

Huge tumor of the intracranial cavity: a catastrophic imaging on USG and MRI

Published online 20 December, 2005 © <http://www.neuroanatomy.org>

Ibrahim M. ZİYAL ⁺
 Burcak BİLGİNER
 Gökhan BOZKURT

Hacettepe University, Faculty of Medicine, Department of Neurosurgery,
 Ankara–Turkey.



⁺ Ibrahim M. Ziyal, MD
 Associate Professor of Neurosurgery,
 Department of Neurosurgery, Hacettepe University,
 Faculty of Medicine, 06100 Ankara–TURKEY
 ☎ 90-312-305 28 27
 📠 90-312-311 11 31
 ✉ ibrahimziyal@yahoo.com

Received 27 October 2005; accepted 19 December 2005

ABSTRACT

In this case, we report a tumor occupying 76.19% of the intracranial cavity which is diagnosed with magnetic resonance imaging. This tumor is inspected as one of the biggest intracranial mass lesions in literature. In such cases, the postoperative survival rate is low due to several factors such as perioperative bleeding, sudden volume changes in the intracranial cavity and intracranial hypotension. The surgical excision is open to question. *Neuroanatomy; 2005; 4: 55–56.*

Key words [huge] [tumor] [intracranial cavity] [USG] [MRI]

Introduction

Fetal intracranial tumors are often associated with hydrocephalus, polyhydramnios and macrocephaly. The most common location is supratentorial compartment [1–3]. Focal neurological changes are absent in most cases of neonatal brain tumors despite the large head size and the hydrocephalus. These tumors do not interfere severely with normal gestation and parturition. Several histologic types of congenital intracranial tumors have been described, including teratoma, choroid plexus papilloma, craniopharyngioma, meningeal sarcoma, lipoma of the corpus callosum and oligodendroglioma [1–4]. Here, a tumor occupying 76.19% of the intracranial cavity is presented. Our literature review revealed no intracranial tumor with such huge volume ratio.

Case Report

A two month old girl was born to a 35 years old gravida 5, para 3, aborta 2 mother at 38th week gestation via cesarean section. Prenatal ultrasonography at 35th week revealed an enlarged head. An intracranial tumorlike mass without obviously normal intracranial structures was evident on ultrasonography. Because of severe macrocephaly, an elective cesarean section was performed at 38th week of gestation. The infant's Apgar score was low. Her weight was 3045 g, length was 45 cm and head circumference was 47 cm at birth. She was admitted to our hospital because of increase in her head circumference and unconsciousness. Physical examination revealed a huge head with wide bulging fontanelles and a bossing forehead.

She was extremely hypotonic with widely separated cranial sutures. No other congenital anomaly was observed. Transcranial ultrasonographic examination revealed a heterogeneous intracranial mass without normal brain structure (Fig. 1). Magnetic resonance imaging demonstrated a huge intracranial mass lesion occupying 76.19% of the intracranial cavity (Fig. 2). A subtotal tumor removal was performed. During surgery it was observed that the tumor was excessively vascular. Short after the operation the patient was deceased.

Discussion

Congenital brain tumors are rare entities which are nowadays often recognised during pregnancy by ultrasound and magnetic resonance imaging [5]. In literature, first report of a massive congenital intracranial teratoma was published in 1864 by Breslau and Rindfleisch [6]. Giant pediatric tumors may present with seizures due to irritation of the cortical gray matter. In more rapidly growing tumors, signs of increased intracranial pressure such as papilledema may occur. Bulging fontanelles and macrocephaly may be evident in infants [7].

Nerve and glial cells are derived from a specialized portion of the ectoderm termed the neural plate. The edges of the neural plate fold, ultimately appose, and form the neural tube. The cerebral hemispheres and the brain stem develop from the rostral and intermediate portions of the neural tube, and the spinal cord develops from the caudal portions. The neuroepithelium lines the

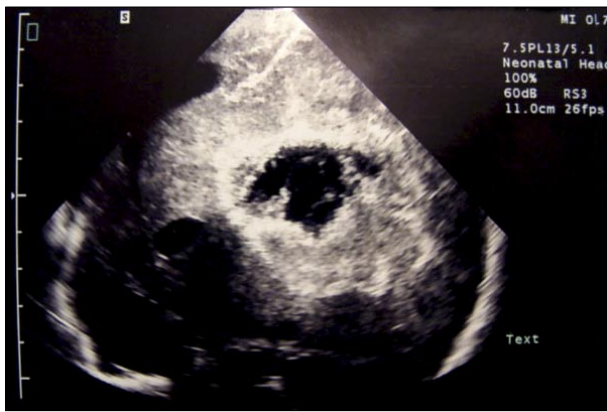


Figure 1. A huge intracranial mass with heterogeneous echogenicity and multilobulated cysts associated with obstructive hydrocephalus is observed in ultrasonography of a 2 months old girl.

neural tube and forms the cellular constituents of the central nervous system. Immature neurons arise from the neuroepithelium. The neural tube has two additional layers: the mantle layer, which becomes the gray matter of the central nervous system, and the marginal layer, which becomes the white matter. The cavity within

the neural tube forms the ventricular system [8]. These tumors are most commonly localized to supratentorial region and generally arise from the cortical gray-white matter or from the ventricles. In this case the congenital tumor prohibits development of normal neural structures. Instead of gray and white matter, corpus callosum, internal-external capsule, thalamus, nucleus caudatus and basal ganglia, there was a huge tumor distorting and compressing the brain.

As far as we know, this case was the biggest intracranial tumor in literature, regarding the ratio of the tumor volume to the intracranial cavity. Such cases may be called “Pandora’s box”. The prognosis is usually fatal in these cases because of rapid, invasive growth of the tumors and the destruction of regular cerebral structures [1]. Surgical excision may be curative at smaller benign intracranial lesions. But in such cases like ours, if the intracranial cavity is opened, control of bleeding is extremely difficult because of large surface of bleeding area in the tumor. On the other hand, sudden intracranial hypotension after removing the tumor may also be fatal. Because of poor prognosis we have to think twice before operating such cases.

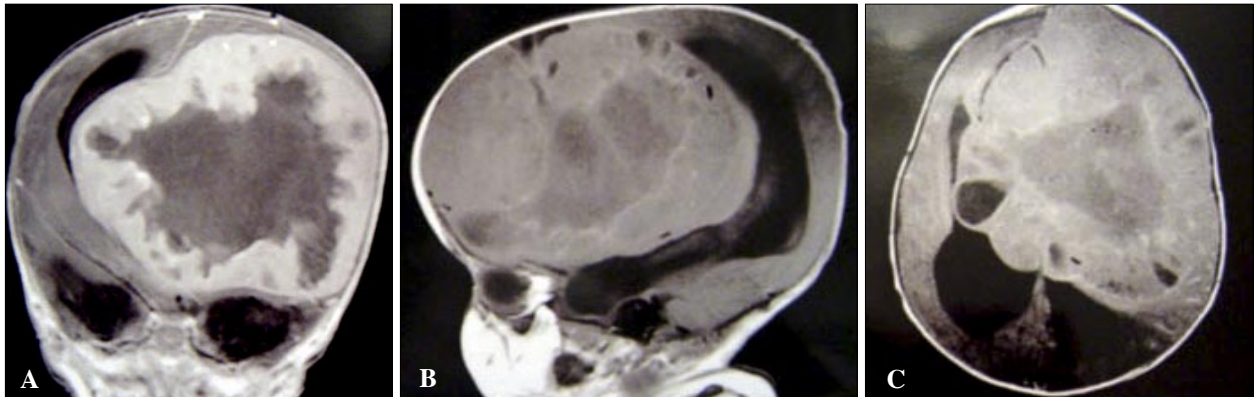


Figure 2. Post-contrast T1-weighted coronal (A), sagittal (B) and axial (C) magnetic resonance images showing the extension of huge intracranial mass occupying 76.19% of the intracranial cavity and the distortion and compression of neural structures.

References

- [1] Horton D, Pilling DW. Early antenatal ultrasound diagnosis of fetal intracranial teratoma. *Br. J. Radiol.* 1997; 70: 1299–1301.
- [2] Palo P, Penttinen M, Kalimo H. Early ultrasound diagnosis of fetal intracranial tumors. *J. Clin. Ultrasound.* 1994; 22: 447–450.
- [3] Chien YH, Tsao PN, Lee WT, Peng SF, Yau KI. Congenital intracranial teratoma. *Pediatr. Neurol.* 2000; 22: 72–74.
- [4] Wakai S, Arai T, Nagai M. Congenital brain tumors. *Surg. Neurol.* 1984; 21: 597–609.
- [5] Mazouni C, Porcu-Buisson G, Girard N, Sakr R, Figarella-Ballanger D, Guidicelli B, Bonnier P, Camerme M. Intrauterine brain teratoma: a case report of imaging (US, MRI) with neuropathologic correlations. *Prenat. Diagn.* 2003; 23: 104–107.
- [6] Breslau, Rindfleisch E. Geburtsgeschichte und Untersuchung eines Falles von Foetus in Foetu. *Virchows Arch. Pathol. Anat.* 1884; 30: 406–417.
- [7] Winn HR, Youmans JR. *Youmans neurological surgery.* 5th Ed., Philadelphia, Saunders. 2004; 3697.
- [8] Martin JH. *Neuroanatomy text and atlas.* 2nd Ed., New York, McGraw-Hill. 1996; 57.