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SCIENTIFIC ARTICLE - CASE REPORT

FETAL INTRAPERICARDIAL TERATOMA: A CASE REPORT

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ABSTRACT

INTRODUCTION: Primary cardiac tumors, including teratomas, are rare and usually diagnosed either intrauterine or postnatally. Intrapericardial teratoma is the second most common benign cardiac tumor in fetuses and can cause complications such as cardiac tamponade and fetal hydrops, leading to death. This report describes a rare case of fetal intrapericardial teratoma detected by ultrasound in a high-risk maternity unit.

CASE REPORT: A 32-year-old pregnant woman was diagnosed with a 32mm mediastinal mass, associated with pericardial effusion, mediastinal shift, and risk of hydrops. The patient was referred to another hospital for pericardiocentesis and tumor sclerosis procedures. Despite these attempts, the fetus developed hydrops and, after delivery by cesarean section, died due to pulmonary hypoplasia.

DISCUSSION: Early fetal diagnosis of cardiac tumors has become more accurate with advancements in imaging techniques. Intrapericardial teratomas are rare tumors that can lead to cardiac compression and fetal death. Early diagnosis is crucial for proper management, allowing interventions such as pericardiocentesis or surgical resection. Fetal surgery is a therapeutic option when available and performed before hydrops develops.

CONCLUSION: Early management and fetal surgery could have changed the prognosis in this case. Prompt referral to specialized centers is essential to improve perinatal outcomes.

KEYWORDS: INTRAPERICARDIAL TERATOMA, FETAL MEDICINE, FETAL CARDIAC TUMOR, PERICARDIOCENTESIS, HYDROPS, FETAL SURGERY.

INTRODUCTION

Primary cardiac tumors are rare diagnoses, typically made intrauterine or during the postnatal period. According to Tagliati et al.[1], these tumors are diagnosed in childhood or the fetal stage as multilocular intrapericardial lesions with cystic and solid components, typically located near the root of the pulmonary artery or aorta. According to Camargo et al.[2], they have an incidence of 0.009% in low- and high-risk ultrasound screenings. Approximately 90% of these tumors are benign and fall into five histological types: rhabdomyomas, teratomas, fibromas, hemangiomas, and hamartomas[2]. Rhabdomyoma is the most common among the benign histological types, accounting for 60-86% of cases, while teratoma is the second most frequent. Teratoma is a rare, histologically complex tumor of embryonic origin, composed of germ cell lineages from the endoderm, mesoderm, and ectoderm. Ultrasound and fetal echocardiography, according to Desmond et al.[3], frequently detect these pathologies. In the case of intrapericardial teratoma, this tumor mass, along with pericardial effusion, may cause cardiac tamponade, which, if relieved, can be life-saving for the fetus. The aim of this report is to describe a suspected case of this rare fetal pathology detected by obstetric ultrasound in a high-risk maternity hospital in Goiânia, Goiás, as well as to discuss the challenges and events related to the case.

CASE REPORT

The State Women's Hospital of Goiás (Hospital Estadual da Mulher de Goiás - HEMU) is a public hospital located in Goiânia (GO) that handles a significant number of fetal medicine cases from across the state of Goiás and is a national reference center in Brazil for surgery to correct imperfect twinning. In 20 years, this is the first suspected case of fetal intrapericardial teratoma.

The case involves a 32-year-old pregnant woman, in her second pregnancy, with a history of a first-trimester miscarriage 10 years prior, overweight, with no laboratory abnormalities or addictions. She was admitted at 23 weeks and 4 days to the High-Risk Obstetrics Department of HEMU with an external obstetric ultrasound describing a solid-cystic formation in close contact with the right side of the heart, measuring 28mm at its largest diameter, and an external echocardiogram reporting an extracardiac tumor associated with significant pericardial effusion.

The institutional ultrasound evaluation revealed a well-defined, circular, and heterogeneous mass with cystic and solid areas inside, pulsatile, predominantly adjacent to the right atrium, measuring 32mm at its largest diameter. It was associated with moderate pericardial effusion and mediastinal shift to the left (Figure 1). Based on these ultrasound characteristics, the primary hypothesis of an intrapericardial teratoma was suggested.

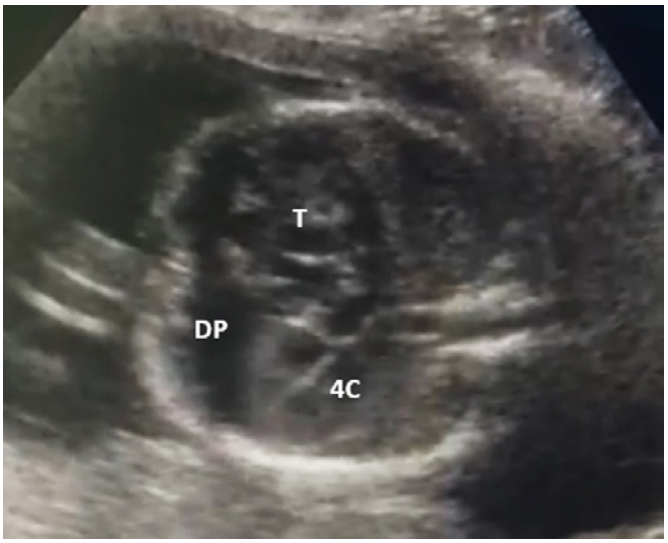
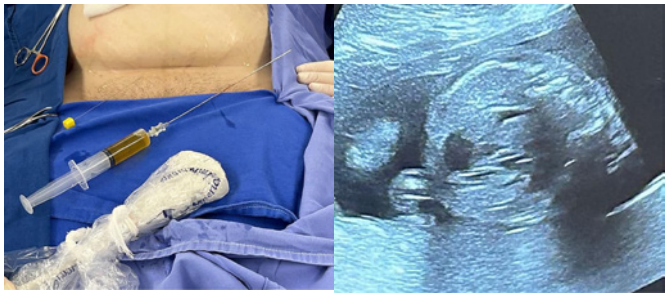


Figure 1: Transverse thoracic section demonstrating a large pericardial mass associated with significant pericardial effusion. 4C – four-chamber view; T – intrapericardial teratoma; DP – pericardial effusion.

Given the likely diagnosis, there was a need for surgical intervention to improve the fetal prognosis due to the possibility of progression in the dimensions of the mediastinal mass, leading to hydrops and fetal death. Due to the unavailability of physical resources at HEMU, the patient was referred to the Hospital de Clínicas of the Federal University of Goiás (HC-UFG) for evaluation.

During the admission examination in the Fetal Medicine department at HC-UFG, worsening of the pericardial effusion was noted, along with scalp edema. A pericardiocentesis procedure was proposed to relieve the fetal effusion and tumor sclerosis, aiming to delay the worsening of the condition and prevent fetal death before the patient could be transferred to a specialized center.

During hospitalization, the previously asymptomatic pregnant woman suddenly experienced severe abdominal pain and vomiting, leading to a diagnosis of intestinal intussusception, likely due to adhesions from a prior bariatric surgery. Consequently, an emergency laparotomy was indicated by the general surgery team, followed by pericardiocentesis to aspirate the pericardial effusion and a fine-needle aspiration of the mediastinal tumor, with sclerosis using hypertonic glucose, performed by the Fetal Medicine team at HC-UFG (Figures 2a and 2b).



Figures: 2a. Citrine fluid aspirated during pericardiocentesis; 2b. Ultrasound appearance after pericardiocentesis and tumor sclerosis.

A few days after the event, the patient was transferred to a Fetal Medicine Service in São Paulo for evaluation of mass resection. Due to the severity of the fetal condition and the immediate maternal postoperative state, the procedure was not indicated. Management continued with daily fetal echocardiograms and cardiotocography three times a day. Due to the recurrence of pericardial effusion in considerable volume, two additional pericardiocentesis were performed. However, during this time, the patient underwent a cesarean section on May 8, 2024, upon confirmation of fetal hydrops associated with cardiac arrhythmia. The neonate was born alive, weighing approximately 1500g, but progressed to death about three hours after birth, likely due to severe pulmonary hypoplasia developed as a result of the cardiac tumor and its complications. A histopathological examination of the tumor was not performed to confirm the ultrasound hypothesis.

DISCUSSION

Camargo et al.[2] reported that in the last decade, there has been a notable increase in the fetal diagnosis of primary cardiac tumors due to advancements in imaging techniques that can diagnose and classify the various histological types of the tumor with high accuracy. Rychik et al.[4] state that such tumors are primarily diagnosed in childhood or during fetal life.

In fetal life, it usually presents as pear-shaped images with a smooth and lobulated surface, heterogeneous echogenicity with cystic areas, with or without calcifications, and almost all are associated with pericardial effusion and located near the fetal right atrium, a description similar to the case in question. Increased vascularization can also be demonstrated using color Doppler. They are rarely diagnosed in adulthood, with few cases reported in the literature.

The presence of a cardiac tumor in the fetus can lead to significant hemodynamic complications that increase fetal and postnatal morbidity and mortality[2]. Garcia et al.[5] described that these tumors are associated with a severe clinical picture, where pericardial effusion combined with cardiac compression due to the mass effect can lead to progressive filling restriction, cardiac tamponade, hydrops, and fetal or neonatal death, reflecting the outcome of the case in question.

According to Garcia et al.[5], clinical management usually involves monitoring and measures for draining pericardial

fluid or delivery when the fetus reaches a viable gestational age for postnatal surgical resection. Prenatal resection, however, would be an ideal treatment option with good outcomes when performed before the onset of severe hemodynamic compromise. In the case in question, due to the non-viable gestational age and unavailability of fetal surgery, it was decided to monitor markers that anticipate intrauterine deterioration, along with pericardiocentesis and tumor sclerotherapy as a therapeutic strategy until access to definitive treatment.

According to Rychik et al.[4], in situations like this, where there is early detection of a tumor suspected to be an intrapericardial teratoma based on its appearance and location, careful and frequent monitoring is necessary to detect changes in tumor size and fetal cardiac output. The goal is to identify changes before the onset of hydrops, in order to avoid treatment in a state of severe hemodynamic instability. An increase in tumor size and abnormally low or declining cardiac output correspond to indications for treatment, thereby reinforcing the urgent nature of the presented case.

Desmond et al.[3] attributed surgical resection as the treatment of choice for cure. Rychik et al.[4] advocate for tumor resection as the most effective treatment since drainage of the cystic component and pericardiocentesis may not adequately relieve tamponade and may not inhibit the rapid growth of the tumor. Due to the progressive increase of these tumors, surgery for preterm infants at 28 weeks is acceptable using the EXIT (extrauterine intrapartum treatment) strategy or, when available, fetal surgery is possible with considerable chances of success if performed before the onset of fetal hydrops. However, given the availability of services, pericardiocentesis followed by fine needle aspiration and tumor sclerosis were the possible therapeutic alternatives to improve the fetal hemodynamic status.

Rychik et al.[4] described the first successful fetal surgery for the resection of an intrapericardial teratoma performed at 24 weeks of gestation and attributed the success of the intervention to early detection and intervention before fetal hydrops. Few studies evaluate the perinatal and long-term outcomes of fetuses with cardiac tumors. According to the cohort by Camargo et al.[2] conducted in 1991 and 2021 at two reference centers in fetal echocardiography, of the four cases of intrapericardial teratoma, three resulted in death, and one remains alive and asymptomatic after tumor excision.

CONCLUSION

Although the postnatal prognosis for newborns undergoing tumor resection is promising, prenatal follow-up presents a challenge. In the case at hand, the fetal intrapericardial teratoma could have been successfully treated with serial monitoring from its detection at an early gestational age and employing effective treatment as soon as signs of imminent fetal hydrops were identified. Therefore, it is crucial to refer patients to a multidisciplinary fetal therapy center at the moment of this diagnosis or upon the first findings predicting hemodynamic decompensation. If referred promptly, within the window of opportunity, it would have been possible to consider fetal surgery with good perinatal outcomes, thus altering the prognosis of this case that presented with a considerable degree of severity, hindering the use of therapeutic options with the best scientific evidence.

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