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Antenatal diagnosis of the congenital craniopharyngioma

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Summary

Background:	Craniopharyngioma is a rare fetal and neonatal tumor.
Case Report:	We report a case of a congenital craniopharyngioma diagnosed by prenatal magnetic resonance. This diagnosis was confirmed by postnatal MR imaging, neurosurgical treatment and histopathological examination.
Conclusions:	Outcome of neonatal craniopharyngioma is very poor, even if radical surgery is performed. The main problems are pituitary insufficiency, diabetes insipidus, and visual disturbance.
Key words:	fetus • congenital brain tumor • craniopharyngioma • magnetic resonance imaging
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Background

Craniopharyngioma is most commonly seen in the first two decades of human life, with predominance in childhood (it constitutes approx. 8–10% of all intracranial tumours in the paediatric age group). It is rare in the foetal and neonatal period [1–3]. One of the articles published in 2002 analysed 250 cases of congenital tumours diagnosed in the foetal and neonatal period. Craniopharyngioma was revealed in 33 of those patients [4].

Modern and non-invasive diagnostic methods (ultrasonography and magnetic resonance imaging) increase the number of tumours diagnosed in foetal life.

Case Report

An elective ultrasonography examination of a foetus revealed a hyperechogenic mass in the brain. The patient was referred for MRI, with suspicion of brain tumour in the foetus.

The MRI examination was performed at 28 Hbd of pregnancy. It revealed a mass lesion measuring approx. 40×35×34 mm, originating from the perisellar region and penetrating into deep brain structures of the right hemisphere. The tumour was compressing the body of the right lateral ventricle and the third ventricle, and was displacing the midline structures to the left, while its posterior part was compressing the brain stem. The tumour was of solid type and it was revealing inhomogeneous signal intensity which could possibly follow from the presence of minor fluid foci. Signal intensity of the tumour was not characteristic for adipose tissue. Due to the location and morphology of the lesion, a diagnosis of craniopharyngioma was suggested (Figure 1A–D).

The pregnancy was terminated in the 36th week, by caesarean section, due to the prenatally diagnosed foetal brain tumour. The newborn baby weighed 3410 g. The condition of the baby was scored 9, 10, 10 points in APGAR scale (in the 1st, 3rd, and 5th minute, respectively). Postnatal ultrasonography of the brain confirmed the presence and location

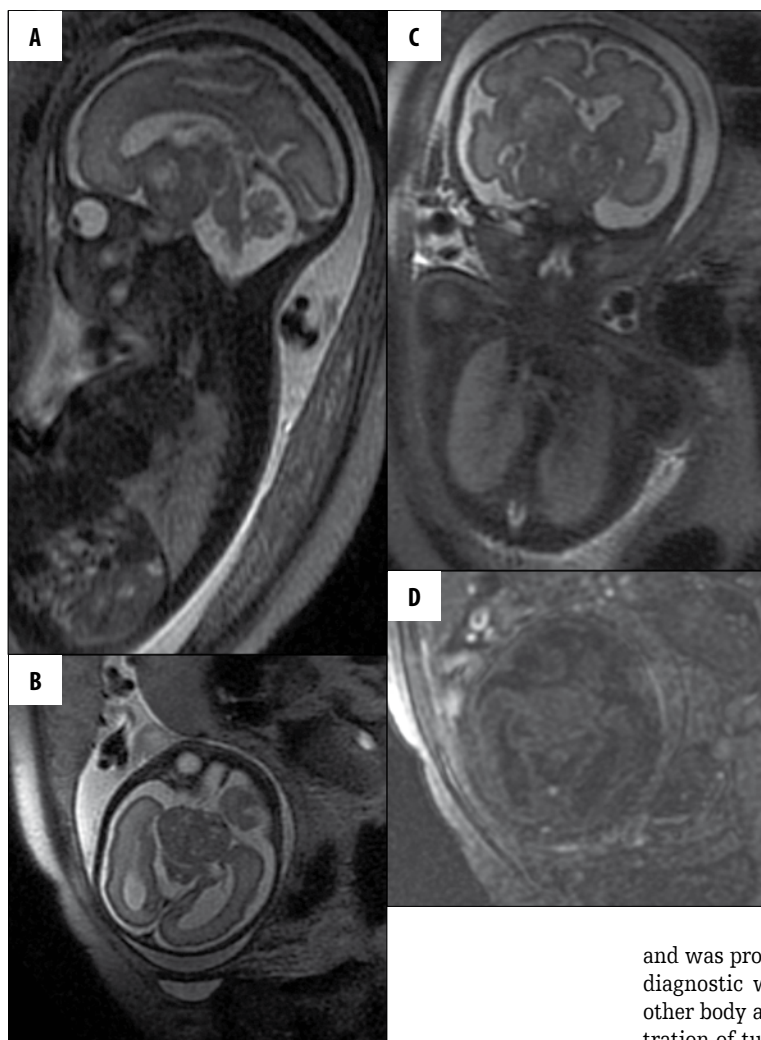


Figure 1. Fetal MRI at 28 Hbd. Solid tumor with cystic elements in the midline and in the right hemisphere, without edema. The mass effect on the right lateral ventricle, IIIrd ventricle and pons is seen. SSFSE/T2, sagittal plane (A), transverse (B) and coronal (C). FGRE/T1, transverse plane – no signal intensity typical for fatty tissue (D).

of the hyperechogenic lesion in the region of skull base and the ventricular dilation. The results of basic laboratory tests were within normal limits.

The patient was admitted to the Department of Pathology and Neonatal Intensive Care of the Children's Memorial Health Institute, with the diagnosis of a congenital brain tumour.

At admission, the baby weighed 3400 g. Its head circumference was 38 cm. It was diagnosed with hydrocele of the right testicle. No other abnormalities were found. Ultrasonography of the brain, performed after admission, revealed a hyperechogenic mass of 60×45 mm, placed medially. Lateral ventricles were moderately dilated. Frontal dimension of the right, lateral ventricle was 10.7 mm, and of the left ventricle 12.8 mm. The III and IV ventricle were not dilated. The results of ultrasonography examination of the abdominal cavity were within normal limits. In the first 24 hours of the baby's life, a brain MRI was performed (at sleep). It showed an inhomogenous, solid-cystic tumour, with the solid part measuring 49×40×58 mm in its largest dimension, and the cystic part: 29×24×31 mm. The tumour was located medially and to the right. It was occupying the suprasellar region

and was protruding into the III ventricle (Figure 2A,B). The diagnostic work-up included the presence of tumours in other body areas. No metastases were found. Blood concentration of tumour markers was monitored (decrease in AFP concentration was revealed). With the tumour remaining unenlarged and the ventricles steadily undilated, no indications were found for neurosurgical intervention or for chemotherapy. Due to the location of the tumour, endocrinological diagnostics was introduced, revealing no hypofunction of the pituitary gland.

The baby was discharged home with indications for follow-up USGs of the brain and an early AFP assay.

After 4 weeks, due to an increasing hydrocephalus, the baby was admitted to the Neurosurgical Department of the Children's Memorial Health Institute. A check-up MRI showed tumour progression, especially in its cystic part, and widening of lateral ventricles. The tumour measured 55×45×63 mm in its solid part, and 44×42×49 mm in its cystic part (Figure 3A–D).

It was decided to perform a surgical procedure. An intraoperative examination of the collected samples of the tumour confirmed the prenatal diagnosis of craniopharyngioma. On the basis of that information, a maximally radical resection of the tumour was undertaken. The procedure revealed atrophy of the optic chiasma and of the optic nerves. The tumour was curetted in the sellar region and the procedure was terminated and assessed as radical. After the surgery,

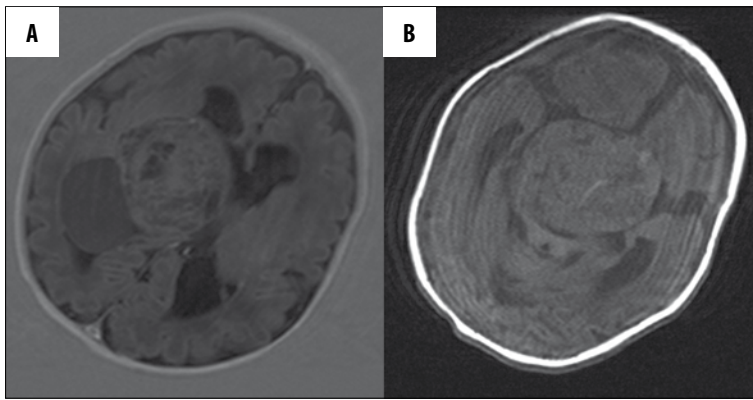


Figure 2. MRI of the newborn at the first day of life. SE/T1, transverse plane (A), (B) Solid-cystic tumor. Solid part of the tumor is in the midline, cystic part is seen laterally. Lateral ventricles are dilated (A). Scan on the level of the solid part, the midbrain is pushed posteriorly, cerebral peduncles are splayed (B).

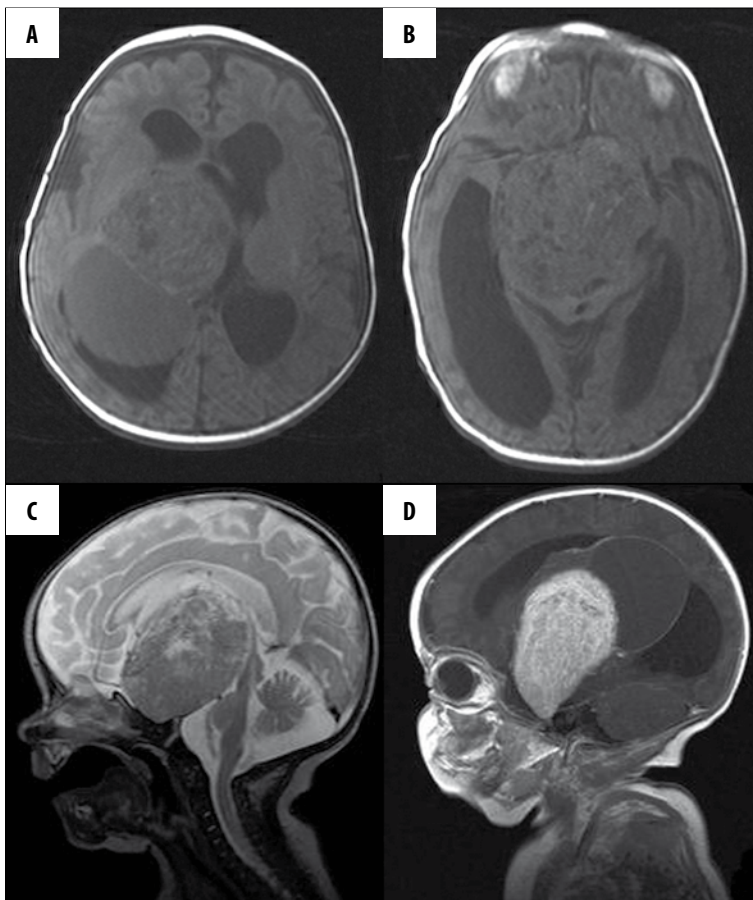


Figure 3. MR at the age of 4 weeks. Progression of the tumour. SET1, transverse plane (A), (B), TSET2, sagittal plane (C), SET1+gad, sagittal plane (D). Enlargement of the cystic part of the tumour with widened lateral ventricles (A). Slight enlargement of the solid part of the mass, with large occipital horns of the lateral ventricles (B). The midline scan, heterogenous tumour in the sellar/suprasellar region, extending posteriorly into the interpeduncular cistern and distorting the midbrain (C). Strong enhancement of the solid mass and thin rim of enhancement of the cystic part (D).

the child was transferred to the postoperative department, for further intensive care, and to the Department of Metabolic Diseases, Endocrinology and Diabetology. The baby was diagnosed with a multihormonal hypoactivity of the pituitary gland and diabetes insipidus. Substitutive hormonal treatment was introduced.

The final result of histopathological examinations was as follows: macroscopically revealed greyish, solid-cystic masses of 70×65×33 mm in size. Microscopically: regions of epithelial cells with characteristic peripheral palisading of cylindrical cells. In the central part, most of the cells were arranged loosely, with foci of keratinisation, multiple calcifications and microcysts including fluid containing desquamated epithelial cells. The

whole macro- and microscopic picture allowed for diagnosing *Craniopharyngioma, adamantinomatous type*. (Figure 4A-C).

At 3.5 month (of the baby's age), a psychological evaluation was carried out. It revealed a delayed psychomotor development. The child is blind and oversensitive to touch.

Discussion

There is no one, uniform classification of brain tumours. Invald et al. include tumours diagnosed in the prenatal and perinatal period in the group of congenital tumours. Sobel et al., on the other hand, believe that only the tumours diagnosed during labour and in the first 2 weeks can be regarded

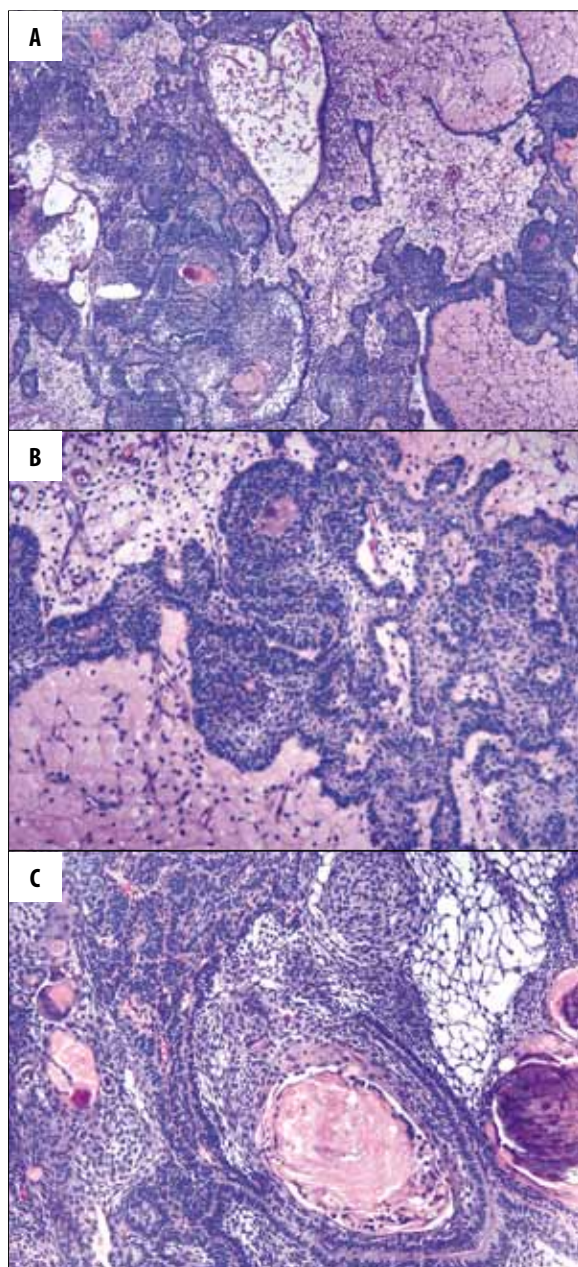


Figure 4. Histopathologic examination. Craniopharyngioma, adamantinomatous type. (A) Epithelial cells with characteristic peripheral palisading. HE ($\times 40$). (B) Epithelial cells with peripheral palisading and loose component so-called stellate reticulum. HE ($\times 100$). (C) Keratinized neoplastic cells and foci of calcification. HE ($\times 100$).

as congenital [5,6]. Other authors believe tumours developing in the first 60 days after childbirth to be congenital [7,8].

Congenital brain tumours constitute approx. 0.5–1.5% of all congenital tumours in new-born babies. Tumours most frequently described in the literature are as follow: teratomas, primitive neuroectodermal tumours (PNET), including foetal medulloblastomas, astrocytomas, and papillomas of the choroid plexus. Tumours found in the prenatal and neonatal period, in contrast to the tumours developing at later stages of life, are often situated supratentorially.

Craniopharyngioma is a benign, epithelial tumour, of stage I, according to WHO criteria. Most probably, it originates from the residues of Rathke's pouch [4]. A typical location of that tumour is the suprasellar region. It frequently penetrates the hypothalamus and the third ventricle.

Microscopic image of craniopharyngioma, adamantinomatous type, resembles the structure of adamantinoma of the mandible. Histologically, craniopharyngioma is a benign tumour. However, due to multiple processes penetrating the brain, it becomes locally invasive.

The prognosis depends on tumour size and the degree of resection. Generally, the prognosis of survival of a foetus and a new-born baby with a large tumour is poor. Due to baby's age, radiotherapy is contraindicated. With improving neurosurgical technology, total resection of such tumours is becoming increasingly feasible.

Complications resulting from tumour location are as follows: abnormal functioning of the pituitary gland (hypo-function) and visual disturbances [4].

Our patient developed symptoms of multihormonal insufficiency of the pituitary gland and diabetes mellitus. Apart from endocrinological symptoms, the baby was diagnosed with blindness resulting from atrophy of the optic chiasma and of the optic nerves, which were persistently compressed by the tumour, for a long time, in the prenatal period. During surgery, these structures were seen as very thin bands of neural tissue.

Foetal MRI is extremely important, as it may be used for planning safe termination of pregnancy, at a proper time. As it was shown in our case, morphological characteristics and location of a tumour may provide us with information on probable histopathological features of the tumour. This was confirmed by an intra- and postoperative examination. Moreover, foetal examination may be easier to carry out than the postnatal one, especially if a new-born baby is in a severe clinical condition and requires monitoring or support of basic vital functions. In the above presented case, this was not necessary. The examination could be easily performed in the first 24 hours of the baby's life, during sleep, and its quality was good. Authors' experience shows that in many cases the quality of prenatal images, especially in late pregnancy when the foetal head is placed near the internal orifice of the cervical canal, is better than after the delivery [9]. What is more, most of the time, the postnatal examination does not reveal any significant, new facts about the pathology, and does not contribute to the results of the prenatal examination [10]. In our case, it only showed larger dimensions of the tumour, which was quite obvious, considering the time which passed from pre- to postnatal MRI (8 weeks). However, this information was obtained earlier, in a transfontanelle ultrasonographic examination. In some countries, surgical procedures based on foetal MRIs are gaining on popularity.

According to our knowledge, there has been 8 published cases of craniopharyngioma diagnosed prenatally so far [2,3,11–16]. Only in four cases, the diagnosis was based on foetal MRI findings [1,3,11,12]. Therefore, our work constitutes the fifth

report on that matter. In the previous four cases known by the literature, neurological procedures were performed in the first year of a baby's life. One of the new-born died, and the

remaining three showed symptoms of multihormonal insufficiency of the pituitary gland and diabetes mellitus, as well as visual disturbances, as in our case.

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