Intrapericardial Teratoma : fetal diagnosis and neonatal management

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ABSTRACT

Intrapericardial teratoma is a rare primary cardiac tumor arinsing from all three germinal layers and usually diagnosed in the featal or neonatal period. They are mostly benign, but life-threating complications have been reported such as hydrops fetalis, cardiac failure and cardiac tamponnade. We report a case of intrapericardial teratoma diagnosed in prenatal life and operated in neonatal period with success.

Keywords : teratoma, fetus, ultrasound, computerized tomography, surgical excision

INTRODUCTION :

Intrapericardial teratoma is a rare form of primary cardiac tumor occurring in 5 to 6 of 10,000 newborns [1]. Less than 100 cases have been reported in the literature [2] since the first case report in 1890 [3]. Although benign, they may cause large pericardial effusion/tamponade and cardiac compression [4]. With prenatal ultrasound, early diagnosis can be accomplished before the onset of symptoms. It permits close prenatal monitoring and post-partum surgical planning. Complete surgical resection is the treatment of choice [5]. Long-term follow-up is required for possible recurrence especially when the tumor is immature or when resection is incomplete [6].

We report a case of intrapericardial teratoma diagnosed in prenatal life and operated in neonatal period with success.

CASE REPORT :

A male fetus, coming from a spontaneous pregnancy followed by a primary care obstetrician, was diagnosed to have a mediastinal mass on ultrasound at 26 weeks' gestation. He was the second child of a non- consanguineous young couple and his mother was 33-year-old, previously healthy and without dysgravidia. Fetal echocardiography showed a heterogeneous multi-lobed mass, measuring 28 x 22 mm and compressing the right atrium with moderate pericardial effusion. Fetal cardiac magnetic resonance imaging showed a large mediastinal mass of heterogeneous signal with an intimate contact with the right heart cavities.

The mother was then referred to a tertiary level maternity and an ultrasound follow-up every 15 days was indicated. No other structural abnormalities in the heart or in the body were detected. Delivery was then planned at term. However, at 35 weeks' gestation, the mass increased in size (41X34mm) with appearance of a pericardial effusion of average abundance. Delivery was then anticipated after a course for fetal lung maturation.

It was a caesarean birth of a boy, clinically term, eutrophic with good adaptation to the extra-uterine life. He had transient respiratory distress without signs of heart failure.

On Chest X ray, lungs were not seen and a thoracic opacity of cardiac tissue tone occupies the thorax.

Computed tomography (CT) showed a large anterior mediastinal mass with predominant cystic component measuring 53 X 48 X 52 mm.

Postnatal echocardiography showed a huge pericardial tumor measuring 48mm × 4mm compressing the upper and lateral face of the right atrium and the left pulmonary artery with the presence of a circumferential pericardial effusion. Its multi-lobed cystic appearance evokes a teratoma. There were no hemodynamic signs of compression of the right cavities. Alpha foeto protein returned very high (111 335.50 IU / mL).

The baby boy was clinically well and the search for another tumor localization came back negative. Surgical excision of the mass was done through a median sternotomy and cardiopulmonary bypass. A moderate pericardial

effusion was drained and complete surgical resection of the tumor was done successfully. The histopathological examination confirmed the diagnosis of benign and mature teratoma.
The post-operative echocardiogram showed no residual mass and good biventricular function.
At one year follow –up, the Child was asymptomatic with no residual mass at echocardiography.

DISCUSSION :

Teratomas are tumors originating from endodermic, mesodermic or neuroectodermal germinal layers: Endodermal components are represented by gastric or intestinal tissue, ectodermal components are neuroglial or neuroendothelial tissue and mesodermal components are bone, muscle, cartilaginous or fatty tissue[7]. The tumor mass may demonstrate compression of the right atrium and/or ventricle, creating hemodynamic compromise, ultimately resulting in hydrops, pericardial effusion [8] and even heart failure [9]. It usually appears as a cystic mass outside the cardiac cavities and is sometimes calcified [10]. These tumors are voluminous and sometimes larger than the heart [11] the majority are located anteriorly to the right side, where they may compress the heart, great vessels, and in some cases the trachea [12]. Echocardiography, computerized tomography (CT) and magnetic resonance imaging (MRI) of the heart are the main non-invasive tools to detect the mass and identify its borders [13,14]. They support the diagnosis if the characteristic features of fat density tissue in combination with calcifications are found in the mass. The findings of a large pericardial effusion or pericardial thickening may suggest teratoma rupture [15]. The differential diagnosis includesa variety of cardiac tumors to include fibromas, hemangiomas, and rhabdomyomas [16].
The management is based on gestation at the time of diagnosis and presence of hydrops fetalis, characterized by fetal anasarca. If hydrops fetalis is absent, the patient should be carried to term, with early neonatal tumor resection. Surgical excision of the tumor without cardiopulmonary bypass can be performed. If hydrops develops during the pregnancy, which is a poor prognostic factor, antenatal pericardiocentesis is the treatment of choice to remove the pericardial effusion and reduce cardiac obstruction [17, 18]. Pericardiocentesis is a temporizing strategy to allow progression of fetal development and lung maturation before delivery [19]. Complete surgical resection is the treatment of choice and early resection immediately after diagnosis improves the outcome [20].
Long-term follow-up is required especially when the tumor is immature or when resection is incomplete.

CONCLUSION :

Intrapericardial teratoma is a rare primary cardiac benign tumor. Prenatal diagnosis, close monitoring for hydrops fetalis, antenatal pericardiocentesis for cardiac obstruction, and early neonatal tumor resection provide a good prognosis.

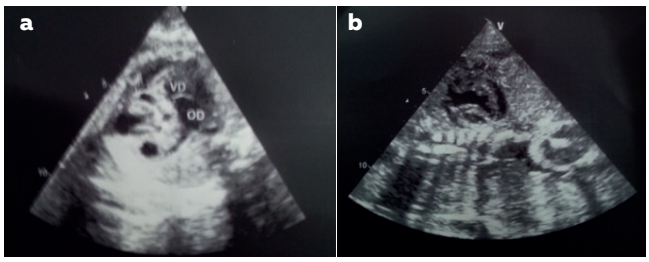

Figure 1 : Fetal echocardiography at 26 weeks' gestation showing a mediastinal mass : axial view (a) sagittal view (b).

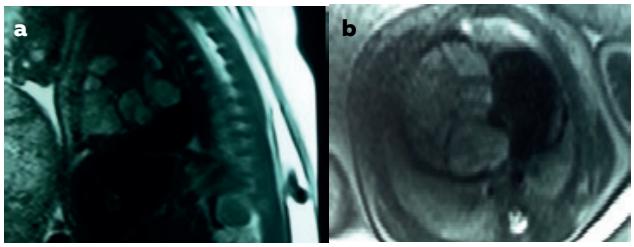

Figure 2 : Coronal view of fetal cardiac MRI showing a heterogeneous mediastinal mass(a). Sagittal view of fetal cadiac MRI showing a large mediastinal mass with an intimate contact with the right heart cavities(b)

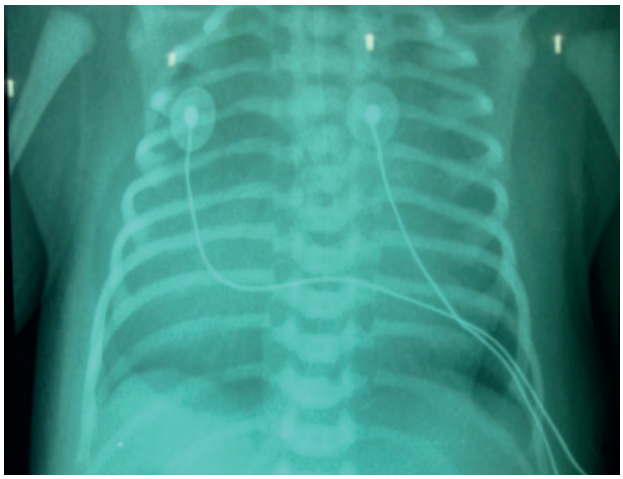

Figure 3 : Chest X ray showing a thoracic opacity with unseen lung fields

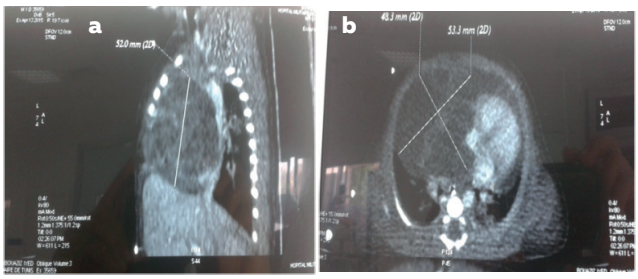

Figure 4 : a-b Axial view of thorax on CT scan showing anterior mediastinal mass with predominant cystic component (a). Sagittal view of thorax on CT scan showing a large cardiac cystic mass (b).

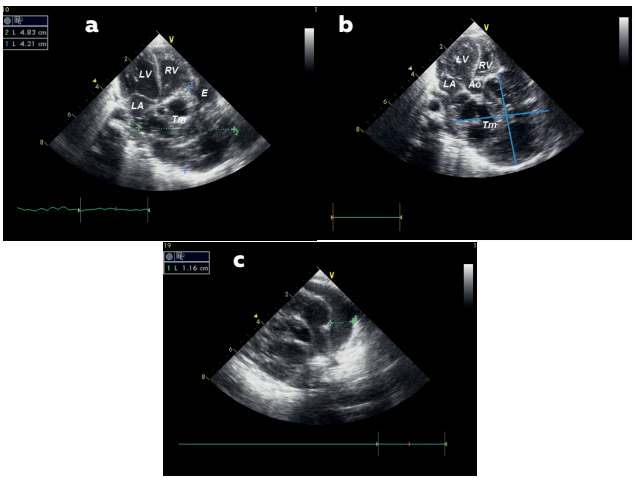

Figure 5 : Echocardiographic apical four and five-chambers views (a+b) demonstrating a heterogeneous mass (arrow) with multiple hypolucent cystic areas compressing the right atrium, with mild pericardial effusion. The tumor mass measures 4.8 cm × 4.2 cm. Short axis view (c) mild pericardial effusion regarding the unfundibular tract.

LV : Left ventricle, RV: Right ventricle, LA: Left atrium,E: pericardial effusion.

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