Case Report

Posterior Fossa Hemorrhage in a Term Neonate with Hemophilia A

Ping-Hung Tsai1,2*, Hui-Ju Chen1, Che-Sheng Ho1,3, Nan-Chang Chiu1,3

1Department of Pediatric Neurology, MacKay Children’s Hospital, Taipei, 2Keelung Hospital, Ministry of Health and Welfare, 3Department of Medicine, MacKay Medical College, Taiwan

Abstract

Posterior fossa hemorrhage is rare in term baby and difficult to assess. The clinical signs are nonspecific and usually delay the diagnosis. We present a 5-day-old male neonate of posterior fossa hemorrhage with the initial presentations of fever and seizure and early deduced by cranial ultrasonography findings as hyperechoic, asymmetric, ill-defined density and complicated with hydrocephalus. Magnetic resonance imaging of the head verified the diagnosis. Hemophilia A was confirmed thereafter by serology.

Keywords: Hemophilia, intracranial hemorrhage, posterior fossa, term neonate, ultrasonography

INTRODUCTION

Intracranial hemorrhage (ICH) of the posterior fossa is rare in term neonates. The clinical symptoms of posterior fossa hemorrhage are nonspecific and often similar to meningitis, sepsis, and extracranial bleeds. The typical signs include pales, vomiting, hypotonia, tense anterior fontanelle, increased of head circumference, seizure, and respiratory distress.1,2 The dorsal respiratory group located in the medulla oblongata is the most important respiratory center and can result in dramatic presentation to death when being involved. It is unknown about the accurate incidence of ICH of the posterior fossa because asymptomatic posterior fossa hemorrhage was underestimated and rarely diagnosed in term neonate.1,3

The risk factors for ICH in term neonate include delivery methods, bleeding disorder, maternal hypertension, perinatal asphyxia, and low Apgar score.4 Hemophilia resulting in ICH of term neonate is uncommon. The incidence was reported from 3.4% to 4.0% of the newborn with hemophilia.5

We reported a case of term neonate of hemophilia A with posterior fossa hemorrhage detected by cranial ultrasonography (USG).

CASE REPORT

The male neonate was born at 40 + 1 weeks’ gestational age to a 30-year-old mother, gravida 1, para 0 with a birth weight of 3030 g through cesarean section. The pregnancy had been complicated by maternal right Bell’s palsy during gestation 30–35 weeks, and the delivery course was prolonged for 24 h, yet the Apgar scores were 9 at 1 min and 10 at 5 min, respectively. The neonate received prophylactic intramuscular Vitamin K after birth. The patient was well-being initially, but on the 5th postpartal day, he was noted to have fever and convulsion with limbs extension, accompanied with desaturation, and decreased activity. Neonatal seizure was suspected, and after loading dose of luminal at the local clinic, he was transferred to our hospital.

His maternal uncle had a history of unknown congenital disease with mental retardation; otherwise, the family history was unremarkable.

On physical examination, he had body temperature 38.5°C, pulse 173 beats/min, respiratory rate 50 breaths/min, blood pressure 96/66 mmHg, oxygen saturation 98%–99% on room

Address for correspondence: Dr. Ping-Hung Tsai, Department of Pediatric Neurology, MacKay Children’s Hospital, No. 92, Sec. 2, Chung-Shan N. Road, Taipei City 10449, Taiwan. E-mail: mmhpnei@gmail.com

Access this article online

Quick Response Code:  
Website: www.jmuonline.org

DOI: 10.4103/JMU.JMU_10_18

How to cite this article: Tsai PH, Chen HJ, Ho CS, Chiu NC. Posterior fossa hemorrhage in a term neonate with hemophilia A. J Med Ultrasound 2018;26:56-8.
air, and his body weight 2800 g. The anterior fontanelle was flat, and his activity decreased with hypotonia. There was polydactyly of the right hand. The remaining of the physical examination was unremarkable.

The initial laboratory tests showed hemoglobin 12.5 g/dL and platelet count 322 K/µL. The spinal tapping for ruled out central nerve infection showed bloody cerebral spinal fluid. The cranial USG was arranged on admission day 1 after the cerebrospinal fluid (CSF) study. The coronal view along the plane of the choroid plexus and the midline sagittal view through the anterior fontanel [Figure 1] revealed obstructive hydrocephalus with enlarged bilateral lateral and 3rd ventricles and ill-defined hyperechoic density over the cerebellum with upward compression of the 3rd ventricle and the 4th ventricle hardly to identified. The resistive index (RI) was 0.63 (normal range: 0.6–0.9), which is through the midline sagittal view and spectral waveforms of the anterior cerebral artery. Magnetic resonance imaging (MRI) of the head [Figures 2 and 3] was soon arranged and revealed a subdural hematoma in the region of the left cerebellar hemisphere causing compression and displacement of the 4th ventricle and resulted in obstructive hydrocephalus with dilatation of the lateral and 3rd ventricles.

The patient was intubated with ventilator support due to frequent apnea and cyanosis. Bleeding diathesis evaluation showed his prothrombin time was 10.9 s (reference range: 8.0–12 s), and an activated partial thromboplastin time was >120 s (reference range: 23.9–35.5 s). Factor VIII (FVIII) activity was <6.6% (reference range: 60%–150%). Hemophilia A was diagnosed.

Owing to personal reasons and the economic conditions of family, the parents refused surgical treatment and recombinant FVIII concentrate therapy. The patient was expired at 15-day-old unfortunately.

**DISCUSSION**

ICH in the neonate with hemophilia is rare and focus on posterior fossa hemorrhage in this group had not been specifically discussed in previous literature.[2,6,7] A case report presented a hemophilia term baby born through vacuum-assisted vaginal delivery without positive family history. His posterior fossa hemorrhage improved after surgical and intravenous recombinant FVIII replacement therapy.[6]

For the newborns with hemophilia, the safest mode of delivery still exists controversy. Kulkarni and Lusher reported that ICH in a newborn with hemophilia delivered by the cesarean section was 13% (6/47) in a review article.[6] The failure of vaginal delivery was concluded by Benedetti, as a risk factor to cause ICH and unrelated to subsequent delivery method.[9] A retrospective study to screen 20 newborns with hemophilia in the 1st week of life came out three with ICH, and all of the 3 patients had extracranial signs of bleeding/trauma through the method of instrument-assisted delivery.[7]

The family history, abnormal bleeding issues, and symptoms of extracranial hemorrhage (ECH) may give hint to hemophilia, but there are various reasons for delay diagnosis. Lack of family history in one-third hemophilia patient and the mother’s unawareness as a carrier could make diagnosis a challenge. Anemia, lethargy, hypotension, and shock were common clinical presentation of both ICH and ECH, but seizure and bulging fontanel were more suggestive of ICH. [6]

The cranial USG is usually the initial image examination to screen the neonate for probable intracranial lesions. The USG

---

**Figure 1:** Coronal view along the plane of the choroid plexus of sonogram (a) showed obstructive hydrocephalus with enlarged bilateral lateral ventricle and high echogenicity over the quadrigeminal cistern and cerebellar hemispheres (arrow). Midline sagittal view of sonogram (b) showed enlarged 3rd ventricle and ill-defined hyperechoic density over the cerebellum (arrow) with upward compression of the 3rd ventricle and the 4th ventricle hard to identified.

**Figure 2:** Axial view of cranial magnetic resonance imaging. A large hematoma shadow (arrow) in the region of the left cerebellar hemisphere as mixed iso-signal intensity and part of high signal intensity on T1 fluid-attenuated inversion recovery (a) mixed iso-signal intensity with small faint low signal intensity and marginal rim low signal intensity on T2 fluid-attenuated inversion recovery (b) and diffusion-weighted imaging (c) low signal intensity on apparent diffusion coefficient (d) small focal hemorrhage in the left cerebellar hemisphere was suspected. T1 fluid-attenuated inversion recovery (a) showed high signal intensity and T2 fluid-attenuated inversion recovery (b) showed low signal intensity along the course of bilateral transverse sinuses (arrowhead), suspicion of sinus thrombosis.
had the advantages of easily available, quick, no radiation, and inexpensive and can be done at bedside. It is a useful technique in the neonatal care units. The cranial USG may visualize the posterior part hemorrhage through different planes,[10] and it is as well as computed tomography for imaging intraventricular hemorrhage or periventricular leukomalacia in preterm neonate.[11] It was not so distinct to detect posterior fossa subdural hemorrhage by USG through anterior fontanel, but there are benefits of the cranial USG through the posterolateral fontanelle.[1,12,13]

In our case, there were neither extracranial evidences of bleeding/truma nor family history of hemophilia. The delivery course was smooth through cesarean section. Fever and convulsions as the initial symptoms easily confused with other conditions and lack of awareness that the neonate has congenital hemophilia. With the suspicion of intracranial lesions, it is a dilemma to send the patient for MRI examination while unstable vital signs. Cranial USG examination provided a useful and early detection of underlying intracranial lesions with or without neurological symptoms, such as ventricle dilatation, hemorrhage, ischemic change, and structure anomaly.[14,15] The RI can be used as a tool for monitoring the intracranial pressure but may not reflect immediately. We also considered that the relative invasive CSF study could be postponed or even canceled after the cranial USG in cases with neurologic symptoms. Even though, the diagnosis of hemophilia A in this case is still challenging.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Blauwblomme T, Garnett M, Vergnaud F, Boddart N, Bourgeois M, Dirocco F, et al. The management of birth-related posterior fossa hematomas in neonates. Neurosurgery 2013;72:755-62.
2. Singleton TC, Keane M. Diagnostic and therapeutic challenges of intracranial hemorrhage in neonates with congenital hemophilia: A case report and review. Ochsner J 2012;12:249-53.
3. Looney CB, Smith JK, Merck LH, Wolfe HM, Chescheir NC, Hamer RM, et al. Intracranial hemorrhage in asymptomatic neonates: Prevalence on MR images and relationship to obstetric and neonatal risk factors. Radiology 2007;242:535-41.
4. Gupta SN, Kechli AM, Kanamallu US. Intracranial hemorrhage in term newborns: Management and outcomes. Pediatr Neurol 2009;40:1-12.
5. National Hemophilia Foundation. MASAC Guidelines for Perinatal Management of Women with Bleeding Disorders and Carriers of Hemophilia A and B. MASAC Document 192. Available from: http://www.hemophilia.org/NHFWeb/Resource/StaticPages/menu5/ menu57/masac192pdf [Last accessed on 2012 Jan 17].
6. Kulkarni R, Lusher JM. Intracranial and extracranial hemorrhages in newborns with hemophilia: A review of the literature. J Pediatr Hematol Oncol 1999;21:289-95.
7. Smith AR, Leonard N, Kainth MH. Intracranial hemorrhage in newborns with hemophilia: The role of screening radiologic studies in the first 7 days of life. J Pediatr Hematol Oncol 2008;30:81-4.
8. De Luca M, Carducci FI, Pansini V, Coletti V, Tucci FM, Cirillo M, et al. Unusual presentation of haemophilia in two paediatric patients. Blood Coagul Fibrinolysis 2013;24:645-8.
9. Benedetti TJ. Birth injury and method of delivery. N Engl J Med 1999;341:1758-9.
10. Bejar R, Coen RW, Ekpoudia I, James HE, Gluck L. Real time ultrasound diagnosis of hemorrhagic pathological conditions in the posterior fossa of preterm infants. Neurosurgery 1985;16:281-9.
11. Khan IA, Wahab S, Khan RA, Ullah E, Ali M. Neonatal intracranial ischemia and hemorrhage: Role of cranial sonography and CT scanning. J Korean Neurosurg Soc 2010;47:89-94.
12. Luna JA, Goldstein RB. Sonographic visualization of neonatal posterior fossa abnormalities through the posterolateral fontanelle. AJR Am J Roentgenol 2000;174:561-7.
13. Steggerda SJ, de Bruine FT, Smits-Wintjens VE, Walther FJ, van Wezel-Mejlger G. Ultrasound detection of posterior fossa abnormalities in full-term neonates. Early Hum Dev 2012;88:233-9.
14. Allison JW, Faddis LA, Kinder DL, Roberson PK, Glasier CM, Seibert JJ, et al. Intracranial resistive index (RI) values in normal term infants during the first day of life. Pediatr Radiol 2000;30:618-20.
15. Lowe LH, Bulas DJ. Transcranial doppler imaging in children: Sickle cell screening and beyond. Pediatr Radiol 2005;35:54-65.