AJPMHS GLOBAL RESEARCH NETWORK

AMERICAN Journal of Pediatric Medicine and Health Sciences
Volume 02, Issue 03, 2024 ISSN (E): 2993-2149

ANOMAL OR DELAYED DEVELOPMENT OF THE POSTERIOR MEMBRANOSAL REGION OF THE BRAIN: ANATOMY, ULTRASOUND DIAGNOSTICS, NATURAL COURSE AND OUTCOME OF BLAKE POCKET CYST IN THE FETAL.

Zhuraev Anvar Mamatmurodovich
1, Assistant, Department of Neurosurgery, Samarkand State Medical University

Introduction

Blake's pouch cyst (BPC) is considered a pathological lesion resulting from abnormal development of the posterior membranous area (PMA) of the fetal brain. However, although its first description dates back to 1900, there is little evidence of this anomaly in the literature: less than 10 articles have been published on the postnatal appearance of cerebral palsy in infants and adults. Although cystic posterior fossa malformations are one of the most frequently discussed topics in fetal neurology, we have found very few series addressing fetal cerebral palsy from a diagnostic or clinical perspective, the only exception being pathology studies9 and a couple of other interesting articles10. 11, one of which included the initial diagnostic determination of BOD10.

Materials and Methods

We decided to review the main documents on the normal and abnormal development of PMA in order to clarify the origin of BOD in as much detail as possible. Having done this, we aimed to determine sonographically acceptable criteria for its diagnosis and to review our experience in diagnosing fetal cerebral palsy.

Specifically, the objectives of this study were: 1) to review the normal development and pathogenesis of PMA in the fetal brain as seen in the published literature to date; 2) determine sonographic criteria for diagnosing breast cancer in the fetus; 3) review the ultrasound features, associated anomalies and outcomes of 19 cases of BOD observed in our center over the past 5 years

All articles describing the normal and/or abnormal development of PMA were reviewed, whether or not they were cited in the limited clinical literature on PMA. Specifically, MEDLINE searches for the terms “Blake's pouch,” with or without “fourth ventricle” or “4th ventricle,” with or without “roof,” and all relevant items 1 – 3, 12 – 16 reviewed. Descriptions of the steps leading to normal or abnormal development of the posterior fossa and fourth ventricle were created by collating the data presented in these articles.

Regarding our retrospective clinical and sonographic study, all 19 cases with posterior fossa anomalies and a final diagnosis of cerebral palsy were retrieved from our database. The following variables were assessed: indications for referral to expert targeted ultrasound,17 gestational age at

diagnosis, ultrasound findings, magnetic resonance imaging (MRI) findings, associated anomalies, karyotype, natural history, pregnancy and neonatal outcomes. In all cases, transvaginal 3D ultrasound examination was performed using an E8 ultrasound machine (GE Healthcare Ultrasound, Milwaukee, WI, USA) equipped with a conventional 5–9 MHz endovaginal volumetric probe or a 6–12 MHz high-frequency probe. The ultrasound criteria adopted for the diagnosis of BOD were as follows: 1) normal anatomy (including a normal-appearing fastigium) and vermis size (midsagittal section) of the fetal brain); 2) mild or moderate counterclockwise rotation of the vermis (mid-sagittal part of the fetal brain); 3) normal dimensions of the cisterna magna (midsagittal and axial parts of the fetal brain); 4) the presence of a roof of the BOD inside the cistern magna (mid-sagittal section of the fetal brain); the first three criteria were required to make a diagnosis, while the fourth was inconsistently observed. Nomograms published by Vinals and others18 were used to estimate vermia size. Follow-up scans were scheduled at 3-week intervals up to 35 weeks. Fifteen cases also underwent prenatal MRI at diagnosis or during follow-up. Postpartum transfontanelle sonography or MRI was performed to confirm the diagnosis of BOD in cases of full-term pregnancies. In cases of TOP, only the normality of the size and rotation of the worm can be confirmed, since the fluid collection often disappears once the posterior fossa is dissected at dissection. If the Blake's pouch does not perforate and form the median opening of the foramen of Magendie, it enlarges and develops into a cyst-like structure (CLS) protruding into the cisterna magna and acting as a wedge below the developing cerebellar vermis, which is located cephalad to the unruptured Blake's pouch. As a result, the worm rises and passively rotates counterclockwise due to the increase in volume of the unruptured Blake's pouch, which has meanwhile developed into a cyst (BPC). SI with surface imaging was able to demonstrate extension of the outer wall of the BPC into the cisterna magna (Fig. 3) in only three cases, in most cases due to shadowing from the occipital bone. In most cases, it was noted that the fluid contents of the cisterna magna were hypoechoic, with tiny strands and were not completely transparent, as in a Blake's cyst

Results

On ultrasound observation, the BPC disappeared and the worm returned to its normal position in six of 10 cases where gestational age was reached (excluding Case 1, delivery at 28 weeks due to placental abruption), likely due to late fenestration. In five of these cases observed at our center, the event occurred between 24 and 26 weeks of gestation; in the remaining case, we were only able to collect postnatal information (MRI) after the initial diagnosis at 23 weeks. In case 6, the newborn (dichorionic twin) died shortly after birth; at the time of death, the BOD was still in place according to transfontanelle ultrasound. In three other cases (cases 4, 10, and 15), moderate counterclockwise rotation of the worm persisted until neonatal observation.

MRI was performed in 15 of 19 cases at diagnosis or during follow-up, but in no case did it add anything to the ultrasound diagnosis; in the five cases where it was performed at follow-up, after sonographic confirmation of late fenestration of the PPC, it confirmed the normal position and anatomy of the vermis and the absence of cysts in the posterior fossa.

Associated major anomalies were present in eight (42%) cases, five of which were or included congenital heart disease (Table 1). The karyotype was normal in 12 cases (five by amniocentesis and seven postpartum observations) and abnormal in two (both trisomy 21); it was not assessed in the remaining five cases, four of whom underwent TOR and one died neonatally.

Regarding pregnancy outcome (Table 1), there were eight (42%) TOPs, two (10%) neonatal deaths, and the remaining nine (48%) survivors.

Conclusion

In conclusion, we described the pathogenesis of BOD by comparing data from various embryological studies 1, 2, 12 – 16 and pediatric neuroradiological 3 – 7 studies. In addition, we proposed criteria that can be used to diagnose BOD in the fetus and also showed that the cyst may undergo delayed fenestration at 24–26 weeks of gestation. Finally, we reported a clearly increased risk of associated anomalies, including congenital heart disease and trisomy 21. Further fetal studies are needed to confirm the concepts expressed in this study. However, it should be considered that, to our knowledge, this is the first analysis of fetal BOD, including diagnosis, natural history, associations, and outcome.

References

1. Blake J.A. Roof and lateral recesses of the fourth ventricle, examined morphologically and embryologically. Jay Comp Neurol1900;10:79–108.
2. Wilson J.T. On the nature and method of occurrence of a hole Magendie.J Anat1936–1937;71:423–428.
3. Tortori-Donati P, Fondelli MP, Rossi A, Carini S. Cystic malformations of the posterior fossa arising from a defect in the posterior membranous region. Mega cisterna magna and the persisting bursa of Blake: two separate entities. Childs' nervous system 1996; 12:303–308.
4. Kollias S.S., Ball W.S. Jr., Prenger EC. Cystic malformations of the posterior fossa: differential diagnosis, clarified using embryological analysis. Radiography 1993; 13: 1211–1231.
5. Strand R.D., Barnes P.D., Poussin T.Y., Estroff J.A., Burrows P.E. Cystic retrocerebellar malformations: association of the Dandy-Walker complex and Blake's pouch cyst. Pediatr Radiol 1993;23: 258–260.
6. Cornips E.M.J., Overliet G.M., Weber J.W., Postma A.A., Hoberigs K.M., Baldewijns M.L.L., Vles J.S. Clinical spectrum of Blake's pouch cyst: a report of six illustrative cases. Childs Nervous System 2010;26:1057–1064.
7. Calabro F., Arcuri T., Jinkins J.R. Blake's pouch cyst: an entity in the Dandy-Walker continuum. Neuroradiology 2000; 42:290–295.
8. Nelson M.D. Jr., Maher K., Gilles F.H. A different approach to posterior fossa cysts. Children's Radiol 2004;34: 720–732.
9. Siebert Jr. A pathologic approach to posterior fossa anomalies. Birth Defects Res A Clin Mol Teratol 2006;76:674–684.
10. Malinger G., Lev D., Lerman-Sagi T. Fetal cerebellum. Errors in diagnosis and treatment. Prenat Diagnostics 2009;29: 372–380.
11. Robinson A.J., Goldstein R. Septum cistern magna: vestigial remains of Blake's pouch and a potential new one marker of normal development of the rhombencephalon. Jay Ultrasound Med2007;26:83–95.

12. O'rahilly R., Muller F. Embryology and human teratology.

Wiley-Liss: New York, 2001.

13. Ki EAH, Retzius G. Study of the anatomy of the nervous systems and

link.. Norstedt and Soner: Stockholm, 1875.

14. Lemire RJ, Loser JD, Leach RW, Alvord Jr EC.

Normal and abnormal development of the human nervous system

. Harper and Row: Hagerstown, 1975.

15. Alexander L. Anatomy der seitentaschen der vierten

hirnkammer.Z Anat 1931;95: 531–707.

16. Brocklehurst G. Development of cerebrospinal fluid pathways

man with special emphasis on the roof of the fourth ventricle.

J Anat1969;105:467–475.

17. ISUOG. Sonographic examination of the central nervous system of the fetus.

system: recommendations for conducting a “basic examination” and “fetal neurosonogram”. Ultrasound Obstetrician Gynecol 2007; 29: 109–116.

18. Vinals F, Muñoz M, Naveas R, Schalper J, Giuliano A. Fetal cerebellar vermis: anatomy and biometric assessment using volumetric contrast imaging in the C-plane (VCI-C). Ultrasound Obstetrician Gynecol2005; 26:622–627.

19. Patel S, Barkovich AJ. Analysis and classification of cerebellar malformations. Am J Neuroradiol 2002;23: 1074–1087.

20. Forzano F, Mansour S, Ierullo F, Homfray T, Thilaganathan B. Posterior fossa malformations in the fetus: a report of 56 additional cases and a review of the literature. Prenatal Diagnostics 2007;27:495–501.

21. Klein O., Pierre-Kan A., Boddart N., Parisot D., Brunel F. Dandy-Walker malformation: prenatal diagnosis and prognosis. Childs Nervous System2003;19:484–489.

22. Paladini D, Volpe P. Posterior fossa and vermic morphometry in the characterization of fetal cerebellar abnormalities: a prospective three-dimensional ultrasound study. Ultrasound Obstetric Gynecol 2006;27:482–489.

23. SIEOG Linee 2010 Manual. Editeam: Cento (Fe), 2010.