Excision of an intrapericardial immature teratoma in a 26-week premature neonate

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A B S T R A C T

We present a case of a 26-week premature newborn with an immature intrapericardial teratoma. The patient was transferred from an outside hospital for management of a large mediastinal mass causing respiratory insufficiency. The newborn was supported with the help of a large interdisciplinary team until day of life 22 when he underwent surgical excision. On follow up the infant is doing very well and is one of the youngest survivors to date.

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Intrapericardial teratoma is a germ cell tumor that typically arises from the base of the heart [1]. While usually benign, intrapericardial teratomas can become relatively very large and cause symptomatic compression or fetal hydrops in utero [2]. Surgical excision is curative, even for most immature variants. However, infants with this condition may require significant support and multiple interventions prior to surgery. Advances in critical care, surgery and interdisciplinary management have resulted in dramatic improvement in treatment outcomes over recent decades.

1. Case report

A 26-week and 3 days old male neonate was delivered by cesarean section at an outside center after an ultrasound performed for premature labor demonstrated a large chest mass in the baby in utero. His birth weight was 1212 g and he required intubation for respiratory distress. After stabilization he underwent chest computed tomography (CT) revealing a mediastinal mass measuring 5 × 3 × 4 cm. He was then transferred to our center. Complicating factors included extreme prematurity, patent ductus arteriosus (PDA), and neonatal respiratory distress syndrome. Ultrasound of the chest at our center demonstrated a 4.4 × 3.1 × 3.6 cm heterogeneous mass adjacent to the right heart with compression of the right atrium and ventricle, a pericardial effusion, and a small left to right PDA. Laboratory tests revealed human chorionic gonadotropin of <0.12 and alpha-fetoprotein (AFP) of >363,000, which is elevated even after adjusting for prematurity. Initially, the baby's status was relatively stable with assisted ventilation, so the decision was made to delay excision to allow for further growth. However, on day 7 of life he developed worsening hypercarbia and had to be placed on high frequency oscillatory ventilation and two days later on the Dräger-VG. Serial ultrasounds demonstrated worsening pericardial effusion, which was drained on day 11 of life with improvement in respiratory status. He again required pericardiocentesis and drain placement on day 13 of life.

With continued respiratory distress requiring prone positioning, the decision to operate was made on day 22 of life. His weight was only 1410 g and the relatively high likelihood of an inability to go on bypass was considered. Collaboration between surgeons from the Divisions of Pediatric Surgery and Pediatric Cardiac Surgery determined the risk of continued observation outweighed the risk of surgery at this time. The baby underwent median sternotomy and upon entry into the pericardial space straw colored fluid was drained. Dissection around the large teratoma revealed that it was well-encapsulated with clean planes. There were connective tissue...
attachments to the right atrium and small vascular attachments to the ascending aorta (Fig. 1). All were divided with electrocautery and no blood was lost during the procedure. The specimen was 6.2 cm in greatest diameter and 68.6 g (Fig. 2). Final pathology demonstrated grade three immature teratoma with predominant neural and hepatic components (Fig. 3).

Upon return to the NICU the baby transiently required dopamine support and did receive a blood transfusion. He was soon extubated and by postoperative day 12 he was tolerating full feeds and was transferred back to the referring hospital for proximity to family. Upon follow up he has been doing very well at home. He underwent repair of bilateral inguinal hernias at 5 months of age.

2. Discussion

Intrapericardial teratomas are germ cell tumors, predominantly found in newborns and infants [1]. They typically consist of all three germ layers, but can contain fewer, and the tissues can range from mature to grade three immature. When they are immature, or involve yolk sac tissue, the AFP may be markedly elevated. As many as 15% of immature teratomas will be malignant, most often due to immature neuroectodermal components [3–5]. They are typically encapsulated with a vascular stalk, arising from the aorta or pulmonary artery, with the mass anterior to the heart with compression of the right atrium and ventricle [6].

The incidence of cardiac tumors was first estimated using autopsy studies and found to be between 0.001 and 0.2% with the incidence of teratomas in infants accounting for 9% of those cases [6]. More recent estimates have been made using echocardiographic databases with an incidence of cardiac tumors between 0.17 and 0.20% in the largest studies [2,7]. Intrapericardial teratomas comprised approximately 2% of tumors in both studies.

Infants are now often diagnosed in utero during screening ultrasound. The tumor may be associated with hydrops fetalis and pericardial effusion. After birth symptoms are determined by the size of the teratoma and the presence of pericardial effusion that can result in mass effect and/or tamponade physiology [2]. Imaging after birth should include echocardiography to evaluate the size and characteristics of the tumor and effusion, cardiac function and mass effect. The diagnostic findings include multicyctic structure, location at the right anterior heart border, attachment to the great vessels, and pericardial effusion [6]. Cystic components of the lesion appear echolucent and calcifications appear echogenic. Consistent findings on CT are a combination of fatty tissues and calcifications in the mass.

Treatment of neonates and infants with intrapericardial teratoma has greatly improved as technology and resources have advanced. Care involves airway management, typically requiring intubation. Pericardiocentesis is often required as well, with either drain placement or repeat procedures depending on timing of surgery. The pericardiocentesis is often nondiagnostic, but typically nonbloody. Successful excision is occurring in increasingly premature neonates. This patient represents one of the youngest and smallest to undergo successful excision reported to date. In order to continue achieving such outcomes it is imperative that low birth weight neonates with intrapericardial teratomas be treated at a tertiary or quaternary center by a multidisciplinary team. At our institution the neonatal intensivists worked closely with pediatric cardiology, pediatric surgery, congenital cardiac surgery, pediatric oncology, radiology and respiratory therapy services.

The treatment is ultimately complete resection as this has been associated with improved survival irrespective of the teratoma’s maturity [8]. Careful histologic sectioning is required to analyze for malignant components in immature or mixed teratomas. Other considerations include AFP measurement for prognosis as well as recurrence later. In extreme premature babies, peripheral bypass is not possible due to vessel size and central cannulation may not be feasible based on the size and location of the tumor. Timing of surgery is determined by the clinical status of the affected baby. Temporization to allow growth is warranted if the baby is stable from a respiratory and hemodynamic standpoint.

Fig. 1. Anterior mediastinal mass with attachment to the right atrium and ascending aorta.

Fig. 2. Cross pathology of mediastinal mass, whole (A) and bisected (B).
3. Conclusion

The treatment of intrapericardial teratoma is feasible even in the setting of extreme immaturity. With multidisciplinary care and the development of a coherent treatment plan the neonate may be able to grow and develop prior to surgical excision. After excision these patients tend to do very well, and our patient was transferred closer to home two weeks after excision.

References


Fig. 3. Hematoxylin and eosin stain at 4× (A) and 20× (B) magnification.