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CASE REPORT

A CHALLENGING PRENATAL CASE: UNRAVELING THE RARE AND LETHAL SACROCOCCYGEAL TERATOMA

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ABSTRACT

This is a case report referring to prenatal care and follow-up in the third trimester of pregnancy, which was referred to confirm the initial diagnostic hypothesis of Myelomeningocele. After morphological ultrasound an even rarer and lethal pathology was diagnosed: Sacrococcygeal Teratoma (SCT). Sacrococcygeal teratoma is a germ cell tumor, and such fetal neoplasia is associated with complications such as prematurity, hydrops and fetal death, and may also lead to maternal complications such as maternal mirror syndrome, where the mother presents the same fetal complications such as generalized edema and pleural effusion.

The case in question evolved with a favorable outcome for the pregnant woman and an unfavorable one for the fetus, culminating in death, even with the neonatal ICU apparatus, thus reinforcing the severity of the malformation.

KEYWORDS: SACROCOCCYGEAL TERATOMA, GERM CELL NEOPLASM, NEURAL TUBE DEFECTS, HIGH RISK PRENATAL CARE, FETAL MALFORMATION.

INTRODUCTION

Sacrococcygeal teratoma is a type of tumor that originates from germ cells and is most common in childhood, representing approximately 40% of all tumors of this type. It can also be identified during the fetal stage, with an approximate incidence of 1 in every 27,000 cases.

In this specific case, the diagnosis was made late through an ultrasound performed at a specialized and high-risk service. Obstetric ultrasound was used with morphological parameters, even though the ideal gestational age for this type of evaluation had already passed.

Despite being a relatively common neoplasm, its diagnosis is complex, and when detected, it allows for a more active approach to prevent unfavorable outcomes, as occurred in this case. Therefore, the aim of this study is to present ultrasound images suggesting the presence of this tumor, with the goal of early diagnosis of this condition. Additionally, it highlights possible complications associated with this condition, both during the prenatal and postnatal periods.

CASE REPORT

A 28-year-old patient, in her third pregnancy and with two previous cesarean sections, was referred to her first high-risk prenatal consultation at the Hospital da Mulher de Goiânia (HEMU). The initial suspicion was a possible myelomeningocele, due to changes detected in a previous ultrasound. In this initial assessment, the patient did not present specific complaints, having sought assistance only due to changes in the previous ultrasound examination.

The previous ultrasound examination showed a mild increase in the amniotic fluid index (AFI) and the presence of a hypoechoic oval-shaped image in the lumbar region, which could correspond to a myelomeningocele. Laboratory tests and the patient's physical examination did not show significant abnormalities. Therefore, the patient was advised to undergo an obstetric ultrasound with morphological parameter evaluation, although it was outside the ideal timeframe for the second-trimester morphological examination (typically between 20 and 24 weeks). The objective was to confirm the diagnosis and determine the appropriate course of action for prenatal care and delivery.

The second ultrasound was performed seven days after the first consultation and showed an amniotic fluid index (AFI) with values above the normal range, confirming the diagnosis of polyhydramnios. Additionally, an image consistent with a sacrococcygeal teratoma measuring 11.5 x 9.5 cm was identified, changing the diagnostic suspicion to this rare condition with a worse prognosis (Figure 1).

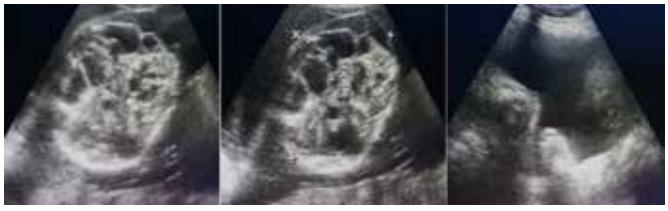

Figure 1. Ultrasound shows a mixed tumor image in the fetal sacral region and polyhydramnios.

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Six days after the first ultrasound, during a new fetal assessment, significant fetal anemia was observed due to the large accumulation of fetal fluids caused by the tumor. At this time, the middle cerebral artery peak systolic velocity was measured at 60.3cm/s (> 1.5MoM), indicating moderate anemia. Given the gestational age of 32 weeks and 3 days, the patient was admitted for lung maturation and, subsequently, the pregnancy was planned to be resolved by cesarean section.

The patient remained in the hospital ward for another three days to stabilize before the cesarean section was performed. The indication for cesarean section was related to fetal malformation, moderate anemia and worsening Doppler flowmetry results.

During delivery, a single female fetus was extracted, which presented an APGAR score of 3 in the first minute and 6 after 5 minutes. The fetus presented a large mass in the sacral region, without ulcerations, with several nodulations and hardness to the touch, in addition to a lower ear implantation.

The newborn (NB), despite not crying or breathing immediately after birth, received resuscitation, including positive pressure ventilation, and was transferred to a stabilization room. The pediatric team described a 15-20cm lesion in the sacral region, without ulcerations, with several hardened nodules and significant collateral circulation (figure 2). Additionally, a low auricular implantation was observed.

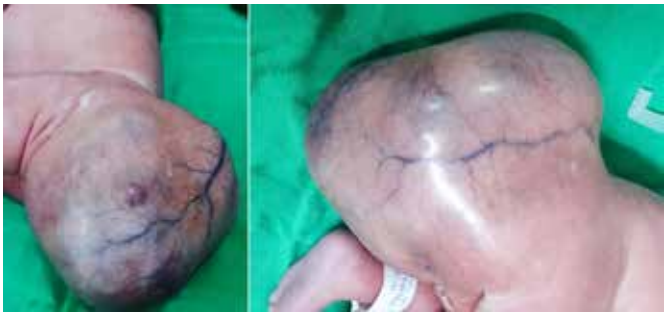

Figure 2. Neonatal image shows tumor mass in the sacral region with exuberant collateral circulation.

The NB remained in the stabilization room before being transferred to the neonatal ICU. However, after 28 hours of life, the newborn presented a significant worsening in herr overall condition, including hemodynamic instability, cold extremities and slow peripheral perfusion, indicating shock (figure 3). Non-invasive ventilation, orotracheal intubation, central venous access puncture and administration of adrenaline were necessary to stabilize her. Measures were adopted for hemodynamic compensation, such as expansion with saline solution and transfusion of packed red blood cells.

Figure 3. Neonatal image 28 hours after birth shows a significant increase in the tumor mass.

After these interventions, the NB underwent a chest X-ray, which revealed a bilateral pneumothorax. Chest drainage was performed, but the NB did not show hemodynamic or respiratory improvement. Unfortunately, the NB went into cardiorespiratory arrest, with unsuccessful cardiopulmonary resuscitation attempts, leading to the patient's death.

This case report highlights the complexity of diagnosing and managing sacrococcygeal teratoma, as well as the serious complications associated with this condition, both prenatally and postnatally.

DISCUSSION

Sacrococcygeal tumor is the most common germ cell tumor of childhood. In the pediatric population they account for 40% of all germ cell tumors, rarely in adulthood. It is the most frequently recognized fetal neoplasm, with an estimated incidence of approximately 1 in 27,000, and more common in females [1].

Teratomas are generally composed of cells representing all three germ cell layers, and present in utero as a mass extending outward from the caudal end of the fetus, or as a childhood tumor that may be asymptomatic, or show signs of obstruction of the rectum or bladder [1,2].

The Altman classification describes the extent, whether the tumor is external and/or internal. Type I tumors are mainly external, while type IV lesions are entirely internal. Type I and II tumors are the most evident on prenatal ultrasound and clinical examination. Type IV tumors are generally found later, in early childhood, compared to tumors with an external component (types I-III) [1]. Compared with the case in question, it is clear that the corresponding type is type I.

Prenatal diagnosis typically occurs during the second trimester, which underscores the difficulty in diagnosing the case in question, as it was identified at the beginning of the third trimester. Most SCTs diagnosed prenatally are solid or mixed, with both cystic and solid components; calcifications are often present, consistent with the reported case, which corresponds to a mixed tumor with solid and cystic components and interspersed calcifications[2,3].

Associated structural abnormalities may include bladder

outlet obstruction and hydronephrosis, rectal stenosis or atresia and cardiomegaly secondary to vascular shunt and high-output heart failure, characteristics that were not observed in the case analyzed [3].

Fetal magnetic resonance imaging (MRI) is recommended when available. Compared to ultrasound, MRI more accurately characterizes the intrapelvic and abdominal extent of the tumor and the compression of adjacent organs. This information can assist in prenatal counseling and preoperative planning for surgical resection. Although these options were considered, due to the rapid progression of the condition, it was not possible to obtain a more precise diagnosis through MRI, and the primary course of action was based on the ultrasound diagnosis.

The most important differential diagnosis of an exophytic cystic sacral mass in the fetus is a distal neural tube defect (such as myelomeningocele or myelocystocele). This fact was confirmed when analyzing the reason for referring the case to the referral hospital, and the initial suspicion of the investigation was indeed this highly relevant differential diagnosis. Therefore, it is essential to keep in mind that both pathologies should be considered when encountering such abnormalities[4]. It is important to note that SCTs always have a portion near the coccyx; they can extend to the sacrum, but the mass effect is usually pre-sacral and not posterior, as would be the case with a neural tube defect[4].

Serial ultrasound evaluation of the fetus, placenta, and tumor throughout pregnancy is an essential component of the overall treatment plan. The primary goal is to identify fetuses at increased risk of fetal death due to hydrops resulting from high-output cardiac failure related to vascularization and tumor size and to intervene as appropriate. This is another crucial point highlighted in the article by Egler et al [1], which also inherently connects with the case reported in this article. Through fetal middle cerebral artery Doppler, fetal anemia can be identified, which was consistent with cardiac failure and hydrops that led to fetal death in the outcome of the case in question. Therefore, it emphasizes the importance of early identification and proper management of this fetal pathology.

The size of the tumor should be measured in each ultrasound examination, and the solid portions of the tumor should be assessed with Doppler ultrasound to evaluate vascular flow 1,5. Large lesions (>10cm), such as the one in this case, which was around 15-20cm, especially vascular ones, are associated with a high rate of perinatal mortality, consistent with the unfavorable outcome in this case. Relatively cystic lesions with absent or mild vascularity tend to exhibit slow growth and a favorable outcome, even when large (>10 cm) 1,5.

The amniotic fluid volume and placental thickness should also be assessed because polyhydramnios and placental thickening are markers of hydrops, and oligohydramnios can result from bladder obstruction due to SCT [4]. In the analyzed case, there were significant polyhydramnios and signs of hydrops.

The frequency of ultrasound examinations depends on the composition of the tumor (i.e., cystic or solid), its vascularity, and any associated findings. Follow-up imaging can be as often as twice a week for high-risk tumors or as infrequently as every two weeks for small or predominantly cystic lesions [4]. This point could not be analyzed in the case due to its severity, which required a more rapid and decisive resolution.

Fetal echocardiography is recommended in fetuses with predominantly solid and/or vascular tumors. Echocardiography is used to identify a high-output cardiac state that precedes the onset of hydrops. A fetal cardiac profile consisting of the evaluation of the cardiothoracic ratio, cardiac dimension scores, combined ventricular output, and valvular regurgitation can be used to identify fetuses with a poor prognosis [4].

Therefore, it can be observed that several factors described in the specialized literature were consistent with what was found in the present case. Even though some important points could not be applied, it became evident that early diagnosis is essential for a favorable outcome and increased survival of fetuses with such malformation.

CONCLUSION

This case report highlights the severity of this fetal malformation, which should be diagnosed early, aiming for individualized prenatal care with the support of a tertiary hospital due to its potential lethality. It is also essential to provide appropriate assistance to the pregnant woman, offering counseling and medical interventions to minimize both her physical and mental distress.

The fetal tumor in this reported case could be classified as type I or II, given its large volume in the sacral region, likely greater than 10cm, and its highly vascular nature, which contributes to increased severity and higher perinatal mortality, as detailed in the outcome of this case.

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