Mediastinal masses: a case of fetal teratoma and literature review

A. Giancotti, R. La Torre, E. Bevilacqua, V. D'Ambrosio, G. Pasquali, P. Benedetti Panici

Department of Obstetrics, Gynecology and Urologic Sciences "Sapienza" University, Umberto I Hospital, Rome (Italy)

Summary

Fetal mediastinal masses are rare congenital formations that could complicate pregnancy. They are usually discovered as space occupying lesions in the fetal chest during routine ultrasound scan. The most important prognostic factors of mediastinal masses are mass location, compressing effect causing pulmonary hypoplasia and/or heart failure, and the presence or absence of hydrops. We report a case of fetal mediastinal teratoma and a review of the literature. A 32-year-old woman carrying a fetus with hydrops due to a mediastinal mass underwent cesarean section at 32 1/7 weeks' gestation. A well encapsulated tumor was excised by surgery at one day of life. The baby is now eight months old without respiratory difficulty. To our knowledge, this is the fourth case report of a mediastinal teratoma associated with nonimmune hydrops in a fetus that survived the neonatal period. Fetal mediastinal teratoma requires close surveillance and multidisciplinary management by obstetricians, neonatologists, and pediatric surgeons.

Key words: Mediastinal masses; Fetal mediastinal teratoma; Prenatal diagnosis; Fetal management.

Introduction

Fetal mediastinal masses represent a wide diversity of disease states.

The mediastinum is demarcated by pleural cavities laterally, the thoracic inlet superiorly, and the diaphragm inferiorly. It is further compartmentalized into anterior, middle, and posterior divisions based on structural landmarks seen on the lateral radiograph. The anterior mediastinum contains the thymus, fat, and lymph nodes. The middle mediastinum contains the heart, pericardium, ascending and transverse aorta, brachiocephalic veins, trachea, bronchi, and lymph nodes. The posterior mediastinum consists of the descending thoracic aorta, esophagus, azygous vein, autonomic ganglia and nerves, thoracic lymph nodes, and fat [1]. Heart masses are not described. Teratoma and lymphangioma are the most common tumors developing in the anterior mediastinum [2]. Congenital thymic cysts are remnants of the thymopharyngeal duct [1]. Laryngeal atresia is an exceedingly rare anomaly in which the high airways are completely obstructed and the lungs appear severely enlarged and hyperechoic [2]. Fetus in fetu is a particular form of teratoma. This tumor is a parasitic twin that develops within the "main" twin [2]. Neuroblastoma can develop in the posterior mediastinum, appearing as a solid mass [3]. Cystic masses in the posterior mediastinum are most commonly esophageal duplication or neurenteric cyst (presence of both enteric and neural tissue in surgical specimens). Vertebral segmentation anomalies are commonly associated [1]. These tumors may extend both below and above the diaphragm [4]. The term congenital diaphragmatic hernia (CDH) encompasses a range of closure defects of the diaphragm with the abdominal viscera, located near the defect, migrated into the thorax [5]. Congenital intrathoracic stomach may be due to a short esophagus or hiatal hernia.

The likelihood of malignancy of mediastinal masses is influenced primary by the following factors: mass location, compressing effect causing pulmonary hypoplasia and/or heart failure, and the presence or absence of hydrops.

Ultrasound (US) assessment of the mediastinum can be carried out easily until 25-26 weeks of gestation. After this period, the increased mineralization of the ribs lead to significant acoustic shadowing, which limits the display of the intrathoracic area. However, it has to be underlined that a good number of mediastinal anomalies evolve: they can appear only in the 3rd trimester, or, on the contrary, they can regress before birth [5]. The initial workup of a suspected mediastinal mass involves obtaining the classic 4-chamber view of the fetal heart. If the results from this view are abnormal, and a mediastinal lesion is found, this should be explored further using coronal and sagittal views [5].

In the present study, we report a case of fetal mediastinal teratoma associated with nonimmune hydrops and a review of literature.

Case Report

A 32-year-old white gravida 2, para 1, mother was referred to the Centre of Prenatal Diagnosis at the Hospital Umberto I of Rome at 29 6/7 weeks of gestation for evaluation of a fetal mediastinal mass. Her previous medical and obstetric history as well as the course of her current pregnancy had been unremarkable. The mother was group O Rh (+) with a negative indirect Coombs test. On presentation, ultrasound revealed a 47 x 35 x 44 mm left anterior mediastinal mass with displacement of the heart and the left lung, and associated bilateral pleural effusions (Figure 1). The growth of the fetus was consistent with its ges-

Revised manuscript accepted for publication September 26, 2011

Table 1. — *Literature review of fetal mediastinal teratoma*.

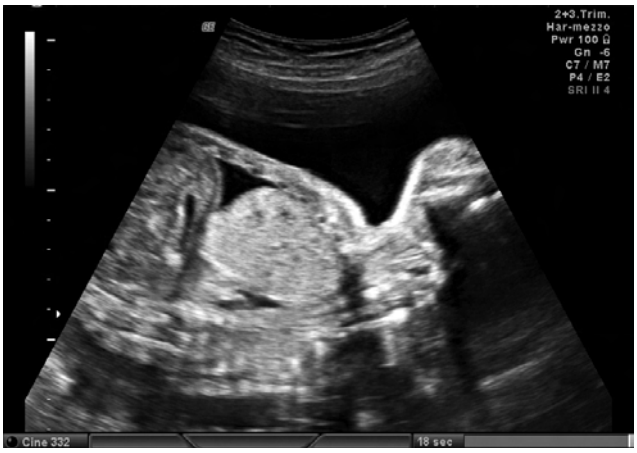
Authors and year	Outcome	Comment
Merchant *et al*. [9] 2005	Case 1: delivered at 25 weeks; alive at 9 months follow-up Case 2: delivered after 34 weeks; alive at 1 year follow-up	Hydrops fetalis; mediastinal mass found on fetal US; in utero resection at 23 weeks Mediastinal mass found on fetal US; surgery in the immediate newborn period
Kuller *et al*. [10 1991	Failed neonatal resuscitation at 27 weeks	Hydrops fetalis; diagnosis at autopsy
Schild *et al*. [11] 1998	Failed neonatal resuscitation at 27 weeks	Hydrops fetalis; mediastinal mass found on fetal US; diagnosis at autopsy
Wang *et al*. [14] 2000	Delivered at 39 weeks; alive at 3 months follow-up	Surgery at 4 days age age; diagnosis at histology
Liang *et al*. [15] 1998	Delivered at 39 weeks; alive at 5 months follow-up	Mediastinal mass found on fetal US; surgery at 7 days age; diagnosis at histology.
Weinraub *et al*. [16] 1989	Fetal demise at 29 weeks	Hydrops fetalis; mediastinal mass found on fetal US
Froberg *et al*. [17] 1994	Fetal demise at 27 weeks	Hydrops fetalis; mediastinal mass found on fetal US
Dumbell *et al*. [18] 1990	Delivered at 36 weeks; alive at 18 months follow-up	Mediastinal mass or cystic malformation of the lung found on fetal US; surgery in the immediate newborn period and at the age of 3 months
Takayasu *et al*. [19] 2010	Delivered at 39 weeks; alive at 6 months follow-up	Hydrops fetalis; mediastinal mass found on fetal US; aspiration of the fetal tumor cyst fluid at 29 weeks; surgery at 30 days of age
Noreen *et al*. [20] 2008	Case 1: fetal demise at 19 weeks Case2: stillborn at 27 weeks Case 3: fetal demise at 23 weeks	Hydrops fetalis; diagnosis at autopsy Hydrops fetalis; diagnosis at autopsy Hydrops fetalis; diagnosis at autopsy
Wesolowski *et al*. [21] 2008	Delivered at 33 weeks; died at 50 days of age	Hydrops fetalis; mediastinal mass found on neonatal US surgery at 7 days of age
Akosy *et al*. [22] 2002	Delivered at term; died at one day of life	Mediastinal mass found on fetal US; surgery in the immediate newborn period
Allman *et al*. [23] 2001	Delivered at 36 weeks; alive at 3 years follow-up	At birth mild hydrops; heart beat detected by 15 minutes of age; surgery at 3 days of age; diagnosis at histology
Present case	Delivered at 32 weeks; alive at 8 months follow-up	Hydrops fetalis; mediastinal mass found on fetal US; surgery in the immediate newborn period and at 18 days of age

tational age, with an estimated body weight of 1,480 g (between the 25th and the 50th percentile). Fetal magnetic resonance imaging (MRI) at 31 weeks of gestation confirmed a heterogenous anterior mediastinal mass suggestive of a mediastinal teratoma. There were sequelae of mediastinal compression including pleural effusions, ascites, hydrocele, and polyhydramnios. Fetal echocardiography at 31 weeks of gestation found no evidence of structural or functional abnormality. Hospitalization was performed at 32 weeks because the following US examinations showed a rapidly growing mass (56 x 42 x 52 mm) with more severe sequelae of mediastinal compression (Figure 2) and signs of hydrops: pleural effusions, ascites, hydrocele, and polyhydramnios. A massive polyhydramnios with an amniotic fluid index of 23 cm was observed and the mother began to complain of mild contractile activity. Moreover, Doppler flow studies revealed increased resistance in the umbilical arteries (pulsatility index, PI, 1.12) and in the ductus venosus (PI 0.75), presumably due to low-output heart failure. Tocolysis was administered (nifedipine 20 mg x 3 on daily) while a course of antenatal corticosteroids for fetal lung maturity was given (betamethasone 12 mg intramuscularly 24 hours apart for a total of two doses). An elective cesarean section was performed at 32 1/7 weeks' gestation. The neonate was a male and he had a birth weight of 1,950 g with Apgar scores of 1 - 3 - 6 - 8 at 1 - 5 - 10 - 20 min, respectively. At 5 min adrenaline was administered. The heart rate exceeded 100 beats per minute by 12 min of age. An initial arterial blood gas assessment showed a pH of 7.35, PCO_2 of 45 mmHg, PO_2 of 20 mmHg, bicarbonate of 22.8 mmol/l, and base deficit of 0.8 mmol/l. Surgery was performed on day 1 of life. A well encapsulated tumor was excised, complete and intact. Histological examination confirmed the diagnosis of teratoma. The baby's postoperative course was complicated by left vocal cord paralysis and left diaphragm paralysis. At 18 days of age a second surgery was performed and the left diaphragm was plicated. The baby has been followed up regularly, and he is now eight months old without respiratory difficulty.

Discussion

Teratomas are tumors composed of multiple tissue elements derived from all three germ layers. These neoplastic lesions could present various degrees of differentiation, ranging from primitive somatic elements to highly organized axial structures [6]. Whereas teratoma is the most common congenital neoplasm, it rarely occurs in the mediastinum in fetal life (4-11% of cases) [7, 8]. While US is usually diagnostic, MRI using both T1-W and T2-W sequences is helpful in determining the extent of the tumor and its content [2]. These congenital malformations could cause compression of vital structures that may result in nonimmune hydrops leading to fetal demise, late gestational polyhydramnios, preterm labor

Fig. 1



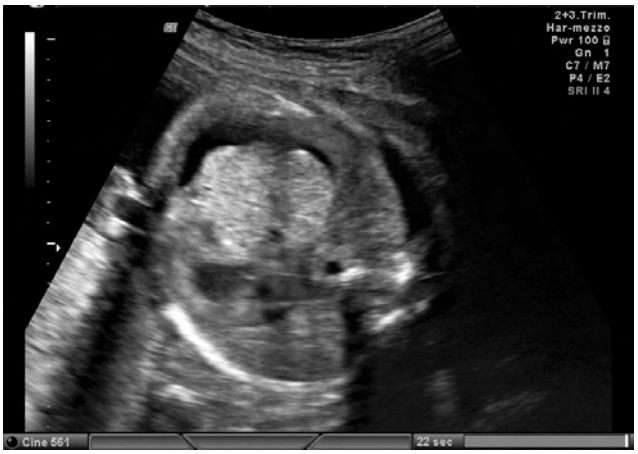


Fig. 2

Figure 1. — Sonogram showing mediastinal mass and pleural effusion.
Figure 2. — Sonogram showing the large formation occupying all the anterior mediastinum.

and respiratory distress at birth [9]. Hydrops fetalis and polyhydramnios are poor prognostic signs. The diseases considered in the differential diagnosis of such lesion are congenital cystic adenomatoid malformation, pulmonary sequestration, intrapericardial teratoma, cardiac rhabdomyosarcoma and diaphragmatic hernia [9-11]. Since each of these pathological conditions may be associated with other congenital and chromosomal abnormalities, a careful anatomic survey and chromosomal analysis are indicated [11]. In particular, karyotyping should be carried out because the prevalence of mediastinal germ cell tumor is 39 to 50 times higher in Klinefelter' syndrome than in the general male population [12, 13].

This tumor is extremely rare and there are just few reported cases of fetal mediastinal teratomas in the literature [9-11, 14-23] (Table 1).

To our knowledge, this is the fourth case report of a fetus with mediastinal teratoma associated with nonimmune hydrops that survived the neonatal period [9, 19, 21]. Merchant *et al*. proposed a management algorithm for large fetal mediastinal teratomas [9]. Management depends on fetal gestation, the presence of hydrops, and the risk of airway compromise. A fetus younger than 30 weeks of gestation with hydrops may require open fetal surgery. A non-hydropic fetus can be managed expectantly with serial US and regular echocardiographic surveillance. Fetuses older than 30 weeks of gestation must be assessed for airway compromise and lung development. In the case of airway compromise, an ex utero intrapartum therapy (EXIT) may be required [9]. Takayasu *et al*. recommend aspiration of the tumor cyst fluid as first-line therapy when the tumor is cystic in nature [19]. In the case of Wesolowski *et al*., the patient succumbed to his illness at 50 days of life [21]. Among the four case reports, Wesolowski's study is the only one with the diagnosis in the neonatal period rather than in the antenatal period. Detection of teratomas before birth with US or MRI is important because the mortality rate is three times greater with a postnatal diagnosis [24].

Liang *et al*. point out the importance of Doppler velocimetry: there may be a better postnatal outcome when the hemodynamic changes only involve the central vessels and not the peripheral vessels than in those cases in which flow in both central and peripheral vessels is decreased [15]. Wang *et al*. state that in the case of immature teratoma the correct diagnosis from the prenatal US can be difficult. Instead, the diagnosis of mediastinal teratoma is comparatively easy to make if a multilobular mass has both cystic and solid components with calcification and acoustic shadows [14].

In 2007, Grethel *et al*. examined their institutional database and looked at patients with fetal mass lesions to evaluate survival with or without intervention. The development of hydrops conferred a dismal prognosis with greater than 95% mortality. Fetal intervention reduced this mortality to 50% [25]. A 32-week gestational age cut-off point has been identified as the critical time-point before which development of hydrops may warrant prenatal intervention, and after which delivery is indicated [26, 27]. It is highlighted that before consideration of fetal intervention, a full evaluation including karyotyping, fetal echocardiography, and level III US must be carried out [25]. In our case, signs of hydrops were present around 31 and 32 weeks of gestation and an elective cesarean section was performed at 32 1/7 weeks' gestation after the administration of a course of antenatal corticosteroids for fetal lung maturity.

Obtaining a precise diagnosis has become easier with advances in prenatal imaging as well as karyotype analysis. Understanding the prognosis of the diagnosed anomaly is a more difficult task. At present it is not possible to predict which fetuses will become hydropic, and premature intervention carries unacceptable risk to those that will not progress to hydrops [25]. We still know too little about non-invasive measures to improve outcomes. The diagnosis is often done in the second or third trimester of gestation and a multidisciplinary diagnostic workup where obstetricians, neonatologists, and pediatric surgeons work closely becomes crucial.

References

[1] Duwe B.V., Sterman D.H., Musani A.I.: "Tumors of the mediastinum". *Chest,* 2005, *128,* 2893.

[2] Avni F.E., Massez A., Cassart M.: "Tumours of the fetal body: a review". *Pediatr. Radiol*., 2009, *39*, 1147.

[3] Rivasi S., Gasser B., Collina G., Massolo F., Philippe E.: "Congenital fetal neuroblastoma". *Ann. Pathol*., 2001, *21*, 76.

[4] Markert D.J., Grumback K., Haney F.J.: "Thoracoabdominal duplication cyst: prenatal and postnatal imaging". *J. Ultrasound Med*., 1996, *15,* 333.

[5] Paladini D., Volpe P.: "Ultrasound of congenital fetal anomalies". Informal Healthcare, London, 2007, 183.

[6] Tapper D., Lack E.E.: "Teratomas in infancy and childhood. A 54-year experience at the Children's Hospital Medical Center". *Ann. Surg*., 1983, *198,* 398.

[7] Ayadi-Kaddour A., Ismail O., Hassen F., Smati B., Djilani H., Kilani T., El Mezni F.: "Benign mature teratomas of the mediastinum". *Revue des Maladies Respiratoires*, 2008, *25,* 531.

[8] Allman A.W., Buss P.W., Spicer R.D., Wake A.: "Mediastinal teratoma presenting as apparent fresh stillbirth". *Arch. Dis. Child Fetal. Neonatal. Ed.* 2001, 84, F65.

[9] Merchant A.M., Hedrick H.L., Johnson M.P., Wilson R.D., Crombleholme T.M., Howell L.J. *et al*.: "Management of fetal mediastinal teratoma". *J. Pediatr. Surg*., 2005, *40,* 228.

[10] Kuller J.A., Laifer S.A., Martin J.G., MacPherson T.A., Mitre B., Hill L.M.: "Unusual presentations of fetal teratoma". *J. Perinatol.,* 1991, *11*, 294.

[11] Schild R.L., Plath H., Hofstaetter C., Hansmann M.: "Prenatal diagnosis of a fetal mediastinal teratoma". *Ultrasound Obstet. Gynecol.,* 1998, *12,* 369.

[12] Lachman M.F., Kim K., Koo B.C.: "Mediastinal teratoma associated with Klinefelter's syndrome". *Arch. Pathol. Lab. Med.,* 1986, *110*, 1067.

[13] Hasle H., Jacobsen B.B., Asschenfeldt P., Andersen K.: "Mediastinal germ cell tumor associated with Klinefelter syndrome. A report of case and review of the literature". *Eur. J. Pediatr.,* 1992, *151*, 735.

[14] Wang R.M., Shih J.C., Ko T.M.: "Prenatal sonographic depiction of fetal mediastinal immature teratoma". *J. Ultrasound Med.,* 2000, *19*, 289.

[15] Liang R.I., Wang P., Chang F.M., Chang C.H., Yu C.H.: "Prenatal sonographic characteristics and Doppler blood flow study in a case of a large fetal mediastinal teratoma". *Ultrasound Obstet. Gynecol.,* 1998*, 11,* 214.

[16] Weinraub Z., Gembruch U., Fodisch H.J., Hansmann M.: "Intrauterine mediastinal teratoma associated with non-immune hydrops fetalis". *Prenat. Diagn*., 1989, *9,* 369.

[17] Froberg M.K., Brown R.E., Maylock J., Poling E.: "In utero development of a mediastinal teratoma: a second-trimester event". *Prenat. Diagn*., 1994, *14,* 884.

[18] Dumbell H.R., Coleman A.C., Pudifin J.M., Winship W.S.: "Prenatal ultrasound diagnosis and successful management of mediastinal teratoma. A case report". *S. Afr. Med. J.,* 1990, *20,* 481.

[19] Takayasu H., Kitano Y., Kuroda T., Morikawa N., Tanaka N., Fujino A. *et al*.: "Successful management of a large fetal mediastinal teratoma complicated by hydrops fetalis". *J. Pediatr. Surg.,* 2010, *45,* e21.

[20] Noreen S., Heller D.S., Faye-Petersen O.: "Mediastinal teratoma as a rare cause of hydrops fetalis and death: report of 3 cases". *J. Reprod. Med*., 2008, *53*, 708.

[21] Wesolowski A., Piazza A.: "A case of mediastinal teratoma as a cause of nonimmune hydrops fetalis, and literature review". *Am. J. Perinatol*., 2008, *25*, 507.

[22] Akosy F., Sen C., Danisment N.: "Congenital mediastinal immature teratoma: a case report with autopsy findings". *Turk. J. Pediatr*., 2002, *44,* 76.

[23] Allman A.W., Buss P.W., Spicer R.D., Wake A.: "Mediastinal tertoma presenting as apparent fresh stillbirth". *Arch. Dis. Child. Fetal. Neonatal Ed*., 2001, *84*, F5.

[24] Isaacs H. Jr.: "Perinatal (fetal and neonatal) germ cell tumors". *J. Pediatr. Surg*., 2004, *39,* 1003.

[25] Grethel E.J., Wagner A.J., Clifton M.S., Cortes R.A., Farmer D.L., Harrison M.R. *et al*.: "Fetal intervention for mass lesions and hydrops improves outcome: a 15-year experience". *J. Pediatr. Surg*., 2007, *42,* 117.

[26] Adzick N.S., Kitano Y.: "Fetal surgery for lung lesions, congenital diaphragmatic hernia, and sacrococcygeal teratoma". *Semin. Pediatr. Surg*., 2003, *12* 154.

[27] Kitano Y., Flake A.W., Crombleholme T.M.: "Open fetal surgery for life-threatening fetal malformations". *Semin. Perinatol.,* 1999, *23*, 448.

Address reprint requests to:
A. GIANCOTTI, M.D.
Department of Obstetrics, Gynecology
and Urologic Sciences
"Sapienza" University
Viale del Policlinico, 155
00161 Rome (Italy)
e-mail: antonellagiancotti@virgilio.it