

Fetal intracranial tumors detected by ultrasound: a report of two cases and review of the literature

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ABSTRACT

Two cases of fetal intracranial tumors detected prenatally by ultrasound, together with a review of the literature on this rare disease, are presented. Sonographic features, histopathological type and location of the tumors, sex distribution, associated anomalies and overall prognosis are described as well as obstetric and postnatal therapeutic management.

INTRODUCTION

Congenital intracranial tumors are rare and usually fatal¹. The incidence of congenital brain tumors has been estimated at 0.5–1.9% of all pediatric tumors^{2–4}, approximately 26–50% of which are teratomas^{1,3–12}. Other tumors that have been found prenatally with an intracranial location include: meningeal sarcoma¹³, cranio-pharyngioma^{14–16}, lipoma^{17–19}, glioblastoma^{7,20,21}, astrocytoma^{22,23}, oligodendroglioma²⁴, cavernous hemangioma²⁵ and undifferentiated malignant tumor²⁶.

The progress of ultrasound imaging has led to safer and more accurate evaluation of this fetal pathological condition. Other imaging techniques such as computed tomography (CT)^{27–29}, magnetic resonance imaging (MRI)^{15,16,29,30} and echo planar imaging (EPI)³¹, may improve diagnosis in the fetus. In most cases, the diagnosis of a fetal intracranial tumor does not alter the prognosis, which is uniformly dismal. There are, however, two exceptions: lipomas (usually associated with agenesis of the corpus callosum) and choroid plexus papillomas.

Early *in utero*, diagnosis and further obstetric management are limited by the fact that these tumors may grow considerably in later pregnancy, causing severe brain damage either by the tumorous mass itself or via increased intracranial pressure and associated hydrocephaly. Macrocephaly may cause dystocia, owing to cephalopelvic dispro-

portion^{1,3,9,12,22,24,26,32,33}. There is also a frequent association with polyhydramnios^{1,7–9,12,14,15,24,26–28,32–38} caused by impaired swallowing due to fetal hypothalamic dysfunction or mechanical obstruction^{7,24,26,33,35}.

The challenge is therefore to detect these lesions in early pregnancy, since this provides an indication of the problems that may develop during pregnancy and permits optimal obstetric management, including:

- (1) Prenatal counselling by pediatricians³⁹;
- (2) Use of other diagnostic methods to confirm the diagnosis and to estimate the prognosis (i.e. evaluation for associated anomalies, estimation of fetal karyotype^{28,38};
- (3) Planning of the time, mode and place of delivery;
- (4) Termination of pregnancy.

This report describes intracranial tumors that have been detected prenatally by ultrasound to examine whether it is possible to distinguish different tumors by ultrasonographic features. To this end, we have studied the relevant literature and analyzed our own experience in two cases.

CASE REPORTS

Two intracranial teratomas were diagnosed in our clinic between 1993 and 1996.

Case 1

A 28-year-old woman, gravida 4, para 2, was referred to our department at 39 weeks' gestation after an ultrasound examination performed at another center had shown fetal hydrocephaly and a suspected brain tumor. Until that time, the pregnancy had been uneventful.

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Ultrasound revealed a biparietal diameter (BPD) of 15.5 cm, which was grossly enlarged, and a solid tumor originating from the base of the cranium. Real-time scanning showed the tumor to be hyperechogenic with discrete hypoechogenic regions. There were also acoustic shadows interpreted as calcifications. The tumor measured 8.4×7.6 cm. The lateral ventricles were dilated and no normal brain architecture was identifiable (Figure 1).

At 39 + 7 weeks of gestation, a Cesarean section was performed, owing to disproportion. A boy weighing 4390 g was delivered. Apgar scores were 8 and 9 at 1 and 5 min, respectively. The infant was then immediately referred to the neonatal intensive care unit.

Physical examination revealed an extreme macrocephaly with bulging and tense anterior fontanel and split cranial sutures. No associated anomalies could be found. Ultrasound, performed on the 2nd day of life, revealed extremely severe hydrocephaly with only 5 mm of brain



Figure 1 Intracranial teratoma: a solid hyperechoic tumor originating from the base of the cranium is destroying the normal brain architecture. Dilated lateral ventricles and grossly enlarged fetal head (biparietal diameter 15.5 cm)



Figure 2 Magnetic resonance image of the fetal head, demonstrating the tumorous mass and a large hydrocephaly with partial hemorrhage

cortex in the parietal region. An amorphous tumor with heterogeneous echodensity and acoustic shadowing was found in the middle of the cranium. MRI showed an undifferentiated tissue mass of varying intensity, associated with massive hydrocephaly with partial hemorrhage. Only a thin coat of cerebral cortex could be found (Figure 2).

After lengthy discussion of the wide destruction of normal intracranial tissue and the dismal prognosis, the parents chose neither to proceed with intensive therapy nor to attempt a surgical approach. The baby died on the 14th day of life.

Autopsy disclosed an immature teratoma, partly solid and partly cystic with hemorrhage, originating from the midline. The stem of the pituitary gland, the optic chiasma and the corpora mamillaria had been destroyed by the tumor. It was invading the basal ganglia, the thalamus, the hypothalamus and the hippocampus (Figure 3). Hyaline cartilage, bone structures, epidermis and undifferentiated central nervous tissue were present on microscopic examination. Normal cerebral structures were narrowed to 1 cm by massive hydrocephaly and there was no normal relief of the cerebral cortex (Figure 4).

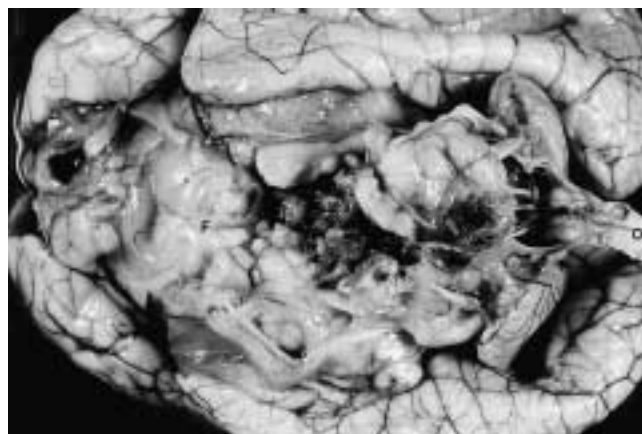


Figure 3 Basal view of the brain. The midline is occupied by a huge tumor. F, frontal; O, occipital

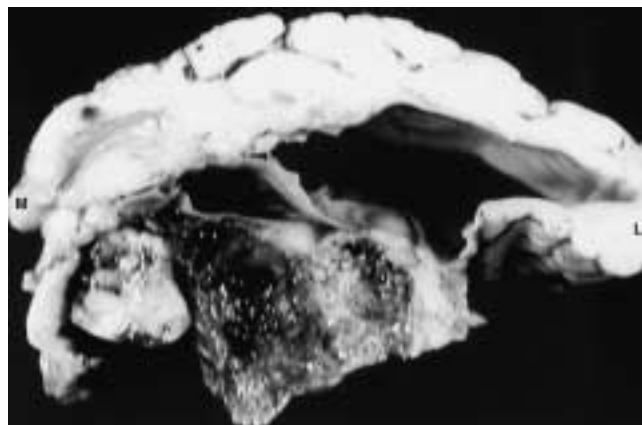


Figure 4 One-half of a pathological frontal brain section at the basal ganglia: massive hydrocephaly constricting the normal cerebral structures to 1 cm. M, medial; L, lateral



Figure 5 Intracranial teratoma: extremely enlarged fetal head with a solid intracranial tumor mass and hydrocephaly. No normal brain structures can be identified

Case 2

A 24-year-old woman, gravida 3, para 2, was referred to our ultrasound department at 30 weeks of gestation because of macrohydrocephaly revealed by an ultrasound examination performed at another center. Until then, the pregnancy had been uneventful and an ultrasound examination performed at 28 weeks of gestation (by her gynecologist, who had also performed the scan at 30 weeks of gestation) was considered to be normal.

At the time of examination in our department, the fetal head was grossly enlarged (BPD 12.8 cm). In addition to hydrocephaly, a large, solid intracranial mass of mixed echodensities was observed, located mainly in the right hemisphere. Doppler flow measurements revealed an abundant perfusion. No normal brain structures were identifiable (Figure 5).

We discussed our findings with the parents who, in view of the poor fetal prognosis in combination with the rapid growth, decided not to continue the pregnancy. The huge macrocephaly and a breech presentation made it necessary to perform a Cesarean section at 30 + 3 weeks of gestation. A girl was delivered weighing 2370 g. The head circumference was 43.5 cm, body length 42.5 cm and Apgar score 8 at 1 min. For further diagnosis, the newborn was intubated and transferred to our neonatal intensive care unit.

Physical examination showed extreme macrocephaly with wide gaping cranial sutures (Figure 6). There were no other anomalies. Only minimal vestiges of normal brain structures were identifiable by ultrasound, and the right lateral ventricle could be seen to be maximally dilated. One solid tumor mass replaced the normal intracranial architecture. The infant died the next day of cardiorespiratory arrest.

Autopsy confirmed the diagnosis of an intracranial teratoma containing cartilage, bone structure, muscle and glandular and immature nervous tissue. The cerebral cortex was extremely atrophic and the ventricles massively dilated.



Figure 6 Fetal head, showing extreme macrocephaly

DISCUSSION

The incidence of brain tumors of congenital origin has been estimated at 0.34 per million live births². Up to now, 48 cases of this rare fetal pathology diagnosed by ultrasound have been reported (authors' case included) (Table 1). Teratomas were the most common type of congenital intracranial tumor, accounting for 30 (62.5%) of the cases. Seven tumors were of neuroepithelial origin (14.6%), three were craniopharyngiomas (6.3%), five were lipomas (10.4%) and there were single cases of a cavernous hemangioma, a meningeal sarcoma and an undifferentiated malignant tumor (2.1%).

Although it was frequently impossible to determine the exact origin of these tumors, most of them were supratentorial, in contrast to the usual infratentorial location of brain tumors in older children, reported in former studies^{1,3-6,11,13,22}.

Reports of a gender bias in cases of congenital intracranial tumors are conflicting: Wakai and colleagues³ and Oi and colleagues¹¹ reported in their studies that female newborns were slightly more affected, while Buetow and colleagues⁴ and Jellinger and Sunder-Plassmann⁵ observed a slight male predominance. The data available from the literature showed that, in 36 cases in which a sex determination was made, 20 were female (55.6%) and 16 were male infants (44.4%).

Little is known about the etiology or pathogenesis of congenital intracranial tumors. A variety of theories have been proposed to explain the origin of teratomas. Among

Table 1 Intracranial tumors prenatally diagnosed by ultrasound: review of the literature. The biparietal diameter (BPD) expected for the gestational age⁴⁰ is shown

First author	Gestational age at diagnosis (weeks)	Sonographic features of the tumor	Measurement (cm)		Other sonographic findings	Obstetric management	Fetal outcome	Sex	Pathological findings	Associated anomalies
			Tumor	BPD						
DeVore ⁴¹	third trimester	complex mass	'large'	NA	NA	NA	NA	NA	teratoma	NA
Hoff ³²	28	complex mass, loss of normal brain architecture	NA	'grossly enlarged head'	macrocephaly, polyhydramnios	Cesarean section (3 days later)	neonatal death (within seconds)	M	malignant teratoma	NA
Grade ⁴²	near term	complex mass, loss of normal brain architecture	filling cranial vault	10.5 (9.7)	macrocephaly	Cesarean section (next day)	neonatal death (2 h)	NA	teratoma	none
Vinters ³⁴	30	solid mass with multiple cystic spaces	NA	15.0 (8.3), 31 weeks	macrocephaly, polyhydramnios, placentomegaly	31 weeks: Cesarean section (prior Cesarean section), cephalocentesis	stillborn	M	teratoma	micrognathia, cardiomegaly, hepatosplenomegaly
Kirkinen ²⁷	32	complex mass, loss of normal brain architecture	14 × 10 × 18, after birth	14.8 (8.5)	polyhydramnios	36 weeks: cephalocentesis, vaginal delivery	stillborn	NA	teratoma	NA
Paes ⁴³	32	complex mass, loss of normal brain architecture	NA	'large'	macrocephaly, hydrocephaly	36 weeks: cephalocentesis, vaginal delivery	neonatal death (4 days)	F	teratoma	none
Gadwood ⁴⁴	31	complex mass, loss of normal brain architecture	NA	'grossly enlarged cranium'	macrocephaly	32 weeks: Cesarean section	neonatal death	M	benign teratoma	none
Hecht ²⁸	31	complex mass	10, after birth	8.7 (8.3)	macrocephaly, polyhydramnios, abnormal facial features	31.5 weeks: vaginal delivery	neonatal death	F	teratoma	NA
Rostad ³⁵	36	complex mass	'large'	NA	hydrocephaly, polyhydramnios	37 weeks: Cesarean section	neonatal death (within minutes)	M	teratoma	horseshoe kidney
	20	complex mass, loss of normal brain architecture	12 × 10.5 × 6, after birth	'massively enlarged head'	macrocephaly, polyhydramnios	21 weeks: induction of labor, Cesarean section	stillborn	M	teratoma	none

Continued

Table 1 Continued

First author	Gestational age at diagnosis (weeks)	Sonographic features of the tumor	Measurement (cm)		Other sonographic findings	Obstetric management	Fetal outcome	Sex	Pathological findings	Associated anomalies
			Tumor	BPD						
Chervenak ³³	NA	complex mass	NA	NA	macrocephaly, hydrocephaly, polyhydramnios	32 weeks: Cesarean section	neonatal death (1 h)	N/A	benign immature teratoma	NA
Lipman ¹	30	complex mass, loss of normal brain architecture	NA	9.6 (8.1), 30 weeks 12.0 (9.0), 34 weeks	macrocephaly, hydrocephaly	induction of labor, cephalocentesis, vaginal delivery	stillborn	M	multicystic immature teratoma	none
Lipman ¹	36.5	complex mass	NA	10.8 (9.4)	macrocephaly, hydrocephaly, placentomegaly	Cesarean section	neonatal death (3 days)	NA	immature cystic teratoma	horseshoe kidney
	20	complex mass, loss of normal brain architecture	NA	12.3 (5.1)	polyhydramnios, mild fetal edema	22 weeks: induction of labor, Cesarean section	neonatal death (5 min)	M	immature teratoma	none
Mueller ⁴⁵	31	multiple cystic structures	NA	11.4 (8.3), 31 weeks 16.4 (9.0), 34 weeks	hydrocephaly	35 weeks: Cesarean section	neonatal death (5 days)	F	teratoma	none
Richards ⁹	24	complex mass, loss of normal brain architecture	'large'	10.8 (6.4)	macrocephaly, polyhydramnios	25 weeks: cephalocentesis, induction of labor (IUD), Cesarean section (pre-eclampsia)	stillborn	F	teratoma	none
Ferreira ⁴⁶	21	complex mass, loss of normal brain architecture	12.5	12.1 (5.5)	macrocephaly, hydrocephaly	23 weeks: induction of labor, vaginal delivery	stillborn	M	teratoma	craniofacial dysmorphism
Daita ⁴⁷	28	complex mass, loss of normal brain architecture	'large'	15.3 (7.6)	NA	Cesarean section	stillborn	F	malignant teratoma	none
Oj ^{48,49}	27	NA	NA	10.5 (7.3)	hydrocephaly	Cesarean section	VPS (3 days) restarted (8 months)	NA	teratoma	none
Dolkart ³⁶	33	complex mass	6 × 8	13.0 (8.8)	macrocephaly, hydrocephaly, polyhydramnios	37 weeks: Cesarean section	inoperable; death (cardio-respiratory arrest) (35 days)	F	teratoma	none

Continued

Table 1 Continued

First author	Gestational age at diagnosis (weeks)	Sonographic features of the tumor	Measurement (cm)		Other sonographic findings	Obstetric management	Fetal outcome	Sex	Pathological findings	Associated anomalies
			Tumor	BPD						
Ol ²⁹	29	complex mass	NA	NA	macrocephaly, hydrocephaly	37.3 weeks: Cesarean section	VPS (3 days) subtotal resection (4 weeks), chemotherapy, psychomotor delay (7 months)	F	malignant teratoma	none
Wienk ²⁴	30	complex mass	7–8	NA	hydrocephaly, polyhydramnios	32 weeks: induction of labor, cephalocentesis	stillborn	NA	teratoma	NA
Saiga ⁵⁰	21	complex mass, loss of normal brain architecture	'large'	abnormal overgrowth	macrocephaly, hydrocephaly, hepatomegaly	Cesarean section	stillborn	M	teratoma	facial deformity
Schwartz ³⁸	17	complex mass, loss of normal brain architecture	NA	'massive'	epignathus (oropharyngeal teratoma), macrocephaly, polyhydramnios	25 weeks: termination of pregnancy	stillborn	F	intracranial teratoma, oropharyngeal teratoma	hepatomegaly, multiple anomalies of the distal extremities
Ferreira ¹²	37	complex mass, loss of normal brain architecture	4.8 × 5.9	10.0 (9.5)	macrocephaly, hydrocephaly	Cesarean section	total resection (1 day), VPS (6 months), delayed development (18 months)	M	immature malignant teratoma	none
Sheret ³⁷	31	complex mass, loss of normal brain architecture	NA	16 (8.3)	macrocephaly, polyhydramnios, scalp edema	34 weeks: Cesarean section	neonatal death (90 min)	F	teratoma	hepatomegaly, generalized edema
Nautin ten Cate ⁵¹	30	echogenic mass, loss of normal brain architecture	6.4 × 5.9	HC: 30.4 (29.2)	macrocephaly, hydrocephaly	35 weeks: rupture of membranes, cephalocentesis, vaginal delivery	stillborn	F	teratoma with six distinct dysmorphic fetuses	none
Authors' case	39 + 7	complex mass	8.4 × 7.6 × 7.2	15 (9.7)	macrocephaly, hydrocephaly	Cesarean section	neonatal death (14 days)	M	teratoma	none
	30 + 2	complex mass, loss of normal brain architecture	'large'	12.8 (8.1)	macrocephaly	Cesarean section	neonatal death (1 day)	F	teratoma	none

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Table 1 Continued

First author	Gestational age at diagnosis (weeks)	Sonographic features of the tumor	Measurement (cm)		Other sonographic findings	Obstetric management	Fetal outcome	Sex	Pathological findings	Associated anomalies
			Tumor	BPD						
Riboni ⁷	33	echogenic mass, shifting the falx cerebri towards the left	'large'	11.0 (8.8)	macrocephaly, hydrocephaly, polyhydramnios, hydrops fetalis	Cesarean section (next day)	neonatal death (20 min)	M	glioblastoma	hydrops fetalis
Alvarez ²⁰	34	complex mass, deviated midline falx	'large'	10.5 (9.0)	macrocephaly, hydrocephaly	Cesarean section	neonatal death (1 day)	M	glioblastoma multiforma, grade III	none
Alvarez ²⁰	30/31	unilateral echogenic mass	4.3 × 4.1, increasing until 33/34 weeks	'normal'	hydrocephaly	at term: vaginal delivery	VPS (4 months), resection of 80% of the brain (6 months), radiotherapy, alive (5 years) with developmental delay	F	glioma grade III	none
Doeren ²¹	29	echogenic mass, loss of normal brain architecture	NA	HC: 34 (27.4)	macrocephaly, hydrocephaly	induction of labor, vaginal delivery	stillborn	F	glioblastoma	none
Vanlieferinghen ²⁵	37	cystic mass	'large'	HC: 36.5 (33.9)	hydrocephaly	37 weeks: Cesarean section	VPS, alive (5 years), retardation	NA	teratoma?	NA
	30	solid echogenic tumor	NA	9.8 (8.3) HC: 34 (30.1), 32 weeks	hydrocephaly	32 weeks: termination of pregnancy	stillborn	NA	capillary and cavernous hemangioma	NA
Wienk ²⁴	32	complex mass	occupying one-half of cranial cavity	NA	macrocephaly, hydrocephaly, polyhydramnios	32 weeks: induction of labor, vaginal delivery	stillborn	F	oligodendroglioma partly with oligoblastoma	NA
Roosen ²²	near term	echogenic mass	10 × 10 × 8, after resection	10.8 (9.9)	macrocephaly, hydrocephaly	Cesarean section (next day)	VPS, extirpation (1 month), nearly normal development	M	anaplastic astrocytoma grade III	none
Heckel ²³	30	echogenic mass, loss of normal brain architecture	6 × 8 × 5, after birth	9.6 (7.8) HC: 31.8 (28.3)	macrocephaly, hydrocephaly	32 weeks: termination of pregnancy	stillborn	F	anaplastic astrocytoma grade IV	none

Continued

Table 1 Continued

First author	Gestational age at diagnosis (weeks)	Sonographic features of the tumor	Measurement (cm)		Other sonographic findings	Obstetric management	Fetal outcome	Sex	Pathological findings	Associated anomalies
			Tumor	BPD						
Snyder ¹⁴	35	echogenic mass, loss of normal brain architecture	NA	12.0 (9.2)	macrocephaly, polyhydramnios	34 weeks: preterm labor 35 weeks: Cesarean section	neonatal death (3 days)	F	craniopharyngioma	NA
Bailey ¹⁵	27	complex mass (mostly solid)	4	NA	hydrocephaly, polyhydramnios	36 weeks: Cesarean section (increasing BPD and hydrocephaly)	CT-guided biopsy, VPS (9 days), death (8 weeks)	NA	craniopharyngioma	none
Kueltuysay ¹⁶	29	complex mass	0.25	NA	macrocephaly	at term: Cesarean section	death within surgery (14 weeks)	NA	craniopharyngioma	NA
van Vliet ¹³	32	complex mass	NA	10.5 (8.5), 32 weeks 11.5 (9.0), 34 weeks	macrocephaly, hydrocephaly	34 weeks: induction of labor, cephalocentesis, vaginal delivery	stillborn	F	undifferentiated sarcoma of the dura	none
Shawker ²⁶	25	triangular cystic space, no normal brain architecture	1 × 2 × 6	8.0 (6.7)	macrocephaly, polyhydramnios	27 weeks: Cesarean section (abruptio placentae)	stillborn	M	malignant tumor (origin of the tumor could not be specified)	NA
Christensen ¹⁷	40	small midline echogenic structure	0.3 × 0.4, after birth	'normal'	none	vaginal delivery	healthy child	M	lipoma of the corpus callosum	none
Mulligan ¹⁸	37.5	bright echogenic masses	NA	HC: 90th centile	hydrocephaly, agenesis of corpus callosum	at term: Cesarean section (breech presentation)	healthy child	NA	lipoma	none
Bork ¹⁹	35	echogenic mass	NA	NA	none	at term: induction of labor, vaginal delivery	healthy child	F	lipoma	agenesis of corpus callosum
	26/30	echogenic mass with irregular margins	1.4, 30 weeks 1.7 × 2.5, 38 weeks	'normal'	mild hydrocephaly, partial agenesis of corpus callosum	40 weeks: vaginal delivery	healthy child (6 months)	F	lipoma	none
	37	echogenic midline mass	4.1 × 1.4	NA	none	37 weeks: vaginal delivery	healthy child (6 months)	F	lipoma	none

HC, head circumference; NA, not available; M, male; F, female; IUFD, intrauterine fetal demise; VPS, ventriculoperitoneal shunting; CT, computed tomography

these are theories suggesting that teratomas originate from displaced germ cells, pluripotent embryonic cells or extra-embryonic cells, or from an included (i.e. phagocytosed at an early embryonic stage) twin pregnancy^{33,50}. Although various drugs are found to cause central nervous system anomalies⁵² and one case report described a possible connection between a sacrococcygeal teratoma and acetazolamide⁵², no links have yet been reported between any drugs or chemicals and intracranial tumors.

Overall fetal outcome was generally poor. Of the cases reported in the literature, 17 babies were stillborn, 16 infants died in the neonatal period and three after this period. Only 11 children were alive at the time of publication^{12,17-20,22,25,29,48,49}. In one case no information was available on the fetal outcome⁴³. This results in an overall mortality of 77.1%. Although long-term survival may be possible^{12,17-20,22,25,29,48,49}, the risk of neurological impairment is high^{12,20,25,29,48,49}. Lipomas, however, seem to be an exception. All five children with prenatally diagnosed intracranial lipomas are reported to be healthy.

Poor prognosis is associated, in most cases, with a large size of tumor, especially in teratomas, as well as poor general condition of the newborn at presentation. The histological type of the tumor may also influence the prognosis, as was shown by Wakai and colleagues³. They found a relatively good prognosis in choroid plexus papillomas (1-year survival 50%) and the next best with astrocytomas (1-year survival 44.4%). In contrast, the survival rate for teratomas was only 7.2%. Our collected data confirm this. As mentioned, lipomas had a good outcome (100% survival, no developmental handicap), while for other tumors the prognosis was relatively poor (teratomas 10% survival, tumors of neuroepithelial origin 28.6% survival).

Ultrasonographic features of the tumor were mostly disordered arrays of echoes varying in distribution and intensity and distortion of the normal intracranial anatomy. Although it is not possible to make a definitive diagnosis of the histological type by ultrasound alone, it is possible to distinguish two groups.

Teratomas, astrocytomas and craniopharyngiomas have a similar appearance (a complex mass distorting the brain architecture, possibly associated with macrocephaly, hydrocephaly and calcifications). Even after birth, a biopsy is frequently necessary to make a definitive diagnosis. The prognosis of these lesions, however, is uniformly poor (overall survival 14%).

Intracranial lipomas are well-defined echogenic areas, usually located in the midline, in the position normally occupied by the corpus callosum, and/or within the bodies of the lateral ventricles; the latter have a good outcome.

Macrocephaly (assessed by an enlarged BPD) was reported in 38 cases (79.2%), hydrocephaly was associated in 28 of all cases (58.3%) and polyhydramnios was also a frequent symptom (18 cases or 37.5%). Intracranial calcifications were usually formed (especially in teratomas and craniopharyngiomas) but were more easily detectable in a CT²⁷ or an MRI scan^{15,16}. High-output cardiac failure, manifested in fetal hydrops, hepatomegaly, extramedullary hematopoiesis, polyhydramnios and a hydropic placenta

was reported in four cases^{7,34,37,38}. This is most probably due to the large cardiac output required by tumor arterio-venous shunting in the massive intracranial tumor.

The widespread use of ultrasound has led to earlier diagnosis of intracranial tumors in some cases^{9,26,35,46,50}. Nevertheless, a normal sonogram performed early in gestation does not rule out the possibility of a brain tumor developing later in pregnancy, as has been reported by some authors^{1,7,12,13,17,19-23,35,36,42} and as we found in one case (normal ultrasound examination at 28 weeks of gestation and a huge tumor at 30 weeks of gestation). This may be explained by the fact that intracranial tumors (especially teratomas) often display rapid growth spurts and then suddenly produce large-for-dates fetuses in otherwise normal pregnancies. Explanation for such massive growth in the fetal period is unknown, although Rostad and co-workers³⁵ have put forward three hypotheses:

- (1) The time of origin of the neoplastic cells is closer to the time of conception, allowing a greater number of days for growth;
- (2) Neoplasms that occur *in utero* have an abnormal growth pattern;
- (3) The fetal period offers a better milieu for the growth of immature elements.

Distinguishing intracranial tumors and other intracranial lesions by ultrasound may not always be easy. The most important differential diagnosis is meningoencephalocele. Lipman and co-workers¹ pointed out that teratomas can be ruled out by the skull defect associated with meningoencephaloceles. Included in the differential diagnosis are cystic lesions of the central nervous system such as porencephaly, hydranencephaly, holoprosencephaly, other neoplastic cystic lesions as well as intrauterine infections and ischemic brain necrosis.

Although real-time ultrasound is still the diagnostic method of choice for fetal tumors, and although it is often more useful to perform repeated ultrasound examinations (e.g. weekly), a further diagnostic approach such as MRI^{15,16} may be used in addition. For teratomas, α -fetoprotein may also be a very useful diagnostic marker. α -Fetoprotein is produced in the liver and the yolk sac, and its concentration in amniotic fluid gradually decreases in normal pregnancy as pregnancy advances. Elevated serum and amniotic fluid levels have been reported in some teratoma cases^{28,29,38,47,50,53}. Nevertheless, although α -fetoprotein is helpful in the diagnosis of prenatal pathology, an increased level cannot be regarded as pathognomonic for teratomas.

Once the diagnosis of an intracranial tumor has been established, a thorough search for other abnormalities should be made. In addition, fetal echocardiography and determination of karyotype are recommended, since the presence of other fetal anomalies would have an important impact on the prognosis and thus the management of pregnancy. However, karyotyping was performed in only three reported cases^{23,28,38}. Hecht and associates²⁸ found an inverted duplication of chromosome 1. The fetus reported

by Schwartz and co-workers³⁸ displayed two different cell lines: [46,XX,t(1;19)(p11;p11)] – 10% and [47,XX,t(1;19)(p11;p11) + der(1)t(1;19)(p11;q11)] – 90%. The latter karyotype was trisomic for both 1q and 19p. The case reported by Heckel and colleagues²³ showed a normal female karyotype.

In the 48 cases of ultrasonographically detected intracranial tumors, six fetuses were reported to have associated anomalies (12.5%); these included three facial dysmorphologies, two horseshoe kidneys and one fetus with multiple anomalies of the distal extremities. This confirms the finding of Wakai and co-workers³, who reported that 14% of the definitely congenital brain tumors are likely to have associated anomalies. The correlation between congenital intracranial tumors and other anomalies indicates that there may be a genetic factor involved in the pathogenesis of these tumors.

Obstetric management was dependent on gestational age at the time of diagnosis. If detected early in gestation (< 26 weeks), the pregnancy was terminated after a detailed consultation with the parents in five of seven cases, owing to the poor prognosis^{1,35,38,46,50}. In two cases, the parents decided to continue the pregnancy^{9,26}. In both cases, however, a Cesarean section had to be performed soon afterwards, owing to intrauterine fetal demise and pre-eclampsia in the case reported by Richards⁹ and abruptio placentae in that reported by Shawker and Schwartz²⁶. In one case⁴¹ no information about the obstetric management was given. Overall, a Cesarean section was performed in 29 cases (60.4%). In most cases, the reason for abdominal delivery was that the head was considered to be too large for vaginal delivery and might have caused dystocia. In 14 cases (29.2%), vaginal delivery was achieved.

Cephalocentesis was performed in eight cases^{1,9,13,24,27,34,43,51} to prevent dystocia (16.7%), and in six cases this made vaginal delivery possible (75%). The remaining two underwent Cesarean section owing to a prior Cesarean section³⁴ and pre-eclampsia⁹. After delivery, ventriculoperitoneal shunting was performed in seven of the 30 newborns. In three of them, this remained the only therapeutic intervention^{15,48,49}. Five infants underwent surgery. Radical extirpation was possible in two tumors^{12,22}. The overall outcome after resection of tumors was poor. Although long-term survivors were reported^{20,22}, most cases had severe psychomotor handicaps.

Less is known about the efficacy or the risks of additional therapeutic strategies such as chemotherapy or radiation therapy. Chemotherapy is the adjuvant therapy of choice according to some authors^{29,54,55}. Although irradiation of the neonatal brain is claimed to be well tolerated in many patients⁵⁶, whole-brain irradiation in this age group may cause severe handicaps later on in the child's development^{22,29,57}.

CONCLUSION

Increasing use of diagnostic ultrasound in pregnancy has improved the prenatal diagnosis of congenital intracranial

tumors. The sonographic features of this lesion are cranial enlargement and gross distortion of the normal cerebral anatomy by a tumor mass with internal echoes of varying distribution and intensity. Hyperechogenic regions may represent calcifications, but are usually more easily detectable under MRI^{15,16}.

The ultrasound features of teratomas, astrocytomas and craniopharyngiomas have a similar appearance: a complex mass distorting the cerebral architecture, possibly associated with macrocephaly, hydrocephaly and calcifications.

Intracranial lipomas are well-defined echogenic masses, usually located in the midline and possibly associated with agenesis of the corpus callosum.

Polyhydramnios is also frequently associated with intracranial tumors.

The most common type of intracranial tumor is teratomas, accounting for 62.5% of cases.

In contrast to the infratentorial location in older children, most congenital intracranial tumors are of supratentorial origin. There is no significant difference in sex distribution.

Other anomalies are associated with intracranial tumors in 12.5% of cases. Since these may have an important impact on the prognosis, a thorough search for secondary fetal anomalies should be made directly after diagnosis of an intracranial tumor.

The overall prognosis for intracranial tumors is poor, depending on the size and histological type of the tumor. Lipomas are the only tumors associated with a good fetal outcome.

Obstetric management is dependent on gestational age at the time of diagnosis. If the tumor is detected early in pregnancy, the parents should consider terminating the pregnancy. These tumors may grow to excessive proportions and most of them are first diagnosed at an advanced gestational age. The time and mode of delivery should be determined individually for each patient. However, the incidence of Cesarean section is still 60.4%, because cephalopelvic disproportion is common.

In spite of recent advances in neonatal surgery and the availability of additional therapeutic strategies, all cases with fetal intracranial tumors (except lipomas) still have a very poor prognosis.

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