Successful surgical treatment of a 1160 g neonate with cardiac teratoma and severe foetal hydrops: a case report

Matthias Beichl, Margarita Thanhaeuser, Barbara Ulm, and Daniel Zimpfer

1Department of Pediatrics and Adolescent Medicine, Division of Pediatric Cardiology, Medical University of Vienna, Austria; 2Department of Pediatrics and Adolescent Medicine, Division of Neonatology, Pediatric Intensive Care and Neuropediatrics, Medical University of Vienna, Austria; 3Department of Obstetrics and Gynecology, Division of Feto-Maternal Medicine, Medical University of Vienna, Austria; and 4Department of Surgery, Division of Cardiac Surgery, Medical University of Vienna, Austria

Received 30 June 2020; first decision 10 August 2020; accepted 1 December 2020

Background
Prenatally diagnosed pericardial teratoma present a rare finding with an unfavourable prognosis due to frequently associated Foetal hydrops and limited treatment options. We report a successful surgical resection of a prenatally diagnosed cardiac teratoma in a 1160 g neonate with severe Foetal hydrops and cardiac deterioration.

Case summary
The patient was transferred in utero to our institution due to prior diagnosed pericardial mass and severe foetal hydrops, which necessitated caesarean section one day after arrival at a gestational age of 28 + 0 weeks. After intubation, the patient was stabilized by surgical drainage of 60 mL of pericardial effusion. Further clinical worsening of the patient on the day of life 12 demanded urgent intervention, so that in toto resection of the tumour was performed at a bodyweight of 1160 g. Histopathological analysis revealed a teratoma and the patient is in excellent clinical condition one year after surgery.

Discussion
This case report demonstrates that an interdisciplinary, two-staged approach can be a feasible and promising treatment option in patients with prenatally diagnosed teratoma and severe Foetal hydrops in a critical circulatory state. Furthermore, it illustrated that resection of pericardial masses can be successfully performed at a bodyweight as low as 1160 g.

Keywords
Cardiac teratoma • Foetal cardiac teratoma • Foetal hydrops • Cardiac tamponade • Pericardiocentesis • Cardiac surgery • Case report

Learning points
• Pericardial teratoma is frequently associated with foetal hydrops and demand close monitoring due to possible complications.
• Prenatally diagnosed pericardial teratoma can progress rapidly, which can lead to compression of cardiac structures by the tumour itself and therefore to rapid clinical deterioration.
• An interdisciplinary, two-staged approach is a feasible and promising treatment option even in preterm infants with a weight as low as 1160 g.

* Corresponding author. Tel: +43-1-40400-32320, Fax: +43-1-40400-32380, Email: matthias.beichl@meduniwien.ac.at
Handling Editor: Sylvia Krupickova
Peer-reviewers: Inga Voges and Filippo Puricelli
Compliance Editor: Daniel Tardo
Supplementary Material Editor: Fabienne Vervaat
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Introduction

With an incidence of 0.14–0.25%, primary cardiac tumours are an extremely rare finding in foetuses. Despite more than 80–90% of foetal cardiac tumours being benign, they can cause significant complications depending on their size and location, for example, pericardial effusion and Foetal hydrops, arrhythmias, or severely compromised circulation. Pericardial teratomas are mostly diagnosed prenatally, typically presenting with an intracardiac or pericardial multicystic mass originating anteriorly from the pericardial cavity. After complete surgical resection, teratomas have a very good long-term prognosis, resulting in the highest survival rate among all primary cardiac tumours. However, the outcome of prenatally detected teratomas is much less favourable because of possible compression of the heart structures and pericardial effusion. This case describes a foetus who presented to our institution at a gestational age of 27 + 6 weeks with a pericardial tumour and unfavourable prognosis due to severe Foetal hydrops and imminent circulatory decompensation.

Timeline

Day 0  
A pericardial mass, as well as pericardial effusion was detected during foetal echocardiography at gestational age of 21 + 4 weeks.

Day 42  
Follow-up sonography showed distinct growth of the mass as well as severe Foetal hydrops.

Day 44  
The patient was transferred to our centre in utero at a gestational age of 27 + 6 weeks.

Day 45  
Due to the critical clinical condition of the foetus, caesarean section and surgical pericardiocentesis were performed.

Day 53  
As the patient’s condition improved gradually, he was weaned from ventilation.

Day 56  
The patient had to be re-intubated due to increased oxygen demand, respiratory acidosis, and subsequent respiratory failure. As echocardiography revealed a critical hemodynamic situation with increasing obstruction of systemic and pulmonary venous return by the tumour, surgical resection was performed on the same day at a body weight of 1160 g.

Day 62  
Histological analysis confirmed the suspected diagnosis of a cardiac teratoma, precluding any further oncological treatment.

Day 114  
The patient was discharged in good clinical condition without any cardiac medication at a corrected gestational age of 37 + 6 weeks.

1 year  
The patient is in an excellent clinical condition with a normal echocardiogram, no signs of residual tumour and normal motor and psychosocial development.

Case presentation

The pericardial tumour was first diagnosed at another institution by foetal echocardiography at a gestational age of 21 + 4 weeks in a 34-year-old healthy primipara. Up to this point, the course of pregnancy was uneventful. During follow-up, progressive growth of the tumour as well as increasing Foetal hydrops demanded timely management. The mother was then transferred to our hospital for further evaluation. On admission, the mother presented in a good clinical condition, laboratory parameters showed no infection, and TORCH serology was negative.
Foetal ultrasound at admission to our hospital revealed compromised circulatory status of the foetus. The pericardial mass was displayed with partial compression of the right atrium and ventricle. Both cardiac output of the left (195 mL/kg/min) and right (100 mL/kg/min) ventricle were impaired, resulting in a decreased total cardiac output of 295 mL/kg/min. Furthermore, significant pericardial and pleural effusions (Figure 1), ascites, oedema, and bradycardia with a heart rate of 113/min were detected.

Foetal MRI, which was performed subsequently, confirmed the pericardial mass with a size of 2.8 cm × 2.4 cm × 2.7 cm and multicystic appearance, which was highly suspicious for a pericardial teratoma (Supplementary material online, Figure S1).

Lung induction therapy was initiated by the application of prenatal steroids. The initially established plan was to postpone delivery as long as possible under close surveillance. However, progressive worsening of cardiac filling due to compression of the tumour and initiating cardiac tamponade as well as significant bradycardia required immediate action within 24 h of admission. We estimated that there existed a viable treatment option in a two-staged approach, despite the unfavourable circumstances. Thus, caesarean section was performed at a gestational age of 28 + 0 weeks.

At initial examination, the female newborn presented with compromised circulatory state, respiratory distress, and bradycardia. With a birth weight of 1600 g, the patient was in the upper range (97th percentile), which could be explained by the severe Foetal hydrops. Immediately after intubation, surgical drainage of the pericardial effusion (60 mL) was performed in the neonatal intensive care (echocardiography see Figure 2, Video 1), together with drainage of both pleural spaces and ascites, application of surfactant, and...
temporary inotropic support with dobutamine. These measures led to stabilization of the patient (see chest X-ray, Supplementary material online, Figure S2A) so that the child could even temporarily be weaned from mechanical ventilation a few days after primary care.

On the day of life 12, the patient had to be re-intubated due to increased oxygen demand, respiratory acidosis, and subsequent respiratory failure. Echocardiography revealed not only new pericardial effusion but also a significant growth of the tumour (see chest X-ray, Supplementary material online, Figure S2B), which caused critical congestion of the superior and inferior vena cava (Video 2) as well as incipient obstruction of pulmonary venous return (Video 3).

Despite the low body weight of 1160 g and the substantial risk of the procedure, surgical resection of the tumour was thought to be the only viable option at that time. Intraoperatively, a tumour adhesive to the ascending aorta with the size of 5 cm × 3 cm × 2.5 cm was resected in toto without any complications (Figure 3). The post-operative clinical course was uneventful. The infant was extubated and all drainage tubes were removed on the third post-operative day; the remaining stay at the neonatal intensive care unit was uneventful.

The histopathological analysis of the tumour revealed an immature, cystic teratoma. The case was discussed at a local interdisciplin ary tumour board supported by an additional international expert opinion and was evaluated to be cured by surgery only, with no need for further chemotherapeutic treatment.

The patient was discharged home at a corrected gestational age of 37 + 6 weeks with a body weight of 2660 g. After 1 year of follow-up, the patient is in excellent clinical condition with normal motor and psychosocial development. Echocardiography depicted normal systolic and diastolic heart function, no residual tumour, and no evidence of residual obstruction of the systemic venous drainage.

Discussion

Rapid growth of the pericardial mass and progression of Foetal hydrops in association with immaturity the foetus can be very challenging clinical issues in patients with foetal cardiac teratomas, as demonstrated in this case. At admission to our institution, the patient already presented with an impaired circulatory state. This was reflected by the critically reduced biventricular cardiac output of 295 mL/kg/min, which was distinctly below reported normal values of 553 ± 153 mL/kg/min.7

Within the first 24-h, worsening of cardiac filling, especially of the right ventricle as well as bradycardia led to the suspicion of cardiac tamponade, which required immediate consideration to proceed with further procedural intervention. One strategy which has been suggested by some institutions 8–11 to allow circulatory improvement and thereby delay delivery until a later gestational age, is prenatal pericardiocentesis. However, depending on the mechanism of cardiac impairment, pericardiocentesis is not always effective, in fact it might even increase the compression that is caused by the tumour itself. Therefore, caesarean section and subsequent surgical drainage of the pericardial effusion was the best treatment option in our opinion. A multidisciplinary team was available postpartum for primary neonatal care, cardiac surgery, and cardiologic management.

In our case, the tumour measured up to 5 cm in diameter at the time of surgery. The tumour showed an impressive growth within 13 days from a maximum diameter of 2.8 cm up to 5 cm comparing the measurements from the foetal MRI and the resected tumour. This substantial increase in size was also accompanied by significant hemodynamic alterations, namely systemic and pulmonary venous congestion. At the time of tumour resection our infant still had a very low body weight of 1160 g, but delaying surgery further was precluded by clinical worsening unresponsive to medical treatment. The presented infant is the smallest reported patient surviving successful resection of a cardiac teratoma. Surgical risk was substantial, as mechanical circulatory support during surgery by cardiopulmonary bypass with anticoagulation was not an option due to small vessel sizes, tumour location, and the high risk of intraventricular haemorrhage of the preterm. Furthermore, complete tumour resection may be restricted by technical limitations with a patient this size.

In conclusion, this case report demonstrates that even in emergency situations accurate diagnosis and haemodynamic judgement of a patient with prenatally diagnosed cardiac tumour and interdisciplinary management by an experienced team enabled successful treatment despite numerous risk factors and an initial situation in which there appeared to be no viable options.

Lead author biography

Dr Matthias Beichl graduated from the Medical University of Vienna in September 2014. Since 2015, he has been a resident at the Division of Pediatric Cardiology at the Department of Pediatrics and Adolescent Medicine at the Medical University of Vienna, Austria. His main clinical and scientific interest is interventional pediatric cardiology, echocardiography, especially 4D-echocardiography and speckle tracking imaging, as well as pediatric intensive care.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient’s next of kin in line with COPE guidance.

Conflict of interest: none declared.

Funding: none declared.
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