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CASE REPORT

FETAL CRANIOPHARYNGIOMA: CASE REPORT

MARCELLO VIGGIANO[1], GUSTAVO TEIXEIRA[1], CAIO MATIAS[1], ROGÉRIO FAGUNDES[2]

ABSTRACT

INTRODUCTION: Craniopharyngiomas are the most common parasellar tumors in children and adults, but rare in the perinatal period (representing only 5.6% of all fetal and neonatal tumors). They are of benign origin from the histopathological point of view, and may present an evolution that is difficult to control due to their location and ability to infiltrate surrounding tissues.

CASE REPORT: This paper reports a case of fetal craniopharyngioma diagnosed by histopathology and suspected based on an obstetric ultrasound examination.

DISCUSSION: Despite the diagnostic difficulty, fetal craniopharyngioma can be suspected in the prenatal period

CONCLUSION: Rare condition during pregnancy. It is concluded that craniopharyngiomas can be suspected during pregnancy through ultrasound and nuclear magnetic resonance, however the definitive diagnosis can only be established with anatomopathological study.

KEYWORDS: BRAIN NEOPLASMS, PREGNANCY, PRENATAL DIAGNOSIS, PREMATURITY

INTRODUCTION

Congenital brain tumors are a rare condition that have an incidence rate of 1.7 to 13.5 per 100,000 live births, representing 10% of all prenatal tumors [1,2]. Congenital tumors of the central nervous system (CNS) can be divided into teratomas and non-teramatous tumors. Teratomas are the most frequent, but there are also neuroepithelial and mesenchymal tumors (such as craniopharyngioma) and others of different origins [3,4].

The association between ultrasound (US) and magnetic resonance imaging (MRI) has allowed more accurate diagnoses of congenital CNS tumors during pregnancy, although a normal ultrasound examination in the first trimester of pregnancy does not rule out the late appearance of a brain tumor in the fetus [3] .However, the final diagnosis can only be confirmed after birth, through histological examinations, molecular analyzes or genetic tests [2].

Craniopharyngiomas are the most common parasellar tumors in children and adults, representing 5-10% of all pediatric tumors, despite being rarely found in the perinatal period, representing only 5.6% of all fetal and neonatal tumors [1]. These tumors develop from embryonic remains of squamous cells originating from Rathke's pouch (ectodermal diverticulum originating from the upper limit of the oropharynx), a structure that extends from the sella to the pharynx, located at the origin of the adenohypophysis. They are of benign origin from the histopathological point of view, and may have an evolution that is difficult to control due to their location and ability to infiltrate surrounding tissues [1,5].

Thus, the aim of this study is to report a case of fetal craniopharyngioma.

CASE REPORT

An 18 years old primigravida patient with no comorbidities was admitted to a public maternity hospital that is a reference for high-risk pregnancies in the state of Goiás, with extreme preterm labor at 23 weeks and 3 days of gestation. Ultrasound was requested to assess presentation, biometrics and fetal morphology.

The ultrasound examination, carried out in the unit, revealed a complex, solid-cystic image, with regular contours (15.7 x 10.5 x 9.4 cm) contiguous to the skull and face, and the diagnosis of fetal teratoma was questioned (Figure 1). Thus, the patient underwent cesarean delivery due to preterm labor, breech fetus, and cephalopelvic disproportion marked by an extensive fetal cranial mass.

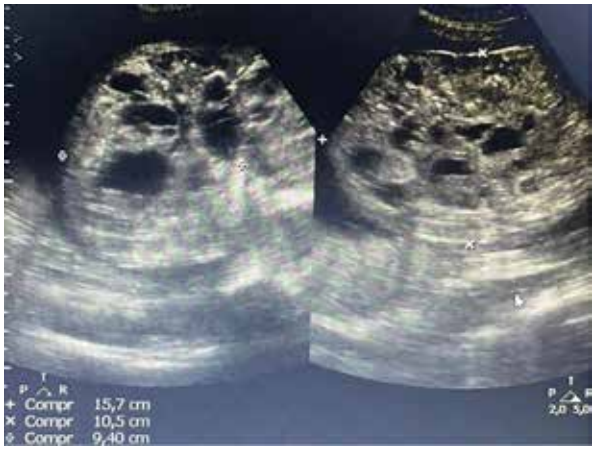

Figure 1 – Ultrasound image of complex image, contiguous to the skull and fetal face

1. UFG
2. UniEvangélica

MAILING ADDRESS:
DR. MARCELLO VIGGIANO
Rua 1028 n.70 apt. 902 - Setor Pedro Ludovico
Goiânia-GO CEP 74823-130
Email: marcelloviggiano@ig.com.br

After extracting the newborn, the female gender was comfirmed, weighing 1465g, with a height of 28cm and a head circumference of 42cm. In addition, she had malformations on the face and skull on the left, with an extensive mass that made it impossible to perform neonatal resuscitation, and he was then declared dead due to the absence of a breathing pattern or heart rate (Figure 2).

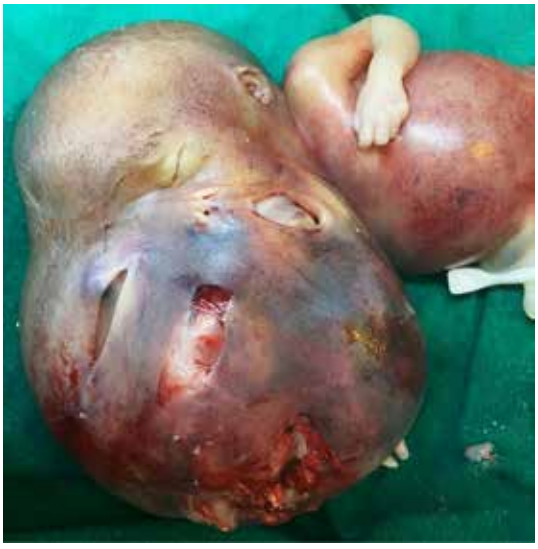

Figure 2 – Craniopharyngioma

The conceptus was sent for anatomopathological study, where it was confirmed a craniopharyngioma and chorioamnionitis, with no other findings.

DISCUSSION

The main finding in the prenatal diagnosis of a congenital tumor is an intracranial mass with a solid, cystic or mixed pattern, with or without visualization of hypervascularization on fetal ultrasound and magnetic resonance imaging. Unlike older children, congenital fetal brain tumors occur mainly in the supratentorial region, in about 70% of cases, and only in 30% are infratentorial[3,4,6].

Most newborns with congenital CNS tumors have a poor prognosis and die shortly after birth, with a survival rate of around 28% [4]. Totally cystic craniopharyngiomas evolve better than those with solid or mixed lesions [5].

Most of these tumors are diagnosed by routine prenatal ultrasound, during the second or third trimester of pregnancy [2,6]. The ultrasound is the main method used to establish the correct diagnosis during pregnancy, as it can detect solid, cystic or calcified lesions [4]. Magnetic resonance imaging can help in determining the remaining brain structures and in the exact location of the tumor, as well as in differentiating between calcifications, hemorrhages and fat deposits, allowing a detailed assessment of the relationship between the tumor and the adjacent brain parenchyma [4,6].

The most common clinical manifestations of these tumors are macrocephaly (28%), due to the presence of a tumor mass and/or fluid and hydrocephalus (17.3%) due to compression of the ventricular system [3]. The presence of macrocephaly can even cause damage such as uterine rupture or severe dystocia with cephalopelvic disproportion, justifying the concern about performing a cesarean section as in the case described [6]. Some factors suggest a poor prognosis, such as tumor location, histological type, surgical resection and general conditions at the time of diagnosis [3].

CONCLUSION

Fetal brain tumors, such as craniopharyngioma, are an extremely uncommon condition and their diagnosis during the prenatal period is challenging. Prenatal care involves a multidisciplinary team, in addition to the use of advanced imaging techniques, such as high-quality ultrasound and fetal magnetic resonance, together with clinical information to guide the decision-making process.

Therefore, despite the imaging exams helping in the process, it is concluded that the anatomopathological analysis remains the gold standard for the definitive diagnosis of this tumor.

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