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Brief Report

Intrapericardial immature teratoma in a newborn: a case report

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Abstract Intrapericardial teratomas are extremely rare and most often benign tumours. In this paper, we have described a case of intrapericardial teratoma diagnosed prenatally and successfully operated. The presented case is noteworthy as an example of potentially catastrophic cardiorespiratory distress caused by the space-occupying nature of the tumour. A multi-disciplinary approach is mandatory because the tumour most often arises from the ascending aorta and in some cases may require the use of cardiopulmonary bypass.

Keywords: Teratoma; intrapericardial; newborn

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MEDIASTINAL TERATOMAS ARE COMMON PERINATAL tumours, second only to neurogenic tumours, with a yearly incidence of ~1 in 40,000 live births.[1] On the other hand, intrapericardial teratomas are exceptionally rare and sparsely reported,[2] although the first description dates back to 1890.[3] Remarkable progress of prenatal ultrasonography has led to a more accurate diagnosis of this potentially fatal condition. Timely diagnosis allows immediate response both in terms of planning the timing of delivery and emergency surgery in order to prevent the effects of compression or rupture of the tumour.

Case report

The newborn is one of the twins from an in vitro fertilisation pregnancy, born of a 27-year-old primigravida. Initial ultrasound screening revealed normal twin pregnancy. However, repeated foetal ultrasonography done at 32 weeks of gestation showed a large tumour mass in front of the heart with hydrops fetalis and pleural effusion in one of the twins (Fig 1). Considering the safety of the mother and the healthy twin and a risk of cardiac compression and hydrops for the sick baby, a caesarean section was performed.

Upon delivery, a 2170 g female neonate with diffuse oedema experienced cardiorespiratory distress and had to be resuscitated and intubated. After this, repeated right thoracocentesis was performed in order to remove a total of 200 ml serous fluid. Blood screenings were normal with no signs of infection, and biochemical findings demonstrated a highly elevated level of α-fetoprotein (110,995 μg/L). The chest X-ray showed an enlarged mediastinum. Transthoracic echocardiogram revealed a huge mediastinal tumour mass without revealing any further cardiac abnormalities. However, heart contractility was impaired. Thoracic computed tomography scan confirmed the presence of a homogeneous, well-circumscribed tumour mass 53 × 48 × 28 mm in the anterior mediastinum, and multiple localisations were excluded. The low density of the tumour corresponded to fat tissue, and there were no calcifications.

The baby was operated upon on the third day of life. During the introduction of anaesthesia and positioning for surgery, she experienced a cardiac arrest and had to be resuscitated with adrenaline. A median sternotomy was performed, and upon pericardiotomy the heart was not visible. The huge and well-demarcated tumour mass was compressing the right atrium and ventricle, superior caval vein, aorta, and the pulmonary artery (Fig 2). After full mobilisation, it was obvious that it arose from the anterior portion of the ascending aorta. Subsequently, it was completely removed; a portion of the ascending aorta adventitia was also resected. There was no need for cardiopulmonary bypass.

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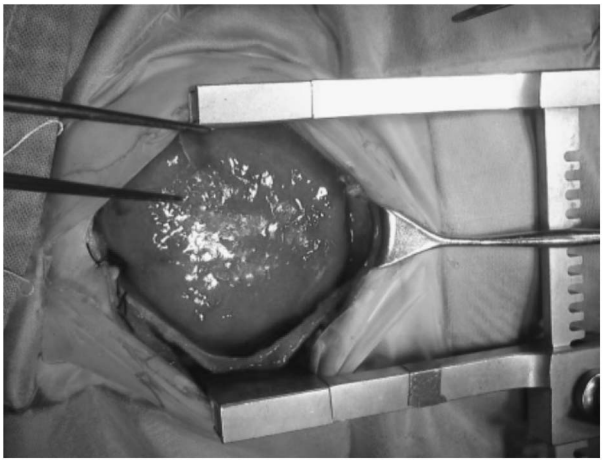

Figure 1.
Foetal ultrasound study demonstrating a tumour mass in front of the heart with pericardial effusion and ascites.

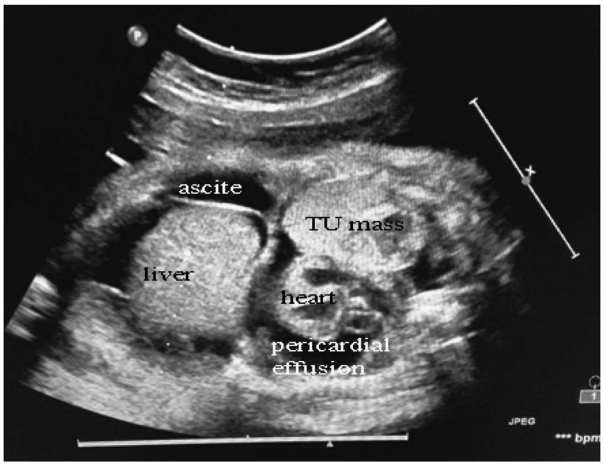

Figure 2.
Intrapericardial tumour mass completely covering the heart.

The post-operative course was uneventful, and she was extubated on the second post-operative day and shortly after transferred to the neonatal department. Histopathology confirmed the diagnosis of immature teratoma grade III. The tumour was mainly composed of immature neural tissue, with smaller areas of epithelial and mesenchymal structures. α-Fetoprotein dropped immediately after surgery to 62,215 μg/L, and continued decreasing to 205 μg/L after 3 months, and 37 μg/L after 5 months. After 11 months, the child is well, symptom free, and with no signs of tumour recurrence.

Discussion

Teratomas are embrional neoplasms, derived from omnipotent cells that contain tissue from at least two or three germ layers. Their localisation in the mediastinum, most often anterior, is not uncommon among neonates and children.[4] On the other hand, their intrapericardial presentation is extremely rare. The most comprehensive structured review of the English literature by MacKenzie et al[5] identified 46 case reports. Among the 22 described survivors, there are only three cases with a lower body weight than our patient. The tumour size and the degree of prematurity emphasise the complexity of the presented case as well.

These tumours tend to grow rapidly between the 20th and the 40th week of gestation.[6] The accelerated growth may result in fatal compression to the heart, as well as pericardial and pleural effusion, due to mechanical obstruction of systemic venous and lymphatic drainage. Hydrops fetalis is another well-known risk factor for fatal termination of pregnancy.[5] A significant rise in cardiac compression during vaginal delivery may result in fatal chest and heart compression.[7] These factors are most likely responsible for poor prognosis for these babies if the diagnosis is not established prenatally.

A foetus with an intrapericardial cystic or mixed mass arising from the anterior cardiac surface and associated with pericardial or pleural effusion is suggestive for this diagnosis. Early manifestation in the second trimester with hydrops and effusions is a severe problem because of the prematurity. Some centres advocate transabdominal pericardiocentesis to allow progression of foetal development or even open foetal surgery.[8] Fortunately, a vast majority of the cases are presented in the third trimester when caesarean delivery can be performed in cases of cardiac compression or hydrops.

Following birth, these babies can suffer from cardiovascular compression and respiratory distress, and thus it is necessary to ensure close monitoring of their clinical condition. The diagnosis is usually confirmed by a computed tomography scan. However, sometimes it can be difficult to distinguish whether the origin of the tumour is intrapericardial or anterior mediastinal. This can be the case especially in low-body-weight neonates with enormously large tumour masses. In our case, pre-operative cardiovascular collapse was caused by chest hyperextension during positioning for surgery, which most likely caused preload heart reduction by extensive compression of the tumour mass. The post-operative course is usually uneventful, although haemodynamic instability is reported sporadically.[2]

Surgery is absolutely mandatory even in patients with deteriorated clinical condition. Despite the benign nature of these tumours, one should bear in mind that almost 15% of immature teratomas are classified as malignant, depending on the neuroectodermal cell composition.[7,9,10] Surgical procedure

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for removal is usually uncomplicated. Although the literature supports resection without the use of cardiopulmonary bypass, in some cases it had to be used.[7] Having in mind this possibility, multidisciplinary approach is mandatory.

Long-term follow-up is obligatory for all patients operated for immature teratoma. α-Fetoprotein is well known as a tumour marker indicating relapse. It has been suggested that the rapidity of α-fetoprotein decay after resection is a more accurate prognostic factor than any isolated value.

In conclusion, accurate prenatal diagnosis, close monitoring for hydrops and signs of cardiac compression, timely planned caesarean delivery, and early surgery provide good prognosis for patients with immature intrapericardial teratoma.

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