Case report

Intrapericardial teratoma in newborn babies

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Abstract

We report two cases of intra pericardial tumor with pericardial effusion, diagnosed in utero by echocardiography at 21 and 28 weeks of gestation. Both fetuses underwent an intra uterine pericardiocentesis to treat a hydrops fetalis. Surgical resection of the tumor was undertaken immediately after birth and histological description reported cystic teratoma. Both babies had a favorable post operative course.

Keywords: Intrapericardial teratoma; Hydrops fetalis; Pericardiocentesis; Unstable hemodynamics

1. Introduction

Intrapericardial teratoma is a rare tumor, usually benign, that often causes severe cardiovascular and respiratory distress in newborn babies and infants. Teratoma are frequently seen in the anterior mediastinum. They can be associated with non-immunologic hydrops fetalis revealed by fetal anasarca. We report two cases of hydrops fetalis diagnosed in utero by echocardiography and associated with an intrapericardial tumor. Both neonates underwent surgical excision of a large cystic lesion shortly after birth.

2. Patients

A 30-year old woman, gravida 2 para 1, was seen at 21 weeks of gestation for a routine obstetric examination. Fetal ultrasonography revealed a thoracic tumor with pericardial effusion and hydrops fetalis. The tumor appeared solid, with cystic and echogenic components (diameter 2.5 cm) and was larger than the heart. The size of the tumor and the amount of pericardial effusion increased as assessed by serial examinations. Ultrasonography at 34 weeks of gestation indicated the deterioration of hydropic condition. A pericardiocentesis was performed in utero and removed 80 ml of serous fluid. Two days later, the hydropic condition re-occurred and prompted delivery of a 2750 g baby. Transthoracic echocardiography demonstrated a 65 × 30 × 20 mm intrapericardial cystic mass with large pericardial effusion. The tumor was attached to anterior portion of the ascending aorta. The heart was otherwise normal (Fig. 1).

A 27-year old woman gravida 3 para 1 was referred to our institution for evaluation at 28 weeks of gestation, because of a possible fetal thoracic mass with non-immune hydrops fetalis. Fetal ultrasonography revealed a solid mass with cystic or echogenic components with a large pericardial and plural effusion, skin edema and ascite. Intrauterine pericardiocentesis was performed to remove 22 ml of serous fluid. Preterm labor developed at 32 weeks of gestation and was unresponsive to medical management. A 1700 g female baby was delivered by cesarean section. Two dimensional echocardiography showed a 40 × 35 × 40 mm intrapericardial cystic mass and large pericardial effusion. The tumor was attached to anterior portion of the ascending aorta and compressed the right atrium, right ventricle and superior vena cava.

Despite a stimulation of lung maturation with corticosteroids (Betamethasone, Celestene® Schering–Plough) during the last weeks of gestation, both neonates were in respiratory distress and required immediate intubation and ventilatory support with high-frequency oscillation, conventional mechanical ventilation and surfactant therapy. Both babies were operated on 3 days after delivery. After median sternotomy and pericardiectomy, the cystic mass was
dissected and completely removed (Fig. 2). Histological study of the masses revealed a variety of tissues, including three germ cell layers and immature neural elements confirming a diagnosis of benign teratoma. The immediate postoperative period was remarkable for a low arterial pressure due to depressed systemic vascular resistance requiring prolonged vasopressive support. The babies were extubated on the 4th and 10th postoperative day. Three months later the babies were well and remained symptom free with no signs of tumor recurrence.

3. Discussion

Intrapericardial teratoma is a rare often benign congenital tumor, that was first described by Joel in 1890 [1]. Before birth the diagnosis of thoracic tumor is established by fetal two-dimensional echocardiography which, besides delineating the tumor, often points out a hydrops fetalis. This non-immunologic hydrops fetalis is characterized by a fetal anasarca which can lead to massive pleural and pericardial effusion with the risk of fatal cardiac compression [2–4]. In our cases, a pericardiocentesis was performed to confirm the diagnosis of intrapericardial teratoma and prevent the development of a tamponade [3]. Suction of fetal pericardial fluid is a simple procedure that can be lifesaving. Pericardial effusion is rarely associated with another cardiac tumor [4]. Rupture of cystic areas in the pericardium due to the multicyctic nature of pericardial teratomas [4], associated with an obstruction of the systemic venous return and of the thoracic duct, interfering with pericardial lymphatic drainage leads to the development of effusions in the pleural, peritoneal, and pericardial spaces. Pericardial fluid is serous and contain immature mesothelial cells [4]. Early intra uterine fetal hydrops worsens the prognosis and makes the management of the newborn a challenge. Because a rise in cardiac compression during vaginal delivery may result in fatal chest and heart compression [5], we elected a delivery by cesarean section after appropriate maturation of the lungs.

All large intrapericardial teratomas eventually become symptomatic and usually require drastic respiratory support and early tumor removal [5,6]. Teratoma are frequently seen in the anterior mediastinum and attached to the heart or great vessels by a pedicle [6]. Nevertheless teratomatous lesions of pericardium generally arise out of the base of the heart and encircle the great vessels as was the case in our patients [7]. If the diagnosis of intrapericardial tumor with pericardial effusion can be routinely performed with two dimensionnal echocardiography, only histological description can prove the diagnosis of a germ cell tumor of which teratoma are the most common. The resection is easy and cardiopulmonary bypass is usually not required due to the lack of myocardial involvement. Multiple intrathoracic locations are rare but another location should always be searched. Both our patients had a relatively unstable hemodynamic course after surgery, a fact that has not been reported in the literature. Several hypothesis may explain this unstable period: a generalised inflammatory response, a vasodilatation following anaesthesia, a chronic tamponade in utero, or a premature baroreflex. We did not
measure pro inflammatory cytokines in blood samples and mRNA expression of pro inflammatory cytokines in the tumor. True teratoma tissue are known to express Interleukin-1, Interleukin-6 and Interleukin-8 mRNA [8].

In conclusion, this article presents two preterm infants with an intrapericardial tumor associated with hydrops fetalis. Prenatal diagnosis of the tumor is essential for planning fetal management, delivery and perinatal surgery. Unstable hemodynamics can to be seen during the postoperative period. The prognosis, however, looks excellent.

References