

Rapid Growth of Congenital Diffuse Brain Tumor Considered to Be Teratoma

—Case Report—

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Abstract

Prenatal ultrasonography of a 17-year-old pregnant female detected ventriculomegaly of the fetus at 31 weeks of gestation. Her medical and family histories were unremarkable. Fetal magnetic resonance imaging taken at 33 weeks of gestation showed a tumorous lesion with ventriculomegaly. A male baby was delivered by cesarean section at 36 weeks of gestation. The Apgar scores were 9 and 9 at 1 and 5 minutes after the delivery, respectively. The head circumference at birth was 41.5 cm with bulging anterior fontanel, but no other congenital anomaly. He showed relatively good activity with satisfactory feeding. Computed tomography performed on postnatal day 5 revealed a massive brain tumor of mixed density, with multiple lobulation and cystic and calcified components. The tumor had rapidly grown with diffuse appearance. The patient underwent endoscopic biopsy with installation of an Ommaya reservoir to control the hydrocephalus on postnatal day 6. The tumor appeared hypervascular and bled profusely on resection maneuver, so the endoscopic procedure for histological verification was abandoned. Cerebrospinal fluid taken intraoperatively revealed marked elevation of the alpha-fetoprotein level and mild increase of the human chorionic gonadotropin level, strongly suggestive of teratoma. Neuroimaging performed on postnatal day 11 indicated significant additional tumor growth which occupied nearly the whole cranial cavity. His activity began to deteriorate on postnatal day 13 and he died of respiratory distress on the 15th day of life.

Key words: congenital brain tumor, intracranial teratoma, endoscopic biopsy

Introduction

Congenital central nervous system tumors are uncommon, accounting for 0.5–1.5% of all childhood brain tumors,³⁾ but are nowadays often recognized during pregnancy by ultrasonography or magnetic resonance (MR) imaging.⁶⁾ The most common histological type is teratoma, present in one-third to one-half of all cases, followed by medulloblastoma, astrocytoma, choroid plexus papilloma, and ependymoma.^{3,4,8,9,11–14,16–20)} Congenital brain tumors show significant differences in etiology, histological type, topographic distribution, clinical presentation, and prognosis from tumors originating after the first year of life.¹⁹⁾ Massive congenital intracranial teratoma is a rare neoplasm with poor prognosis because the tumor tissue has already replaced most of the brain tissue before first identification on fetal

ultrasonography.^{1–5,7,10,13,15,19)} Surgical treatment is seldom undertaken because of the high frequency of fatal and postnatal death. The surgical treatment of a large intracranial tumor with partial destruction and replacement of brain structure also represents an ethical dilemma.¹⁵⁾ The definitive diagnosis is usually achieved by histological evaluation, but the diagnosis may remain uncertain because of the malformative origin of this type of tumor. The histological diagnosis is only achieved at autopsy in many cases following the failure of endoscopic biopsy.^{2,5,6,19)}

Here we present a case of massive congenital brain tumor showing rapid growth which was most probably teratoma but endoscopic biopsy failed to provide definitive identification.

Case Report

A male neonate was the first child of a 17-year-old mother, gravida I, para 0, whose medical and family histories were unremarkable. Regular prenatal ex-

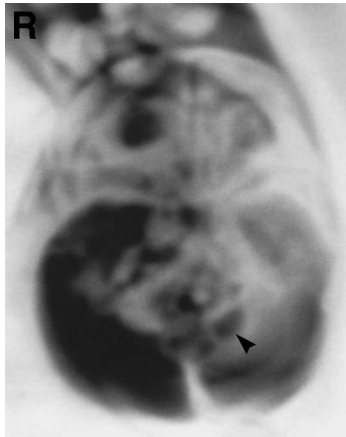


Fig. 1 Fetal T₁-weighted magnetic resonance image taken at 33 weeks of gestation showing a multilobulated mass adjacent to the choroid plexus of the left lateral ventricle (arrowhead) with ventriculomegaly.

aminations were uneventful until prenatal ultrasonography performed at 31 weeks of gestation revealed fetal hydrocephalus. She was referred to the Department of Obstetrics of our university hospital. Fetal MR imaging taken at 33 weeks of gestation showed a multilobulated mass adjacent to the choroid plexus of the left lateral ventricle with ventriculomegaly (Fig. 1). After consultation with the Department of Neurosurgery, a cesarean section was performed at 36 weeks of gestation to allow treatment of the newborn. The Apgar scores were 9 and 9 at 1 and 5 minutes after the delivery, respectively. The birth weight was 2536 g, height 51.5 cm, and head circumference 41.5 cm (>95th percentile).

Physical examination revealed bulging anterior fontanel 5 × 6 cm in diameter and bossing of the forehead. No other congenital anomaly was observed and neurological examination revealed no abnormality. Cardiopulmonary dysfunction was not found and he showed satisfactory feeding. Cranial computed tomography (CT) performed on postnatal day 5 revealed a multilobulated diffuse brain tumor of mixed density, including cystic and calcified components, and heterogeneously enhanced by contrast agent (Fig. 2). The tumor had rapidly grown with diffuse appearance during the 4 weeks between neuroimaging examinations. Normal brain structures were hardly recognizable including the brainstem.

The parents were informed of the poor prognosis and the significant risks associated with a radical resection, and following adequate discussion and consent to the treatment, selected endoscopic biopsy

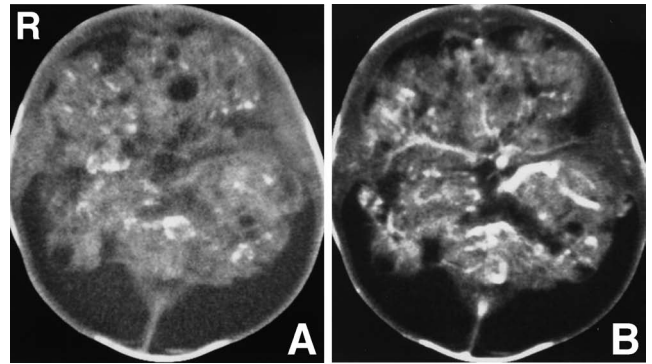


Fig. 2 Computed tomography scans performed on postnatal day 5 demonstrating a diffuse brain tumor with multiple lobulation, including cystic and partially calcified components (A), and heterogeneously enhanced by contrast agent (B).

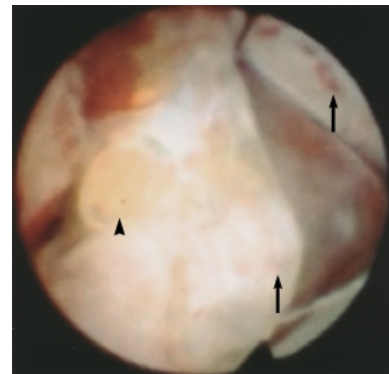


Fig. 3 Endoscopic view showing the grayish surface of the tumor, with multilobulated appearance, cyst formation (arrowhead), and hypervascularity (arrows).

for histological verification, with installation of an Ommaya reservoir to control the hydrocephalus. Surgery was performed on postnatal day 6. A burr hole was made 3 cm superior and 3 cm lateral to theinion, for safe entry into the cranial cavity and adequate distance between the inner table of the skull and the surface of the tumor. Endoscopy revealed the grayish, multilobulated, cystic hypervascular tumor (Fig. 3). The tumor bled profusely on resection maneuver, so the endoscopic surgery for histological verification was considered too dangerous to proceed.

Examination of cerebrospinal fluid taken intraoperatively revealed marked elevation of the alpha-fetoprotein level (49 300 ng/ml) with mild increase of the human chorionic gonadotropin level (9.2

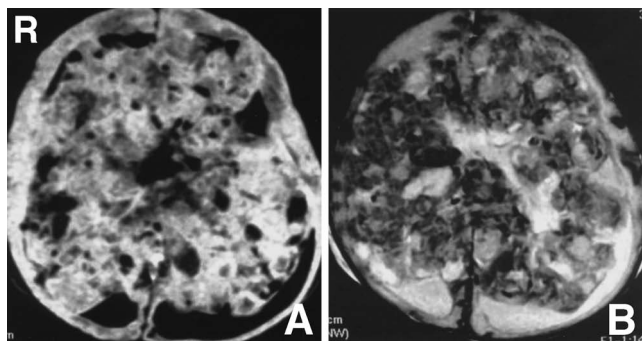


Fig. 4 Axial magnetic resonance images taken on postnatal day 11 showing the tumor as heterogenous, but mostly hyperintense on the T₁-weighted (A) and heterogeneously intense on the T₂-weighted (B) image, demonstrating significant additional tumor growth occupying nearly the whole cranial cavity.

mIU/ml), strongly suggestive of teratoma. Surveillance CT performed on postnatal day 11 revealed significant additional tumor growth which occupied nearly the whole cranial cavity. MR imaging taken on the same day showed the tumor as heterogenous, but mostly hyperintense on T₁-weighted and heterogeneously intense on T₂-weighted imaging (Fig. 4). His activity began to deteriorate on postnatal day 13 and apnea developed. The infant died on the 15th day of life. His parents refused to permit autopsy of their son.

Discussion

In the present patient, fetal MR imaging taken at 33 weeks of gestation revealed an apparently treatable brain tumor and hydrocephalus, which necessitated cesarean section. However, the tumor rapidly grew and became enormous during the next 4 weeks with little brain tissue discernible, which forced us to abandon radical resection surgery. Most of 39 cases of congenital intracranial teratomas had a fetal head size larger than expected for the gestational age and massive destruction or compression of the brain, which is too late to begin management.¹⁰⁾ In the present case, an Ommaya reservoir was installed to control the hydrocephalus combined with endoscopic biopsy as palliative measures.

Surgical treatment for massive congenital intracranial teratoma is rare. The affected fetus or neonate has already been severely impaired by diffuse extension of the tumor before identification, which may partly explain the poor prognosis. Surgical treatment represents an ethical dilemma in a ne-

onate with diffuse intracranial tumor and little brain functioning structure.¹⁵⁾ In addition, the long-term functional and intellectual outcomes of the operated neonates are unknown. Termination of the pregnancy has been recommended if the tumor is detected before the 24th week of gestation.^{1,7)} Surgery may achieve survival and better prognosis for the patient if complete resection of the tumor can be achieved before enormous growth occurs. Future methods of intrauterine surgery may offer the chance to treat congenital intracranial teratoma before the affected fetus is severely impaired.

Endoscopic surgery is less invasive and easier to perform than open surgery, especially for histological examination of homogeneous lesions. However, this method is not appropriate for diffuse tumors containing variable histological components, which may obscure the interpretation of biopsy specimens, even if multiple samples are obtained from different areas.⁶⁾ The limited surgical view also presents significant risk if the lesion is hypervascular.

In the present case, the intracranial localization, enormous extension and size, hypervascularity, and rapid progression of the tumor all combined to form a very poor prognosis. The prenatal diagnosis of intracranial teratoma is important for timely counseling of the parents and to aid in obstetrical decision making. Fetal MR imaging is crucial for identifying the tumor localization, extent, and for optimizing plans for postnatal care.¹⁰⁾ More cases will illustrate the medicosocial, medicolegal, and ethical aspects of the management of congenital teratoma.

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