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Massive Congenital Intracranial Teratoma
-An autopsy case-

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Massive congenital intracranial teratoma (MCIT) is a very rare tumor and only one case has been reported in Korea. We report a case of MCIT discovered in a male infant of 25 weeks of gestational age. Prenatal ultrasound revealed a large heterogenous echoic mass which almost replaced the intracranial content. The infant was severely macrocephalic with a small but normal appearing body. The cranial vault was almost entirely occupied by an 18cm sized, multilobulated, partially cystic mass that displaced the severely distorted and attenuated cerebral cortex. Microscopically, the bulk of the tumor was composed of embryonal neuroepithelium with mature glial tissue and choroid plexus. Cartilage, liver, mature squamous epithelium and differentiated mesenchymal elements such as muscle and adipose tissue were found within the tumor.

Key Words: Massive, congenital, intracranial teratoma

Teratoma is the most common congenital brain tumor (Ishikawa *et al.* 1987). Up to the fourth month of life, most teratomas are benign and composed of mature tissue (Avila and Conzalez-Crussi, 1991). Although congenital intracranial teratomas producing hydrocephalus in early life are quite common, massive teratoma replacing almost the entire intracranial content of a neonate is an exceptionally rare event(Odell *et al.* 1987). Only one case has been reported in the Korea (Kim *et al.* 1990). The origin of the extragonadal teratoma may be a misplaced conjoined (Jensen, 1974; Rostad *et al.* 1985; Odell *et al.* 1987; Saiga *et al.* 1991). It is thought that intracranial teratomas usually originate in the vicinity of the pineal gland, quadrigeminal plate, or the walls of the third ventricle, in a median or closely paramedian location. In massive cases, the precise site of origin can not be determined (Odell *et al.* 1987). We report a case of massive intracranial immature teratoma discovered in a male infant of 25 weeks of gestational age.

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CASE REPORT

A 1,614 gm male infant was delivered by inverted T hysterectomy at 25 weeks of gestation to a 30-year-old G_2 P_1 L_1 D_0 A_0 mother. The family history was unremarkable. Prenatal ultrasound revealed a large heterogenous echoic mass which almost totally replaced the intracranial content (Fig. 1). Therefore, termination was done. The infant was severely macrocephalic with a small but normal appearing body. The head circumference was 32.5 cm with grossly split sutures and a soft, boggy, almost fluctuant head. The fontanelles were widely separated and covered by membranous, nonossified tissue (Fig. 2). Magnetic resonance imaging of the brain was taken. A huge, lobulated mass was observed. The mass

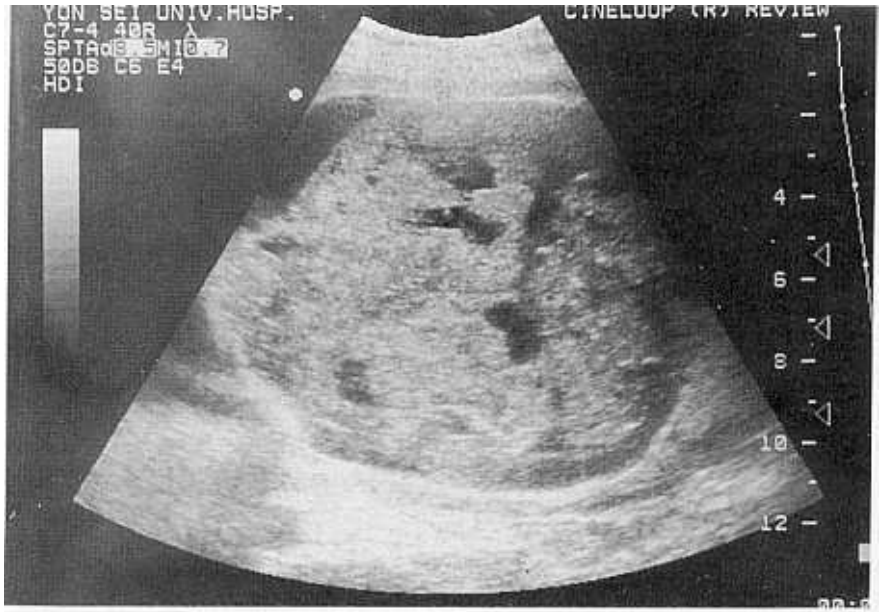

Fig. 1. *Prenatal ultrasound reveals enlarged calvarium with a large heterogenous echoic mass near totally replacing the intracranial content.*

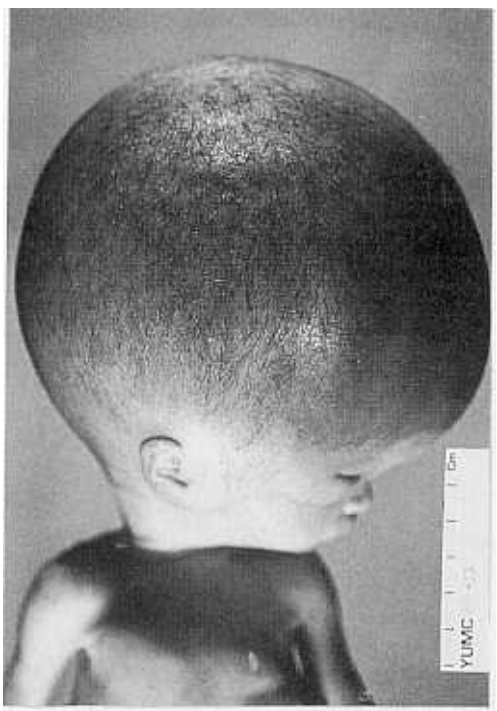

Fig. 2. *The infant is severely macrocephalic with a small but normal appearing body.*

showed homogenous, intermediate signal intensity in T1 WI and heterogenous hyperintensity in T2 WI. Calcified spots were present within the mass (Fig. 3). The cranial vault was almost entirely occupied by an 18cm sized, multilobulated, partially cystic mass that displaced the severely distorted and attenuated cerebral cortex. The solid portions of the tumor were grayish white and firm and contained punctate calcification. The cystic portions contained mucoid materials (Fig. 4). The brain was so markedly effaced that the anatomic orientation of the central nervous system was unrecognizable. Microscopically, the bulk of the tumor was composed of embryonal neuroepithelium with mature glial tissue and choroid plexus (Fig. 5). Cartilage, liver with extramedullary hemopoiesis and differentiated mesenchymal elements such as muscle and adipose tissue were readily found; the cartilage revealed focal ossification. Several dilated lymphatic spaces and several nests of mature squamous epithelia were also found. Other organs were unremarkable.

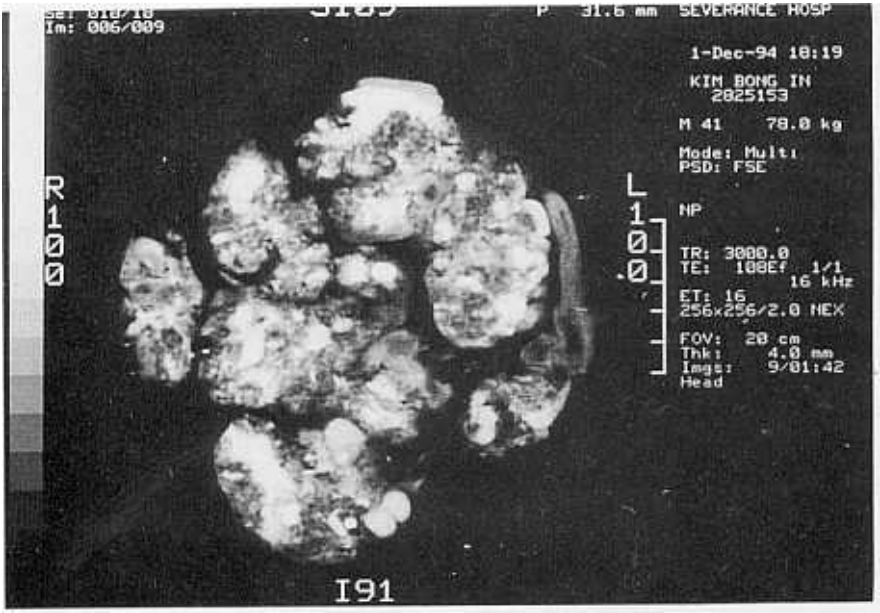

Fig. 3. *T2 WI of specimen MRI reveals a mass showing heterogenous hyperintensity with multiple cystic spaces and calcified spots. The remaining brain parenchyme is noted in the peripheral portion.*

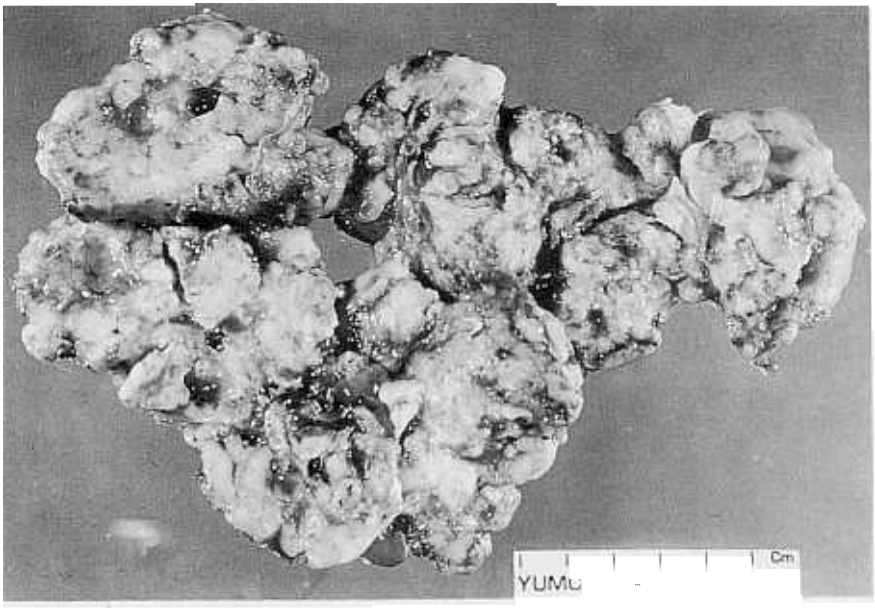

Fig. 4. *The mass removed from the brain is composed of multilobulated, grayish white, firm tissue with multiple cystic spaces.*

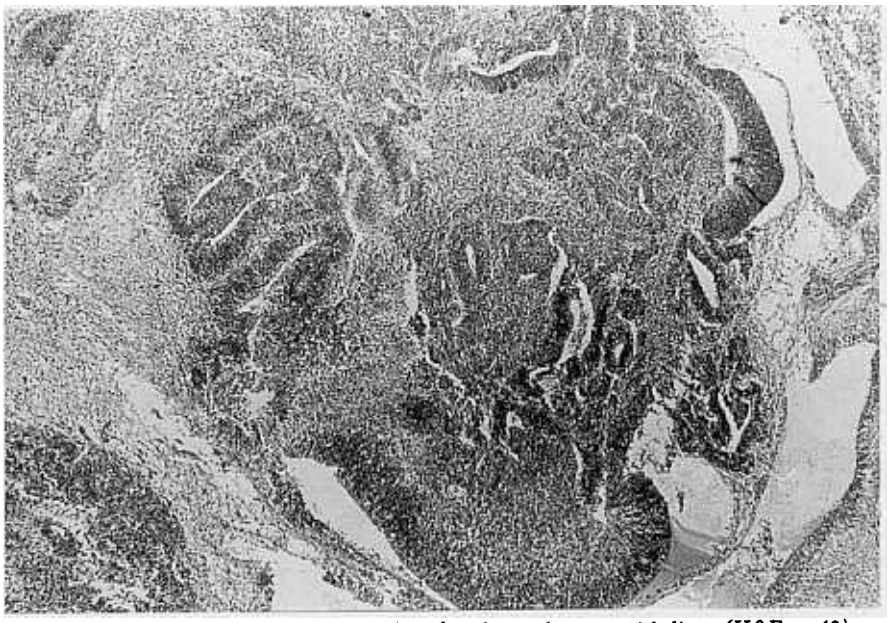

Fig. 5. The tumor is mainly composed of embryonal neuroepithelium (H&E, ×40).

DISCUSSION

A neonatal brain tumor is one which develops within 60 days of birth, and is very rare. Histologically, a teratoma is the most common tumor, with medulloblastoma ranking next. The following have also been reported: astrocytoma, choroid plexus papilloma and ependymoma or ependymoblastoma (Ishikawa *et al.* 1989). Before the fourth month of life, most teratomas are benign and composed of mature tissue (Avila and Conzalez-Crussi, 1991).

An intracranial teratoma has three characteristic clinical patterns. In the first one, cranial enlargement occurs abruptly between two days and three months after date of birth. Typically, the tumor is relatively small and produces hydrocephalus. In the second group, children die soon after birth or survive until nine weeks of age. These present with extensive replacement of brain substance by tumor. A third group presents with massive intrauterine growth. These infants are either stillborn or die soon after birth (Crussi, 1982). Although congenital intracranial teratomas producing hydrocephalus in early life are quite common, massive teratoma replacing almost the entire intracranial content of a neonate is an exceptionally rare event (Odell *et al.* 1987). Only one case has been reported in the available Korean literature (Kim *et al.* 1990). Explanations for massive growth in the fetal period are ① time of origin of the neoplastic cells is closer to the time of conception, allowing a greater number of days of growth before birth and tumor detection, ② these neoplasms which occur in utero have an unchecked growth pattern because of relatively minimal clinical systems produced in the fetus, ③ fetal period offers a better milieu for growth of the immature elements which make up these teratomas (Rostad *et al.* 1985). Many cases have polyhydramnios. Possible explanations include ① extension of the tumor into or around the oropharynx with esophageal and tracheal displacement resulting in the inability to swallow amniotic fluid, ② displacement or destruction of the brain stem or diabetes insipidus from hypothalamic destruction (Rostad

et al. 1985).

The origin of an extragonadal teratoma may be a misplaced conjoined twin pregnancy (Jensen, 1974; Rostad *et al.* 1985; Odell *et al.* 1987; Saiga *et al.* 1991). This is suggested by cytogenetic studies confirming the diploid nature of the tumor (Rostad *et al.* 1985; Odell *et al.* 1987) and observation that they frequently arise at the site of attachment of conjoined twins (Jensen , 1974). It is generally accepted that the distinction between fetus-in-fetu and teratoma is largely determined by whether an axial skeletal system is present or not (Chi *et al.* 1984). Associated factors are chromosomal abnormalities, intrauterine infection, and drug ingestion during pregnancy (Sato *et al.* 1978).

It is thought that intracranial teratomas usually originate in the vicinity of the pineal gland, quadrigeminal plate, or the walls of the third ventricle, in a median or closely paramedian location. In massive cases, the precise site of origin can not be determined (Odell *et al.* 1987). Congenital brain tumors have dominant CT appearance including a large heterogenous lesion with associated hydrocephalus, regardless of histology. However, coarse calcification is a constant feature in the teratomas (Buetow *et al.* 1990).

The prognosis of the congenital brain tumor is related to tumor histology; the prognosis for astrocytoma and choroid plexus papilloma is good, but that for other tumors is poor. Tumors that are of a size and location that render them manageable or potentially curable by surgical resection are typically associated with an increased possibility of surgical survival (Ishikawa *et al.* 1989).

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