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Giant Intrapericardial Teratoma – Enough Space Left in the Neonatal Thorax?

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Authors' contributions

This work was carried out in collaboration among all authors. Author MW treated the patient and wrote the manuscript. Authors CP and FH reviewed the literature and revised the manuscript. Author MV supervised the surgery and reviewed the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Objective: Optimal treatment of fetal intrapericardial teratoma is controversial, especially in regard to fetal intervention. Given the rarity of the disease case reports can assist in decision making.
Case Report: We report on a neonate with a giant intrapericardial teratoma detected in utero, almost filling the entire thorax. Delivery was planned per cesarean section with extracorporeal membrane oxygenation (ECMO) stand-by. As a surprise the child adopted very well after birth, requiring only continuous positive airway pressure (CPAP). The tumor was resected on the next day without injuring cardiac structures. The child was and discharged on day 10 post-surgery.
Conclusion: Our case supports the assumption that even in very large tumors the postnatal course can be benign, if there is no fetal hydrops.

Keywords: Intrapericardial teratoma; fetal intervention; cardiac tumor; conservative management.

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1. INTRODUCTION

Fetal intrapericardial teratomas are rare, affecting less than 0.01% of newborns [1,2,3,4]. Although most often benign they can be lethal due to pulmonary or circulatory compromise related to their size [5,6]. Fetal hydrops seems to be a marker for an unfavorable outcome. Yet, optimal management is controversial regarding the need for fetal intervention [7,8]. The aim of our case report is to add knowledge about the optimal management of this rare disease.

2. PRESENTATION OF CASE

Prenatal ultrasound scan at 30 weeks of gestation showed a large intrathoracic tumor. The tumor showed rapid progression in size over the next weeks. At 37 weeks of gestation, the size of the tumor within the pericardial cavity equaled the size of the fetal heart. In addition there was a massive pericardial effusion Fig. 1.

Fetal hydrops was not prevalent. The lungs were categorized as hypoplastic. Delivery by cesarean section at 38+1 weeks of gestation was planned in the operating room of the heart center with neonatology, cardiology and cardiac surgery stand-by. Need for ECMO therapy immediately after birth was discussed and prepared for due to lung hypoplasia and likely cardiac compromise. Surprisingly the child did very well after birth with good adaption (APGAR 8/9/10). Birth weight was 3210 g. Due to mild dyspnea, the baby was placed on CPAP but did not need intubation despite severely reduced aeration on chest X-ray Fig. 2.

Circulation was stable without the need for catecholamines. Echocardiogram confirmed prenatal findings with a cystic tumor of 4x4x3.5 cm within the pericardium attached to the base of the heart and adjacent to the great arteries Figs. 3 and 4.

Left and right outflow tracts were unobstructed. The circulation was not duct dependent. The pericardial effusion was large, but due to the mild clinical symptoms was not drained before surgery, which took place the next day. At surgery attachment of the tumor at the aortic root was found. In toto resection of the tumor was possible without injuring the vessel walls and no need for cardiac bypass or circulatory arrest. The postsurgical course was uneventful with extubation on the next day. Histology showed mildly immature teratoma (G1 as per Gonzales-Crussi) [9]. The child was transferred to a lower level hospital on day 2 post-surgery to complete treatment and was discharged home 10 days after surgery. No adjuvant chemotherapy was given. One year follow-up showed no tumor relapse.

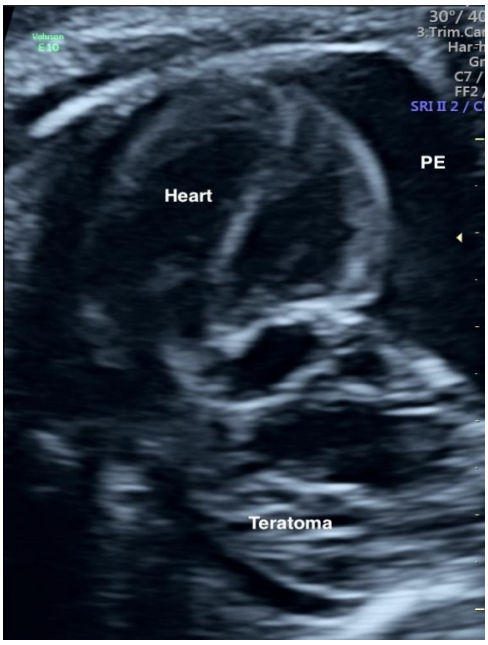

Fig. 1. Fetal ultrasound at 37 weeks of gestation demonstrates a large intrapericardial teratoma about the size of the fetal heart, surrounded by a large pericardial effusion (PE)

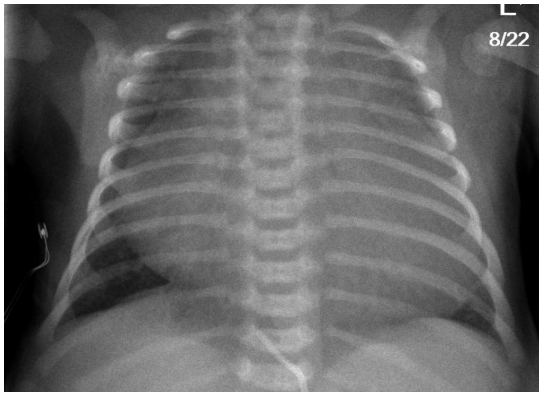

Fig. 2. Chest X-ray after birth shows massive enlargement of the mediastinum

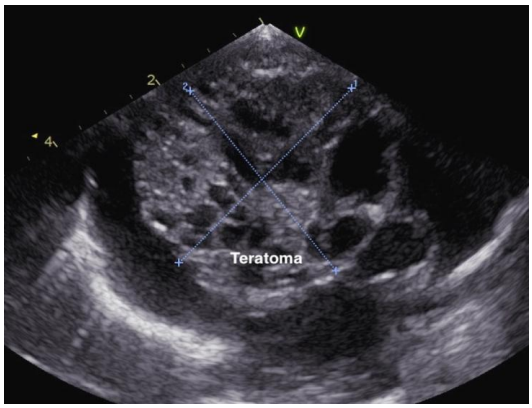

Fig. 3. Postnatal echocardiogram reveals a tumor size of about 4x4 cm

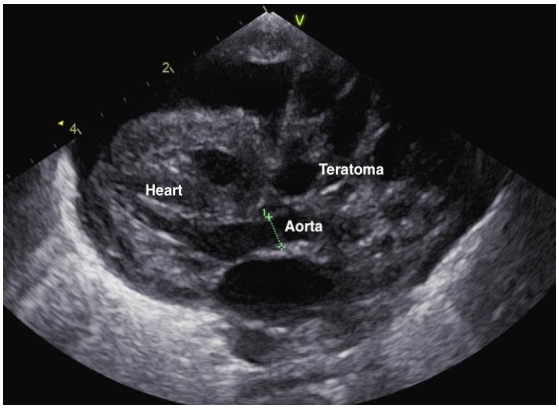

Fig. 4. The tumor originates at the cardiac base adjacent to the aortic root

3. RESULTS AND DISCUSSION

Our case illustrates the difficulty to predict pre- and postnatal course in intrapericardial teratoma.

Nassr et al. [7] reviewed the literature to evaluate the outcome of fetal detected intrapericardial teratoma in 2016. Only case reports or small case series were found. They included 67 fetuses in their review, 46 with hydrops, 21 without. 26 fetuses underwent some kind of fetal intervention (pericardiocentesis in the majority of them) with a favorable outcome in 75%. Of those without intervention, only 30.8% had a favorable

outcome. Six children without hydrops underwent a fetal intervention without any complication. However, in those fetuses without hydrops, survival without intervention was good as well. There was only one stillbirth and one child that died after a serious complication during postnatal surgery.

Due to conflicting results regarding the benefit of fetal intervention and the lack of fetal hydrops we opted to defer treatment to the postnatal period, but were prepared for a complicated course including the need for ECMO support. Despite the size of the tumor and prenatally diagnosed lung hypoplasia, symptoms were remarkably sparse without need for pre-surgery intervention. The child underwent postnatal surgery successfully without complications. Our case thus favors a more conservative approach regarding fetal intervention in cases without hydrops.

4. CONCLUSION

Despite massive proportions, symptoms of intrapericardial teratoma might be sparse. Conservative prenatal management might be justified in cases without hydrops.

CONSENT

The patient's parents have given their informed consent for the case report to be published.

ETHICAL APPROVAL

As the report is a case presentation formal ethics approval is not applicable. The report has been conducted following the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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