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Diagnostic imaging of the congenital brain neoplasms

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Summary

Background:

Congenital brain tumors account for 0.5-1.5 % of all congenital neoplasms in neonates. They are most commonly found in the supratentorial region, with the ratio 4:1 to the infratentorial structures. The neoplasms consist of a variety of histologic types, with predominance of teratoma and rarely astrocytoma or choroid plexus papilloma. Because of the elasticity of the infantile skull, the tumors attain huge size.

Material/Methods:

In the period 1994-2007, 41 congenital brain tumors were diagnosed at the Polish Mother's Memorial Hospital Research Institute. The patients underwent the CT and MR examinations. In 3 cases, prenatal US revealed intracranial masses.

Results:

Histologically the most of the tumors were astrocytoma structures (45%) and the second type was choroid plexus papilloma (15%). A teratoma was diagnosed only in one child (0.24%). In 32 (77%) patients a huge, heterogeneous mass in the supratentorial area was observed at the time of the diagnostic examinations.

Conclusions:

Congenital CNS masses detected after delivery period present different histological types. As a result of this histological variety, we observe different CT and MR images. These truly congenital tumors produce symptoms at birth or within the first 2 weeks of life.

Key words:

Congenital • brain tumor • CT scan • MR image

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Background

Congenital brain tumors are detected in 1.1-3.6 neonates per 100000 live births and are fatal in 0.004-0.18% children below 1 year of age [1].

Brain tumors account for 0.5-1.5% of all congenital neoplasms in neonates [2].

In 3.4% of cases, CNS tumors are diagnosed in the prenatal period during the last trimester of pregnancy [3].

The literature presents various types of classification of congenital CNS tumors. Invald et al. propose the following types: true congenital CNS tumors diagnosed in the perinatal period, neonatal tumors detected in the first month of life and brain tumors detected between 2 and 12 months of age [4]. Another classification presented by Sobel et al.

presumes that true congenital CNS tumors are those diagnosed on delivery and in the first 2 weeks of life. Congenital brain tumors are probably the cause of clinical symptoms observed in the first year of life, and the development of those diagnosed later may be associated with the prenatal period [1].

A characteristic feature of congenital CNS tumors is the occurrence of spontaneous intracranial hemorrhages observed in prenatal examinations [2], due to the presence of anomalous vasculature developing within the tumor tissue. The tumors are most commonly localized in the supratentorial region, with the ratio 4:1 to the infratentorial structures [5].

Immature teratomas (teratoma immaturum) located centrally in the region around the sella turcica or the pineal body, account for 28.8% to 50% of tumors diagnosed in the

Table 1. Histological type of the brain tumors.

Histopathological diagnosis	Number of patients
<i>Astrocytoma</i>	19 (45%)
<i>Choroid plexus papilloma</i>	6 (14.6%)
<i>Ependymoma</i>	4 (8.1%)
<i>PNET</i>	3 (7.3%)
<i>Medulloblastoma</i>	2 (0.05%)
<i>Germinoma</i>	2 (0.05%)
<i>Dermoid Cyst</i>	1 (0.02%)
<i>Teratoma</i>	1 (0.02%)
Others	2 (0.05%)
Total	41

perinatal period [3]. Less frequent tumors include astrocytomas, choroid plexus papillomas and PNET (primitive neuroectodermal tumors) [5, 6].

Because of the elasticity of the infantile skull, congenital tumors attain huge size and cause an increase of intracranial pressure in the fetal period [7]. Increased circumference of the head may result in premature birth, or complications of delivery at term [4].

The most frequent clinical symptom presented by neonates with congenital brain tumors is a rapid increase of head circumference. Above one month of age, the tumor is manifested by symptoms associated with increased intracranial pressure, vomiting and loss of appetite.

Materials and methods

In 1994-2007, 41 neonates and infants were diagnosed with congenital brain tumors in the Polish Mother's Memorial Hospital Research Institute. Their incidence was equal for both sexes, the patient group included 21 boys (50.25%). The first diagnostic imaging modality was head CT in 13

Table 2. Localization of the brain tumors.

CNS tumor localization	Number of patients
supratentorial	32
infratentorial	9

children, MR in 10, and 18 underwent both CT and MR of the brain. Three tumors were diagnosed with USG in the prenatal period.

CT and MR imaging was biphasic, with intravenous contrast medium administration in the second phase of the scan, iodine in CT and paramagnetic in MR. The contrast medium dose was appropriate for the patient's body weight. For MR, the children required short-time sedation, in single cases we managed to take advantage of physiological sleep to perform the scan.

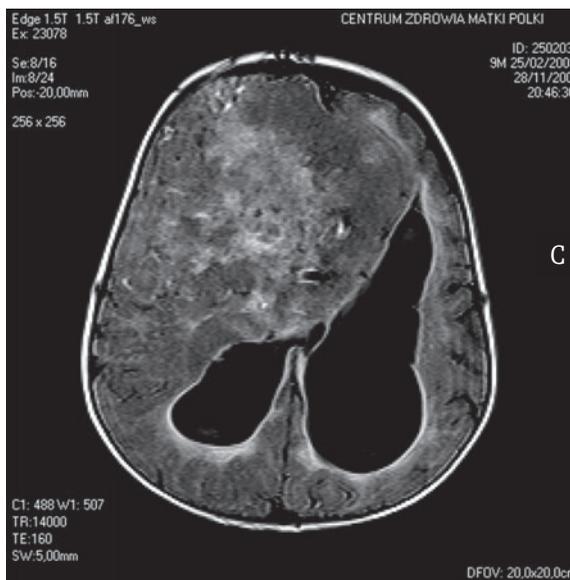
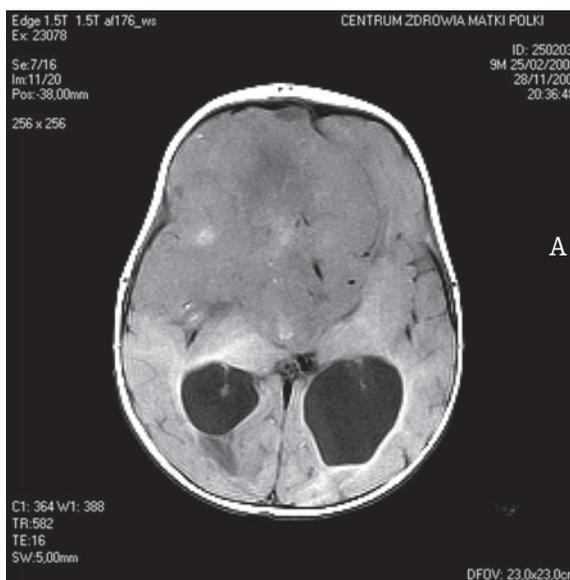
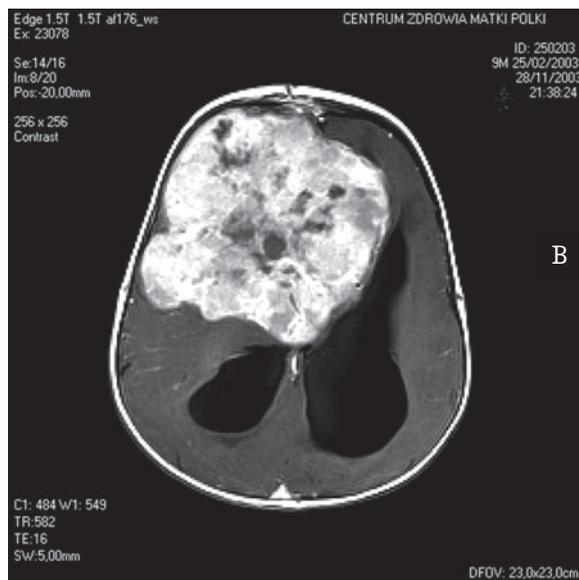


Figure 1. Axial T1-weighted noncontrast (A) and after contrast administration (B) images show huge, heterogeneous tumor (*astrocytoma*) in the anterior fossa. The periferal portion of the tumor localized in the frontal horns of the lateral ventricles.

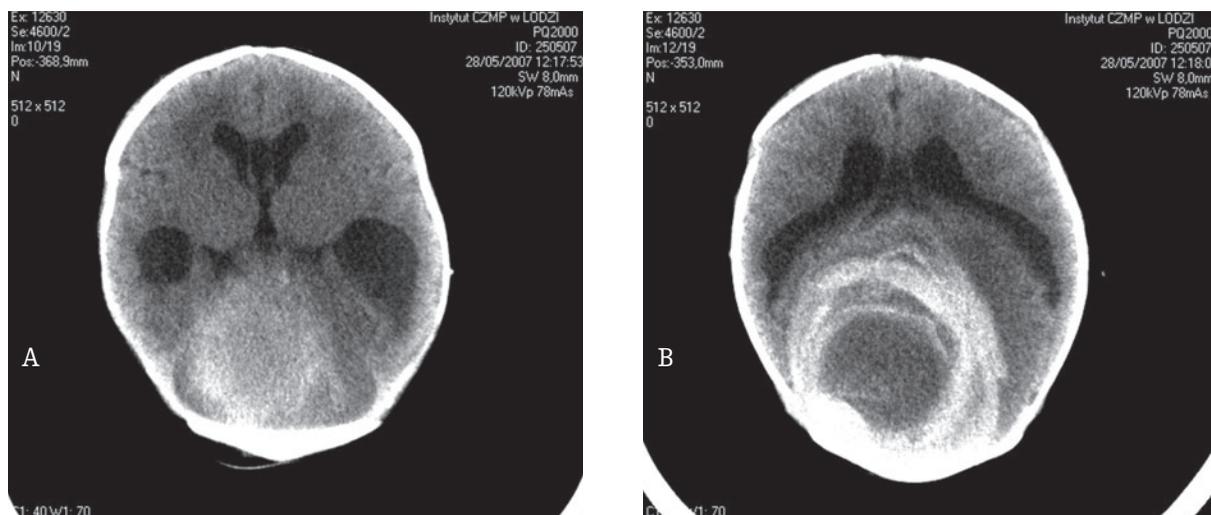


Figure 2. Axial noncontrast CT scan (A) and scan after i.v. contrast injection (B) shows hemorrhage in the tumor (*teratoma*) involving the occipital and parietal lobes of the brain. Postcontrast scan reveals enhancement of the tumor capsule (B).

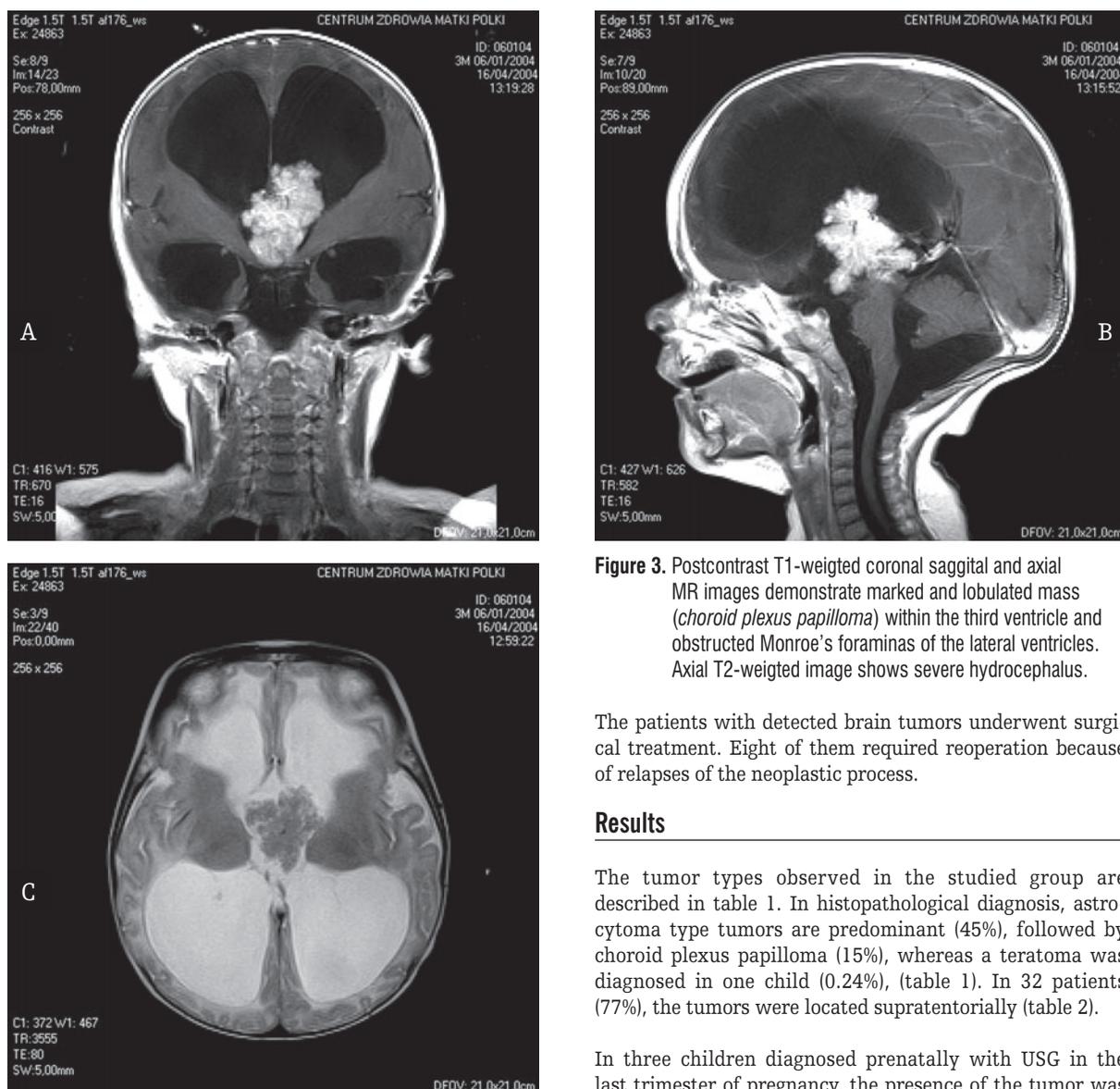


Figure 3. Postcontrast T1-weighted coronal saggital and axial MR images demonstrate marked and lobulated mass (*choroid plexus papilloma*) within the third ventricle and obstructed Monroe's foramina of the lateral ventricles. Axial T2-weighted image shows severe hydrocephalus.

The patients with detected brain tumors underwent surgical treatment. Eight of them required reoperation because of relapses of the neoplastic process.

Results

The tumor types observed in the studied group are described in table 1. In histopathological diagnosis, astrocytoma type tumors are predominant (45%), followed by choroid plexus papilloma (15%), whereas a teratoma was diagnosed in one child (0.24%), (table 1). In 32 patients (77%), the tumors were located supratentorially (table 2).

In three children diagnosed prenatally with USG in the last trimester of pregnancy, the presence of the tumor was

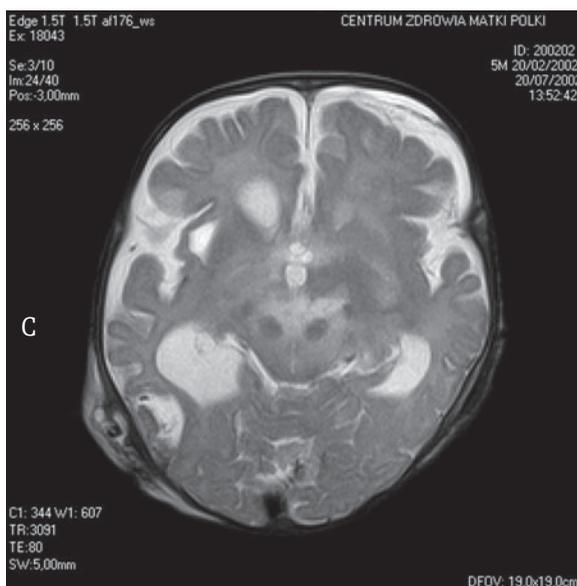
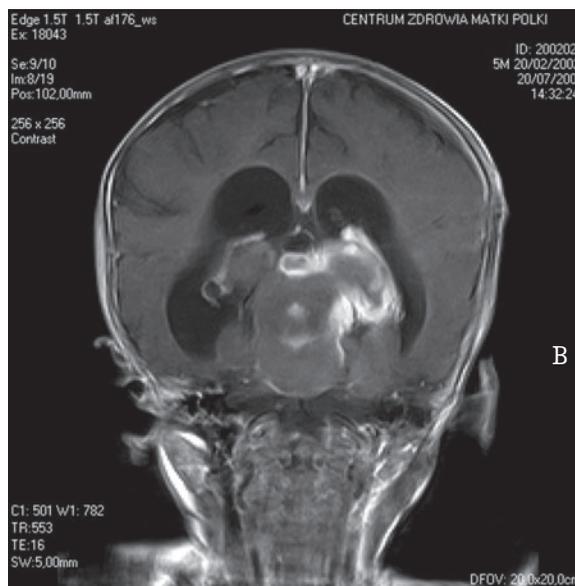
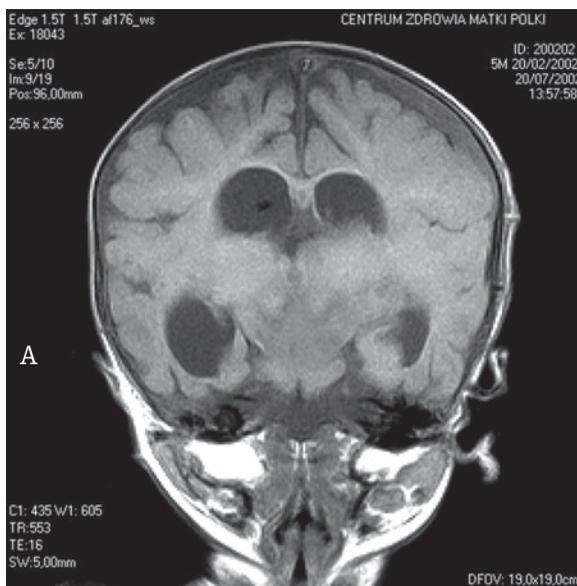


Figure 4. Coronal T1-weighted noncontrast (**A**) and after contrast administration (**B**) MRIs. Diffuse infiltration of tumor (*astrocytoma*) extending from the right basal ganglia and thalamus to the left thalamus. Axial T2-weighted image shows dilatation of the lateral ventricles (**C**).

confirmed after birth by CT and MR of the head. After the surgery, supratentorial astrocytomas were diagnosed in two cases, and a PNET type tumor located within the cerebellum in one child.

The remaining 38 brain tumors were diagnosed in children at the age of 1-12 months, most frequently, in 7 patients, at 7 months, and least frequently, in 1 patient, at 11 months.

In 8 patients a relapse of the neoplastic process was observed and they needed reoperation. In 6 of them, the cause of relapses was primary astrocytoma.

Huge tumor mass and involvement of supratentorial structures was a characteristic feature of congenital brain tumors visualized by both CT and MR. Both CT and MR revealed inhomogeneous structure of the tumors, with fluid-filled spaces within necrotic foci in the tumor tissue (fig. 1). In the children with tumors diagnosed in the prenatal period, CT and MR demonstrated the presence of massive hemorrhages

within the tumor (fig. 2). In 6 patients, choroid plexus papillomas accompanied by distension of the lateral ventricle were visualized (fig. 3). Head CT revealed in 11 patients small, amorphous calcifications within the tumor mass, predominantly located on the periphery. Intravenous contrast administration led, except for one dermoid cyst case, intensive and inhomogeneous enhancement of the tumor tissue.

Infratentorial tumors involved the whole cerebral hemisphere in one case only, and in the remaining eight patients were located medially and accompanied by symmetrical distension of the ventricular system.

Astrocytoma type lesions were most frequent among the tumors involving the supratentorial structures, whereas ependymoma and medulloblastoma were equally frequent (33%) among cerebellar tumors. Astrocytomas localized in the region of the optic chiasm (25%) presented in the scan an infiltrating mode of growth, with involvement of the sella turcica structures, including the intracranial segment of the internal carotid artery (fig. 4). The dermoid cyst type tumor developed in the cerebral hemisphere and was characterized morphologically by polycystic structure, visualized well by MR of the head.

Discussion

Supratentorial localization is a characteristic feature of congenital brain tumors, whereas in older children, above 2 years of age, brain tumors usually develop in the cerebellum. With respect to histopathological structures, teratoma, accounting for 33% of cases, is reported to be most frequent (33%) among all congenital brain tumor types [1, 7]. It is followed by astrocytoma, medulloblastoma and choroid plexus papilloma (8.9% each) [3]. A rarely diagnosed tumor is PNET (ca. 3.4%); it is detected in 4.2% of cases in children above

4 weeks of age [7]. Congenital CNS tumors of vascular origin, such as hemangiopericytoma, are observed occasionally [1]. Brain tumors usually develop in the midline, attain huge size because of elasticity of the fetal skull and cause an increase of head circumference. In prenatal examinations, brain lesions are visible as late as the 3rd trimester of pregnancy, often accompanied by asymmetric hydrocephalus and hemorrhages within the tumor mass [5,6]. The symptoms of congenital CNS tumors in viable neonates present as increased head circumference with wide cranial sutures. On clinical examination, restlessness, generalized convulsions and vomiting are observed. USG can reveal a brain tumor in the fetus in the 3rd trimester of pregnancy, if the tumor has grown to a large size; therefore, only 8% of lesions are detected prenatally or in the perinatal period [4]. In some cases of hemisphere asymmetry, the cause of enlargement of brain structures cannot be obtained unequivocally. Differential diagnosis should take into account, in addition to a neoplastic process, congenital hyperplasia of a cerebral hemisphere (hemimegalencephalia). The doubts can be resolved by MR performed on the fetus or after delivery [8]. Consistently with the reports by other authors, supratentorial localization of the tumors was predominant in the analyzed group [9]. No predominance of the female sex reported for diagnosed CNS tumors was observed among our patients [9]. Astrocytoma type histopathological structure was typical in the investigated tumor material, with a single teratoma case diagnosed in the cerebral hemisphere of a 7-month-old female infant. Only 3 patients were diagnosed in the perinatal period and their tumors can be definitely regarded as congenital, whereas the remaining ones, diagnosed in infancy (38/41%) are, according to Sobel et al., probably congenital tumors [1]. Such classification of tumors in the studied group would explain the histopathological types of the lesions.

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Conclusions

In the analyzed group of children, astrocytoma type tumors were most frequent, unlike the incidencies reported by the cited literature.