Journal of Medical Ultrasound (2017) xx, 1—4

Chinese Taipei Society of Ultrasound in Medicine

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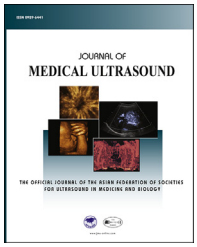
journal homepage: www.jmu-online.com



CASE REPORT

Rare Brain Tumor in a Neonate

Shih-Huan Chen [1*], Che-Sheng Ho [1,2], Nan-Chang Chiu [1,2], Hui-Ju Chen [1], Yi-Jie Lin [1]

[1] *Department of Pediatric Neurology, MacKay Children's Hospital, Taipei, Taiwan, and* [2] *Department of Medicine, MacKay Medical College, Taiwan*

Received 20 June 2017; accepted 20 September 2017
Available online ■ ■ ■

KEYWORDS
Ultrasonography,
Brain tumors,
Neonate,
ETMR

Abstract Neonatal brain tumor is rare and its outcome is generally poor. We reported a 17-day-old neonate presented as enlarged head girth. The pathological finding showed an embryonal tumor with multilayered rosettes.

Introduction

Brain tumors presented during neonatal period are rare that represent only 0.5—1.9% of all pediatric central neural system tumors [1]. Clinical presentation in the neonatal period, as the period of first 4 weeks after delivery, is not specific and the focal neurologic sign is subtle.

The availability of noninvasive imaging such as Transcranial Doppler ultrasonography, computer tomography (CT), magnetic resonance imaging (MRI) during the fetal and neonatal periods makes earlier diagnosis of these tumors possible. Transcranial Doppler ultrasonography may be the first modality to evaluate neonates clinically presenting with increasing head circumference, bulging fontanelle, lethargy or seizure.

Embryonal tumor with multilayered rosettes (ETMR), belongs to a new classification of embryonal tumors of the central nervous system according to the World Health Organization (WHO).

We present a case of ETMR and describe the clinical, pathological, and cranial ultrasonographic features of this rare tumor.

Case report

A 17-day-old male neonate presented with rapidly increased head girth. Head circumference was 39.5 cm (birth head circumference was 36 cm). He was born as a full-term to primigravida mother by lower segment cesarean section with Apgar scores were 9 at 1 min and 10 at 5 min. Birth weight was 3.48 kg. Maternal history and prenatal examination including chromosomal study of cultured amniocytes were normal. Prenatal sonographies were unremarkable until 37 weeks of gestational age. Physical examination revealed frontal bossing with bulging anterior fontanel.

Conflicts of interest: There are no conflicts of interest.

* Correspondence to: Shih-Huan Chen, Department of Pediatric Neurology, MacKay Children's Hospital, No. 92, Sec. 2, Chung-Shan N. Road, Taipei city, 10449, Taiwan.
E-mail address: i2a2n@hotmail.com (S.-H. Chen).

https://doi.org/10.1016/j.jmu.2017.09.004

Please cite this article in press as: Chen S-H, et al., Rare Brain Tumor in a Neonate, Journal of Medical Ultrasound (2017), https://doi.org/10.1016/j.jmu.2017.09.004

Transcranial Doppler ultrasonography (PA6-8-D transducer,4—10 MHz, GE Voluson ultrasound machines) showed severe lateral ventricle dilatation, third ventricle dilatation, heteroechogenicity density over whole posterior fossa with displaced 4th ventricle and visible aqueduct (Fig. 1). The lateral ventricular ratio, between lateral ventricular width and hemispheric width [2], was 62.6% of left ventricle and 56.9% of right ventricle. The Doppler color-flow imaging revealed high resistance index = 0.85 and pulsatility index = 1.70 of anterior cerebral artery. Obstructive hydrocephalus and a suspected posterior fossa space occupying lesion were impressed. MRI confirmed a $4.5 \times 4.9 \times 5.7\ cm^3$ tumor in cerebellar vermis with lateral and anterior extension as well as inferior extension to the foramen magnum. Compression on pons, medulla and 4th ventricle and evidence of obstructive hydrocephalus were seen (Fig. 2).

A suboccipital craniotomy for tumor resection was performed. Histological analysis of the resected specimen showed a hypercellular tumor which contains small cells with round to oval nuclei and scanty cytoplasm. Formation of multilayered rosettes, composed of pseudostratified tumor cells is seen. The primitive small cell areas are immunoreactive for vimentin, while the neuropil-like areas showing expression for synaptophysin. The Ki-67 labeling index is high in the cellular areas. The histopathologic features are compatible with an ETMR.

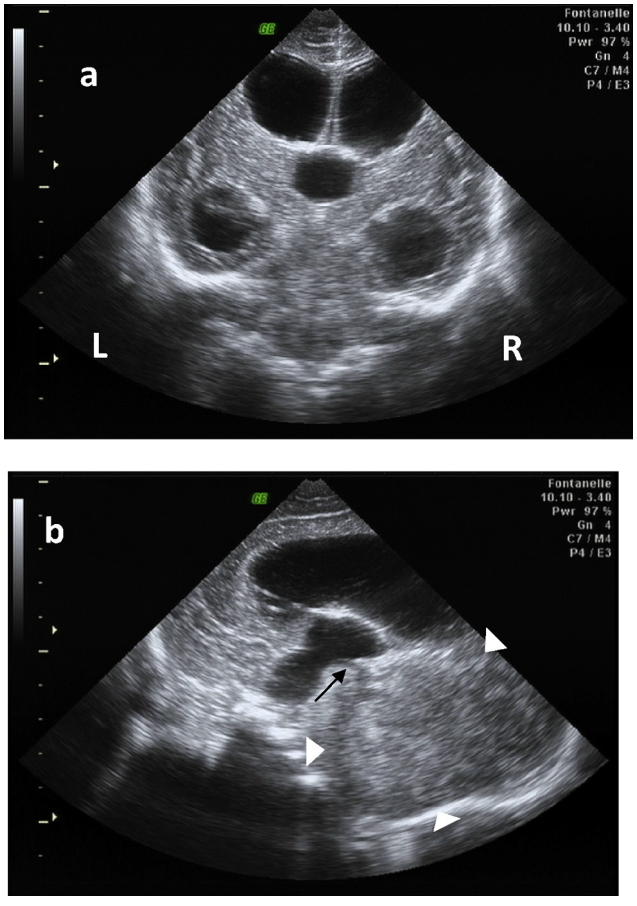


Figure 1 Sonographic image (a) in coronal view demonstrates with bilateral lateral ventricle and third ventricle dilatation. (b) Sagittal midline view shows hydrocephalus with 3rd ventricle dilatation, visible aqueduct (arrow), displaced 4th ventricle, unclear margin of vermis and cistern magna that indicated obstruction of lower CSF pathway. Space-occupying lesion (arrowhead) with heteroechogenicity in vermis is suspected.

Discussion

The most common brain tumors diagnosed in the fetus/neonate were teratomas (29%), followed by astrocytomas (18%). Primitive neuroectodermal tumor (PNET) makes up 13% of all fetal and neonatal brain tumor [3,4]. ETMR constitutes a distinct entity of the PNET family. ETMR was proposed in 2010 as a unifying entity by Paulus and Kleihues, including embryonal tumor with abundant neuropil and true rosettes (ETANTR), and ependymoblastoma [5]. This tumor presents a focal amplification of the C19MC region encoding a miRNA cluster of chromosome 19 (19q.13.42). In 2016 WHO classification of embryonal brain tumors [6], the term of PNET was removed. In the presence of C19MC amplification, a tumor with histological features conforming to ETANTR/ETMR should be diagnosed ETMR, C19MC-altered. In contrast, in the absence of C19MC amplification, ETMR, not otherwise specified should be diagnosed. Chromosome 19 expression studies were not available in our case.

ETMR are rare entities and underdiagnosed in the result of unclassified embryonal tumors and only a few cases reported in the past literature. They occur mostly in children aged less than 2 years old and their prognosis is poor. In the literature, the localization of the majority of ETMR is supratentorial with signs of increased intracranial pressure (66%) [7]. Our case showed a infratentorial tumor with hydrocephalus.

Most embryonal brain tumors in children grow rapidly and spread into other area of the brain. Depending on the type, size, and location of the tumor, obstruction of cerebrospinal fluid pathways can be seen. In the literature, infratentorial tumors were commonly found to have hydrocephalus than supratentorial brain tumors [8,9].

Our case manifested as rapid growth infratentorial tumor leading to hydrocephalus in neonatal period. Neonatal brain tumors are rare and sometimes detected incidentally on routine prenatal or neonatal ultrasonography. These tumors may present with an increasing head circumference, bulging fontanel, vomiting, seizures, sunset eyes and also only with subtle symptoms such as irritability, poor feeding, failure to thrive.

Transcranial Doppler ultrasonography is a quick, convenient and also not expensive tool to evaluate cerebral structure via an open fontanel in neonates who presenting nonspecific symptoms. It can evaluate hydrocephalus in the neonatal patient and can be used to quantitate the progression of hydrocephalus, which can help for decision about neurosurgical intervention. The sonographic feature of brain tumor is a hyperechoic or hypoechoic mass with well-defined or unrecognized border. Intracranial teratoma appears as heterogeneous echogenicity with calcification and cystic lesion. Ependymoma presents as a large cystic mass. Choroid plexus papilloma is hyperechoic on ultrasonography and located in the lateral ventricles [10,11].

In other case reports, sonographic pattern of ETMR was not described in detail. The sonographic feature of this

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