Case report - Congenital

Intrapericardial teratoma in a low birth weight preterm infant: a successful multidisciplinary approach

Gabriele M. Iacona, Miguel A. Barber, Margarita Medina, Raul Abella

1. Introduction

Primary tumours of the heart in infants have an incidence of 0.0017–0.28% [1]. Intrapericardial teratomas are even more rare [2]. They can be diagnosed during foetal life and can be associated with pericardial effusion and hydrops [3]. They often present with severe cardiac and respiratory compromise at birth [4].

We report the case of an intrapericardial teratoma diagnosed during foetal life and treated with success.

2. Case report

A 24-year-old healthy woman, gravida 1 para 0, no risk factors for foetal cardiac abnormalities and normal foetal heart at 20 weeks was referred to our unit for foetal pericardial effusion at 32 weeks gestational age (GA). Foetal ultrasound confirmed the large pericardial effusion and revealed a 2.5-cm intrapericardial cystic mass (Fig. 1a). An intrauterine pericardiocentesis was performed to avoid cardiac tamponade and lung compression, with drainage of 40 ml of serous fluid containing mesothelial cells.

At 33 weeks GA, a new intrauterine pericardiocentesis drained 25 ml effusion.

At 34 weeks GA, the size of the mass (4 cm) and the pericardial effusion were increasing. Considering the risk of hydrops, vaginal delivery was induced after a third intrauterine pericardiocentesis (36 ml) and corticosteroid administration.

A baby girl weighing 1.98 kg was born without cardiorespiratory compromise. Apgar scores at 1 and 5 min were 8 and 9, respectively. On the chest X-ray, the mediastinum was enlarged. Echocardiography showed a 4.0 × 3.0-cm cystic intrapericardial mass above the root of the great vessels, no heart abnormalities, no collapse of the right heart (Fig. 1b). Thoracic CT-scan excluded multiple localizations. The alpha-fetoprotein level at birth was >350 µg/l (no quantitative assessment possible).

At three days of life, the baby was operated with cardiopulmonary bypass (CPB) on stand-by. Ten millilitres of pericardial effusion were drained. The mass arose from the ascending aorta (Fig. 2) without adhesions with other structures. It was completely removed without CPB. The aortic adventitia was partially resected. Postoperative course in intensive care unit was uneventful. The patient was extubated on postoperative day 0 and transferred to the neonatal intensive care unit the day after.

Pathology confirmed the diagnosis of immature intrapericardial teratoma, grade II. It revealed an encapsulated, multilobulated mass consisting of solid and cystic portions.
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Fig. 1. Echocardiography showing: (a) foetal pericardial effusion and cystic mass, (b) postnatal 4.0×3.0 cm cystic intrapericardial mass above the left ventricle.

The tumour was composed of immature neural tissue, focal areas of cartilaginous tissue and smooth muscle cells, cylindrical ciliated epithelium.

Alpha-fetoprotein levels were 2103 μg/l at one month and 169 μg/l at three months, postoperatively. At three-month follow-up, the baby was doing-well and echocardiogram showed a normal heart function without pericardial mass.

3. Discussion

Teratomas are embryonal neoplasms containing three germ layers (endoderm, ectoderm, and mesoderm) [5]. Teratomas in newborns usually occur in the sacrococcygeal area, testis and ovaries and less frequently in the neck or intracranium. Intrapericardial teratomas are a rare occurrence (5–6 out of 10,000 children) [2]. They are usually benign and arise from the root of the great vessels. Their fibrous pedicle has a blood supply coming from the adventitial vasa vasorum.

Diagnosis can be done after 20 weeks GA. Tumour growth can be very rapid. The foetal ultrasound scan usually shows large pericardial effusion with an intrapericardial mass. The mass appearance is multilobulated and cystic with calcificated areas. The pericardial effusion is serous and contains mesothelial cells. Foetal MRI can add information supporting the diagnosis [6].

Pericardial effusion is probably due to the irritative stimulus of the tumour on pericardial layers and rupture of the cysts. Furthermore, the tumour can cause mechanical obstruction of the venous and lymphatic drainage leading to the development of pericardial and pleural effusions, ascites. Esophageal compression can cause polyhydramnios. Pericardial effusion and mass effect are responsible for foetal pericardial tamponade. Finally, the foetus can suffer from hydrops.

Repeated intrauterine pericardiocentesis or placement of a thoraco-amniotic shunt allows postponing birth by reduction of pericardial effusion and cardiac obstruction with normal lung development [2].

Delivery has often to be scheduled before term on the basis of tumour growth and presence of hydrops, always considering the weight of pulmonary immaturity. Some authors suggest caesarean extraction due to the high risk of fatal chest and heart compression during vaginal delivery [4]. When the fetus achieved 34 weeks GA, we induced vaginal delivery to avoid complications from repeated pericardiocentesis and development of hydrops. In our opinion, an intrauterine pericardiocentesis just before delivery can prevent pericardial tamponade during vaginal extraction.

At birth neonates can present pericardial tamponade and respiratory distress from tracheal compression and lung immaturity. The role of an EXIT (ex utero intrapartum therapy) procedure has been described [6].

Diagnosis is confirmed by echocardiography. Contrast thoracic CT-scan or MRI can better describe the relationship of the mass with surrounding structures, and the presence of multiple localizations [7, 8].

Surgery is mandatory in order to eradicate the tumour and to prevent or treat the haemodynamic repercussions. The presence of hydrops is a bad prognostic factor for surgery [3, 9]. Tumoural exeresis is almost always complete and partial adventitial resection should be accomplished. Mass is manipulated with caution in order to avoid its rupture. Stormy postoperative course has been reported [4]. Our neonate was haemodynamically stable without
hydrops. The tumour was huge but it was compressing neither the heart nor the trachea.

Long-term follow-up is required especially when the tumour is immature or when resection is incomplete. Alpha-fetoprotein is a tool to monitor a possible recurrence [10].

In conclusion, we believe that in case of an intrapericardial teratoma the foetal effusions should be treated in order to avoid the development of hydrops. In its absence, expectant management allows somatic growth and lung maturation. When lung maturation has been achieved delivery should be induced in a third-level centre with a paediatric cardiac surgery team on stand-by.

In presence of hydrops, the management depends on the gestational age:

- >30 weeks GA foetus: preterm delivery and postnatal surgery are an option,
- <30 weeks GA foetus: aggressive treatment of the effusions is mandatory, even if some advocate the possibility of foetal tumour resection [3].

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References