Fetal Intrapericardial Teratoma: Diagnostic and Therapeutic Challenges

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Introduction

Primary cardiac tumors are rare in all age groups, with an incidence of 0.002% to 0.3% in autopsy series.[1] During the prenatal period, cardiac tumors are highly uncommon and may be incidental findings in routine ultrasound assessment, which has become more frequent with the widespread use of imaging methods in the last decades. Fetal echocardiography is the main tool for detailed diagnosis of cardiac anatomy and function from the end of the first trimester until term.[2, 3] During fetal life, the most frequently found tumors are benign primary tumors, which include rhabdomyomas (the most common), as well as teratomas, fibromas, hemangiomas, and myxomas.[4] The finding of malignant primary tumors (for example, rhabdomyosarcoma and fibrosarcoma) or metastatic tumors is exceptionally rare. In fetuses, cardiac tumors can be asymptomatic or lead to complications, such as arrhythmias, hydrops, obstruction of ventricular inflow or outflow, heart failure, and death.[5]

Teratomas, although they are usually benign, can have embryonic origin in one or more of the 3 germ layers, resulting in complex histology, with the possibility of simultaneous areas of mature and immature tissue.[6] Intrapericardial teratomas are tumors with a low incidence, but they frequently invade the mediastinum and compress adjacent structures, leading to death.[4] We report a case of intrapericardial teratoma diagnosed in a fetus at 22 weeks of gestation.

Case report

We report the case of a 29-year-old, primiparous pregnant woman, without complications, referred for fetal echocardiography due to the finding of pericardial effusion on a routine morphological obstetric ultrasound. Fetal Doppler echocardiogram was performed at 22 weeks, showing normal biventricular cardiac anatomy and function. Moderate pericardial effusion was observed, containing a heterogeneous mass, with irregular contours, adjacent to the right chambers (8.4 × 7.2 mm), without causing compression of the chambers or alteration of intracavitary blood flow (Figure 1). Additionally, atrioventricular valve, ductus venosus, and umbilical artery flows were normal. The diagnostic hypothesis of pericardial teratoma was suggested. As the fetus was hemodynamically stable, serial monitoring with fetal echocardiography was chosen. Reassessments were carried out every 2 weeks, and there was no worsening of intracardiac and fetal flows up to 32 weeks of gestation. At that time, the pregnant patient had preeclampsia refractory to clinical treatment, requiring interruption of the pregnancy. Pre-delivery fetal echocardiogram showed an increase in the size of the tumor mass (37 × 24 mm), as well as pericardial effusion, but there were no other findings compatible with hemodynamic decompensation. Operative delivery was performed, with a preterm male newborn weighing 3000 g, with Apgar score of 1 in the first minute and 7 in the fifth minute, progressing with the need for orotracheal intubation and mechanical ventilation. The transthoracic Doppler echocardiogram performed during the postnatal period revealed a large heterogeneous tumor mass and the presence of significant pericardial effusion, without signs of tamponade. On the sixth day of life, the newborn underwent thoracotomy for surgical resection of the mass. During surgery, a large mass with irregular contours and a lobulated appearance was located (Figure 2). It was attached to the ascending aorta, to the right of the heart. Macroscopy showed a lobulated, granular appearance, weighing 58 g, with cystic formations when cutting the tumor. Microscopy concluded that it was a mixed mature and immature teratoma, with a resection margin affected by neoplastic cells. The newborn was referred for chemotherapy with the onco-hematology team.

Discussion

Teratomas are rare tumors (approximately 1 per 40,000 live births per year) originating from germ cells (endodermal, mesodermal, and neuroectodermal), and they may contain tissue from one or more layers.[6] They are most frequently located in the gonads, sacrococcygeal region, and mediastinum.[7] Intrapericardial teratoma accounts for less than 2% of all cardiac tumors in childhood, and can be classified as mature (benign) or immature (malignant).[4] Intrapericardial teratoma generally originates from the base of the heart, on the right side of the pericardium, attached to

Keywords

Echocardiography; Prenatal Diagnosis; Pericardial Effusion; Teratoma

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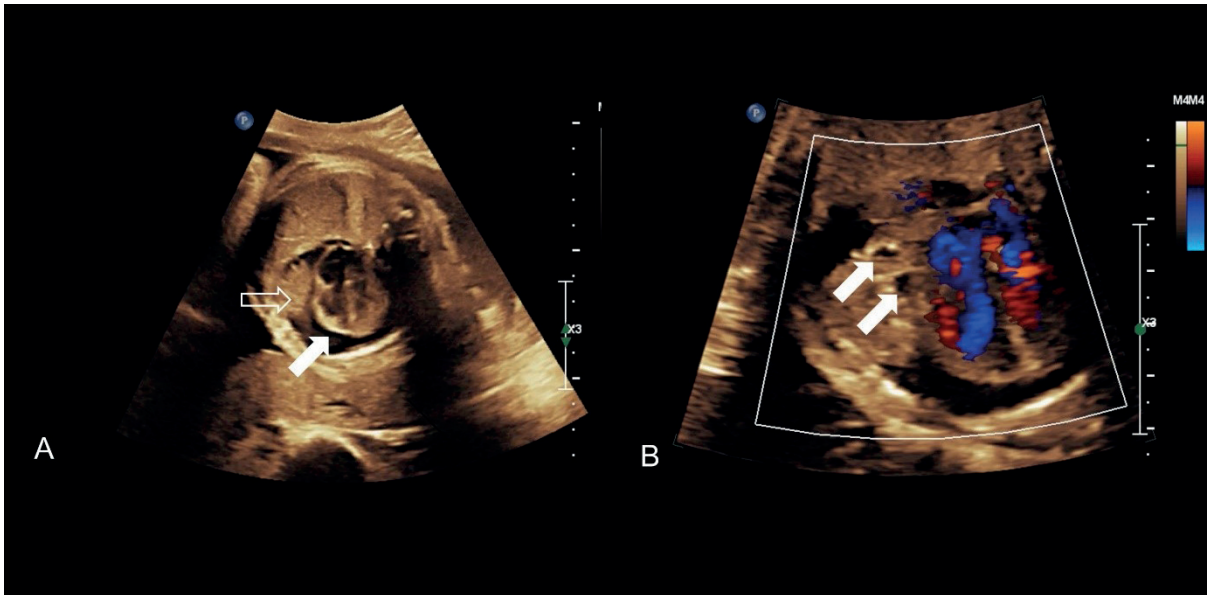


Figure 1 – *A) Fetal echocardiogram in the 4-chamber view showing circumferential pericardial effusion (solid arrow) and mass adjacent to the right atrium and ventricle (open arrow); B) Tumor mass adjacent to the right atrium and ventricle, with a heterogeneous appearance, containing hypoechoic formations inside (arrows), and no detectable blood flow.*

the aorta or pulmonary trunk. Pericardial effusion is invariably present, and the tumor mass can eventually compress the heart and great vessels.[7] In the clinical course, hydrops fetalis (a major prognostic marker), polyhydramnios, and postnatal compression of the trachea and respiratory tree may occur.[5,7] The case described, similar to the experience accumulated in the literature, was diagnosed based on an incidental finding of pericardial effusion on routine obstetric ultrasound. The tumor was attached to the aorta, on the right side of the pericardium, and was accompanied by moderate pericardial effusion, which increased during serial follow-up with fetal echocardiography. Unlike the usually reported clinical evolution, in this case, there was no appearance of hydrops or compression of anatomical structures.

In cases where the tumor and/or voluminous pericardial effusion generate compressive phenomena, prenatal pericardiocentesis can be performed to improve the fetal hemodynamic condition and enable lung growth. Pericardial effusion can recur after puncture.[7] In specialized centers, when available, other invasive fetal interventions can be considered, for example, pericardial-amniotic shunt, thoracentesis, or even open fetal surgery.[8] In the case in question, we opted for serial follow-up, without invasive fetal procedures, because, notwithstanding the increase in mass size and pericardial effusion, the fetus did not show hemodynamic impairment. Follow-up with serial fetal echocardiography showed that biventricular filling, umbilical artery and vein, and ductus venosus flows remained normal throughout the pregnancy, which was interrupted due to an obstetric condition.

Postnatal complications of intrapericardial teratoma, such as respiratory failure and cardiac tamponade, are common. Early surgical procedure can be curative, but perinatal mortality is still high.[8]

Conclusion

Fetal intrapericardial teratoma is a very rare condition that poses major diagnostic and therapeutic challenges. Early diagnosis and serial follow-up are crucial to appropriate

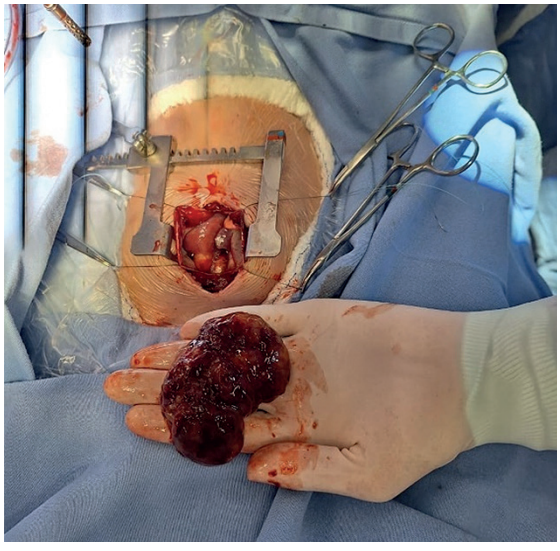
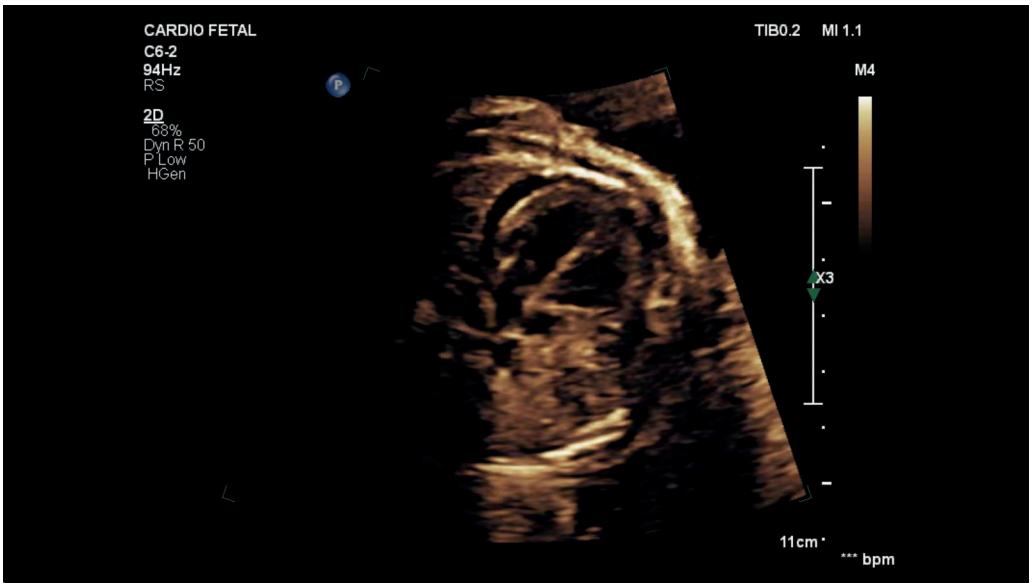
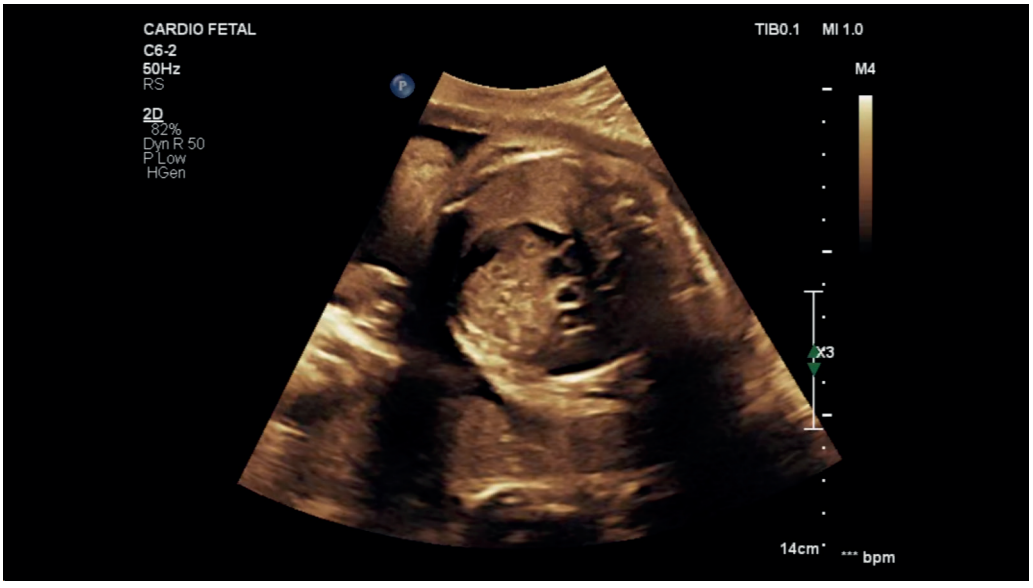


Figure 2 – *Sternotomy and macroscopic sample of the resected tumor, with an irregular and lobulated appearance.*



Video 1 – *Fetus at 26 weeks of gestation, 4-chamber view showing moderate pericardial effusion containing heterogeneous mass adjacent to the right chambers. Link: http://abcimaging.org/supplementary-material/2024/3703/2024-0065_RC_Video_1_1.mp4*



Video 2 – *Fetus at 29 weeks of gestation, 4-chamber view showing increased pericardial effusion and mass size. Link: http://abcimaging.org/supplementary-material/2024/3703/2024-0065_RC_Video_2_1.mp4.*

management. It is fundamental to differentiate hemodynamically unstable fetuses, who are candidates for intrauterine treatment, from those for whom invasive intervention is unnecessary, making it possible to wait for term to perform surgery. Intrapericardial teratoma can have a fatal outcome, but postnatal surgical resection, although complex, can be curative or offer an opportunity to combine with adjuvant therapy (chemotherapy) in immature tumors. This case highlights the importance of multidisciplinary and individualized approaches to improve outcomes in patients with this very rare condition.

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Author Contributions

Conception and design of the research and critical revision of the manuscript for intellectual content: Barberato MFA, Barberato SH; acquisition of data: Barberato MFA; analysis and interpretation of the data and writing of the manuscript: Barberato MFA, Barberato RFA, Barberato FFA, Barberato SH.

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This article does not contain any studies with human participants or animals performed by any of the authors.

References

1. Barberato SHB, Barbosa-Ferreira JM, Fernandes F. Tumores Cardíacos. In: Paola AAV, Barbosa MM, Guimarães JI, editors. Livro-Texto da Sociedade Brasileira de Cardiologia 2015. Barueri: Manole; 2015. p. 1152-56.
2. Pedra SRFF, Zielinsky P, Binotto CN, Martins CN, Fonseca ESVBD, Guimarães ICB, et al. Brazilian Fetal Cardiology Guidelines - 2019. Arq Bras Cardiol. 2019;112(5):600-48. doi: 10.5935/abc.20190075.
3. Morhy SS, Barberato SH, Lianza AC, Soares AM, Leal GN, Rivera IR, et al. Position Statement on Indications for Echocardiography in Fetal and Pediatric Cardiology and Congenital Heart Disease of the Adult - 2020. Arq Bras Cardiol. 2020;115(5):987-1005. doi: 10.36660/abc.20201122.
4. Yuan SM. Fetal Primary Cardiac Tumors During Perinatal Period. Pediatr Neonatol. 2017;58(3):205-10. doi: 10.1016/j.pedneo.2016.07.004.
5. Desmond A, Satou G, Garg M, Kallapur S, Horenstein J, Goldstein J, et al. Fetal Pericardial Teratoma: Perinatal Management and Example of Preterm Cesarean Birth to Resection. JACC Case Rep. 2023;29(3):102169. doi: 10.1016/j.jaccas.2023.102169.
6. Heerema-McKenney A, Harrison MR, Bratton B, Farrell J, Zaloudek C. Congenital Teratoma: A Clinicopathologic Study of 22 Fetal and Neonatal Tumors. Am J Surg Pathol. 2005;29(1):29-38. doi: 10.1097/01.pas.0000146006.46468.ef.
7. Bader R, Hornberger LK, Nijmeh LJ, Al-Kazaleh F, Ryan G, Toi A, et al. Fetal Pericardial Teratoma: Presentation of Two Cases and Review of Literature. Am J Perinatol. 2006;23(1):53-8. doi: 10.1055/s-2005-923433.
8. Nassr AA, Shazly SA, Morris SA, Ayres N, Espinoza J, Erfani H, et al. Prenatal Management of Fetal Intrapericardial Teratoma: A Systematic Review. Prenat Diagn. 2017;37(9):849-63. doi: 10.1002/pd.5113.