et al. observed no long-term clinically apparent morbidity among 11 cases of various third ventricular lesions operated on via this approach. Six of their cases underwent rigorous neuropsychometric testing postoperatively, and no significant deficits in interhemispheric transfer or memory could be demonstrated. Jeewes et al. noted only mild defects in interhemispheric transfer of tactile data at long-term follow-up examination of a 14-year-old girl with a colloid cyst of the third ventricle excised via this approach. Winston et al. studied four children after transcalsosal interfornicial

**Fig. 2. Follow-up digital subtraction cerebral arteriogram at 1 week after surgery.**
Intraventricular hemorrhage in a term neonate

operations for third ventricular lesions and found no significant neuropsychological sequelae. The interhemispheric transcallosal interfornicial approach allowed total extirpation of an angioma on the roof of the third ventricle in our patient. At the age of 8 months, this child appears to be developing normally.

Postoperative subdural hygromas have been reported by Jooma and Grant in two infants undergoing transcallosal excision of third ventricular choroid plexus papillomas. One of these patients required bilateral subdural peritoneal shunts. The extra-axial collection in our patient remains asymptomatic.

Conclusions

Full-term neonates developing intraventricular hemorrhages in the perinatal period without apparent predisposing factors should be investigated further for the possibility of an operable vascular lesion. High-speed digital subtraction arteriography may be required for diagnosing these cases. The interhemispheric transcallosal interfornicial approach offers excellent access to third ventricular lesions in young infants, apparently with minor morbidity.

References


Manuscript received February 4, 1985.
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