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Case Report

Prenatally Diagnosed Malignant Intrapericardial Teratoma with Yolk Sac Element: Importance of Early Diagnosis and Its Impact on Patient Management

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Abstract

Primary cardiac tumors are rare, less than 0.2% of all childhood tumors. They can be diagnosed prenatally. Although they are usually benign, they can be fatal because of the mass effects. We present a case of a prenatally diagnosed cardiac teratoma in a female fetus at 29 weeks gestation. The mass was large and the fetus showed hydrops. She was delivered at 32 weeks and underwent emergency surgery. Histologically, the tumor showed malignant yolk sac elements. One year follow up showed no recurrence. This case represents the importance of prenatal diagnosis and a multi-disciplinary approach to ensure favorable outcome.

Introduction

Fetal primary cardiac tumors are rare and usually benign, yet they often result in serious and potentially life threatening complications. Pericardial teratomas can impede fetal heart functioning due to their mass effects and increase the risk of developing fetal hydrops. Traditionally, sonography and echocardiography are used as the primary diagnostic tools, although fetal MRI has many advantages. Treatments for fetal pericardial teratomas focus on managing hydrops and delaying delivery to allow the lungs to fully mature. Early diagnosis is essential to proper case management. We present a case report on the diagnostic procedures and treatment for a female fetus with a pericardial teratoma with yolk sac elements. Furthermore, we discuss the importance of early diagnosis and a multi-disciplinary treatment approach.

Clinical History

A baby girl born at 32 weeks 2/7 days to a 22-year-old primigravida mother was transferred to our institution. She was diagnosed with an intrapericardial teratoma on antenatal ultrasounds at 29 weeks. The mass measured 48x32mm with moderate pleural effusion and no tamponade, atrial collapse, or ventricular dysfunction. The mass had increased in size since an ultrasound conducted 2 weeks earlier. The mother was then referred for cardiac echo of the fetus and fetal MRI. The baby was born via Caesarian Section. Her APGAR scores were 2 and 8 at 1 and 5 minutes respectively, and she was cyanotic at emergence with bradycardia and feeble respiratory efforts. She was emergently intubated, given a dose of surfactant, and transferred to our Cardiac Intensive Care Unit (CICU). There is no significant maternal history except for a leaky mitral valve.

Precordial exam revealed a systolic murmur of 2/6 at the left sternal border. The heart sounds were muffled throughout the precordium.

AFP and B-hcG were 140,000 and 12.21 respectively. Her other blood work revealed Hb: 11.2, Hct: 33, Na: 140, K: 3.4, iCa: 4.7, and Lactic acid of 1.9.

Prenatal fetal MRI at 29 weeks gestation demonstrated a heterogeneously T2 hyper intense multiseptated mass in the right mediastinum which measured approximately 3.2 x 4.4 x 2.7 cm (Figure 1). The mass appeared to arise from the anterior mediastinum with mass effect on the right hilum, which was pushed posteriorly and caused impingement upon the right atrium. Significant mass effect on the heart displaced the heart and mediastinum to the left (Figure 2). Subsequent fetal echocardiography and ultrasound revealed progression of the mass and development of ascites and hydrops. Therefore, the baby was delivered at 32 weeks (Figure 3). Postnatally, a CT scan showed marked dilation of the pericardial space due to a large pericardial effusion and a low attenuating and faintly enhancing mass with an epicenter in the right anterosuperior mediastinum. The mass was septated and measured 5.6 x 4.7 x 4.4cm (Figure 4). There were variable degrees

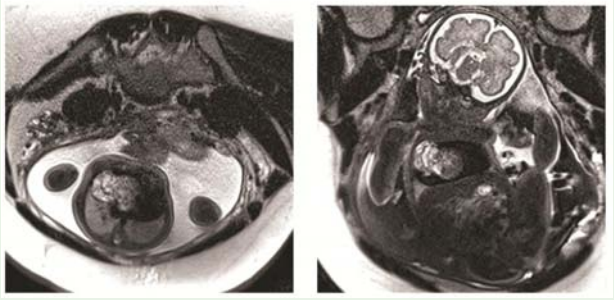

Figure 1: Fetal MR images at 29 weeks gestation. A) axial and B) coronal T2 weighted images oriented to the fetus showing a large T2 hyperintense mass with septations impinging upon the right atrium. The hear appears enlarged. No pleural effusion is identified.

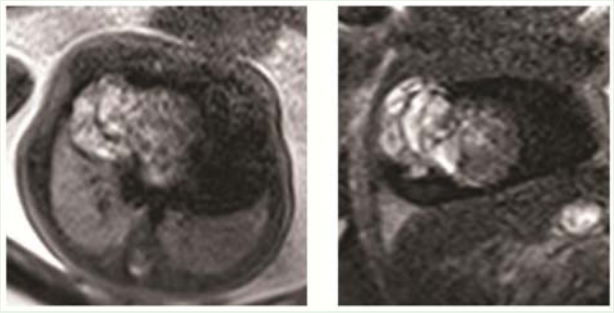

Figure 2: Cropped images of the fetal chest showing the large size of the septated mass with mass effect on the heart, filling the anterior chest compartment.

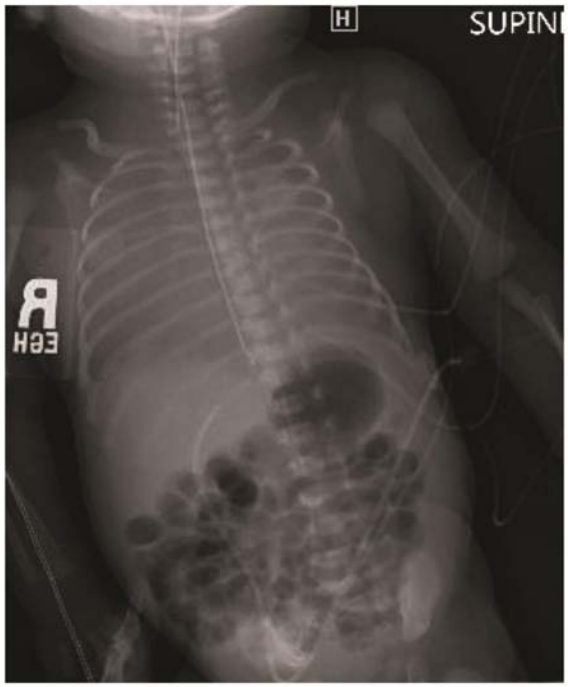

Figure 3: Immediate post-natal radiograph showing opacification of the lungs secondary to the large mass as well as pleural and pericardial effusions seen at ultrasound.

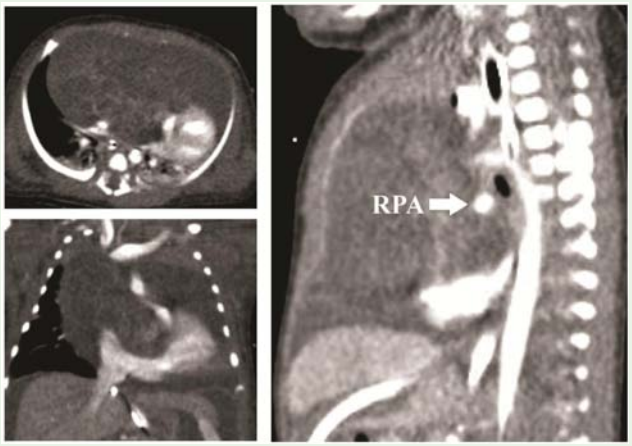

Figure 4: Post-natal CTA of the chest. A) Axial image with large hypodense mass and moderately large pericardial effusion and displacement of the heart to the left and posterior. B) Coronal image shows impingement and mass effect on the right atrium. C) Sagittal image showing the mass partially encasing the right main pulmonary artery and with mass effect, posteriorly displacing the aorta and heart.

of compressive atelectasis in all 5 lung lobes, and no pleural effusion was seen. The mass was displacing the thymus to the left, causing posterior displacement and rotation of the heart and compression of the chambers, particularly the right atrium. It was abutting the descending and transverse thoracic aortic segments. There was some narrowing near the origin of the left pulmonary artery, probably due to an acute angle caused by the mass. The SVC was displaced posteriorly with the central most part compressed prior to entering right atrium. Congestive changes in the liver and moderate free fluid in the upper abdominal peritoneal space were consistent with impaired venous return to the heart.

She underwent emergency surgery. The operation revealed a large tumor arising from the pericardium. Sharp and blunt dissection was performed. The tumor was extremely adherent to the aorta, and the aortic adventitia was inadvertently removed in some areas. Bilateral mediastinal and pleural chest tubes, a peritoneal drain, and 2 intracardiac lines were placed.

Pathological examination of the mass revealed an immature teratoma with yolk sac elements. One year later, the patient has no recurrence and no additional complications.

Discussion

Primary cardiac tumors are extremely rare, especially in fetuses, with an incidence of approximately 0.14% [1]. The majority are benign teratomas arising from the pericardium and rarely from the heart [1,2]. Intracardiac tumors generally originate in the atrial or ventricular walls [2]. The size of intracardiac teratomas can range from 2-9 cm, while intrapericardial teratomas have been reported up to 15 cm in diameter [1]. The most common fetal intracardiac tumor is rhabdomyoma, accounting for over 60% of benign tumors, followed by fibromas, myxoma, hemangioma, and teratoma [1-3].

Grossly, teratomas appear lobulated due to their cystic nature [2]. Cross-sectional examination reveals a cystic mass with intervening solid areas [2]. Histologically, teratomas contain immature elements

from multiple, most often all, germ layers [1-3]. The denser tumor regions contain mature and immature tissues, including neural, pancreatic, thyroid, smooth muscle, skeletal muscle, cartilage, and bone [1,2].

Approximately 65 cases have been reported in literature since 1890 [3]. Intracardiac and pericardial teratoma scan be fatal despite their benign nature [3]. When fetal cardiac tumors grow they can cause pericardial effusion, respiratory distress, cardiomegaly, congestive heart failure, cardiac murmurs and cyanosis [1,2]. Pericardial effusion develops in 92.7% of cases and cardiac tamponade in approximately 75% of cases [2-4].

Fetal cardiac teratomas are most commonly diagnosed antenatally by two-dimensional echocardiography and ultrasound [1-3]. Fetal MRI is useful to evaluate the extent of the tumor and its effect on the fetus as well as to assess more globally the health of the unborn baby [4-6]. In a study evaluating the effectiveness of MRI as a diagnostic tool for cardiac tumors in children, 97% of diagnoses made by evaluating MRI images independent of the pathology report were made correctly. Of those, 55% of the cases had a single correct diagnosis, and 42% had the correct diagnosis as an element of the differential. The 3% of incorrectly diagnosed cases were reported as atypical on cardiac MRI images. The most important factor in accurate diagnosis from MRI was a comprehensive exam, including all key imaging sequences [5]. Therefore, a complete MRI exam can be a valuable diagnostic tool for cardiac tumors. Cardiac teratomas are distinguished by their multiseptated appearance, larger size, and location, which is frequently on the right anterior heart border [3]. Pericardial effusion is also indicative of cardiac teratoma [3]. Most fetal cardiac tumors can be detected late in the second and throughout the third trimester [3]. Cardiac teratomas can also be postnatally diagnosed by sonography [2].

Treatment focuses on preventing premature delivery without risking fetal death [3]. The current practices for tumor management depend on the presence or absence of hydrops [3]. Induced labor and postnatal surgical intervention rely on the degree of hydrops development and maturity of the lungs [3]. In cases of large pericardial effusion and to prevent cardiac tamponade, pericardiocentesis can be attempted [3,7,8]. The first successful report of fetal pericardiocentesis was by Benater et al. in 1992 [8]. Fetal surgery carries low success rates [4].

Occasionally recurrence after tumor resection or malignant differentiation occurs [1]. When compared to other cardiac tumors of fetuses and newborns, cardiac teratomas have the second highest survival rate after hemangiomas [2]. Devileger et al. reported that the absence of fetal hydrops was associated with near term delivery and good prognosis [4]. The development of tamponade or hydrops significantly reduces the likelihood of a positive prognosis [2,3].

Our case is significant due to the malignant nature of the intrapericardial teratoma. The post surgical pathological examination revealed an immature teratoma with yolk sac elements. Foci of yolk sac elements have previously been associated with tumor recurrence [7]. Elevated α-fetoprotein (AFP) levels up to 100,000 are within normal limits for a newborn. However our patient showed 140,000 AFP, which raises suspicion for a malignant component, which was confirmed by histological examination. Follow up of our patient one-year after resection shows no recurrence.

Prenatal ultrasound is a diagnostic screening tool that can clearly show the boundaries of the tumor, the extent into chest cavity, and assess for development of hydrops. Dedicated fetal echocardiography will allow noninvasive monitoring of cardiac functioning [8]. However, fetal MRI is an extremely useful tool to better assess the origin of these masses and provide characterization of cardiac masses without the necessity of contrast or exposure to ionizing radiation. In addition, fetal MRI will provide a complete picture of the entire fetus and placenta to screen for additional abnormalities. Postnatal CT and MRI may be recommended to determine the full extent of the tumor, mark clear boundaries with the use of intravenous contrast, and measure the extent of its effect [8].

Conclusion

The prenatal diagnosis and monitoring of cardiac tumors, specifically intrapericardial teratoma, is essential to ensure the best possible outcome of this rare condition. In our case, even with a malignant component, the outcome was good. Prenatal ultrasound and fetal echocardiography are the primary modalities for screening and follow up of cardiac masses. Fetal MRI may be indicated in cases where further characterization and definitive site of origin are required. Fetal MRI may provide additional information about other anomalies that may not have been seen at ultrasound. Therefore, early diagnosis and intervention can optimize the outcome for infants with this unusual, potentially lethal tumor.

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