Case Report

Intrapericardial immature teratoma in the new-born

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ABSTRACT

Intrapericardial teratomas are rare causes of mediastinal masses in children. Pericardial teratoma is a potentially curable lesion that may become life threatening when it induces mediastinal compression and fetal hydrops. Majority of the reported cases have been diagnosed prenatally. We report a case of an infant with intrapericardial immature teratoma, which was detected in a newborn infant presenting with respiratory distress. The tumor was excised completely, and histopathological examination of the resected tumor was suggestive of immature teratoma. The patient was asymptomatic three months postoperatively.

Key words: Immature, intrapericardial, newborn, teratoma

INTRODUCTION

Teratomas are tumors that present in infants and children as well as adults. Pediatric cases are usually found in the sacrococcygeal region, gonads, mediastinum and intracranially. Intrapericardial teratomas are rare and are usually diagnosed in the neonatal period or in utero.[1] They are associated with large pericardial effusions, hydrops and tamponade.

CASE REPORT

A newborn infant presented with respiratory distress. On echocardiography an intra-pericardial mass was detected causing pericardial effusion [Figure 1]. There was no sign of heart failure or arrhythmia but there was moderate pericardial effusion. On day 3, the infant was operated due to rapid fall in oxygen saturation. The tumor was explored through median sternotomy. Intra-operatively a 5 × 3.5 cm spherical, cystic mass was detected arising from aortic and pulmonary artery adventitia. This tumor was compressing the right atria, right ventricle and superior vena cava. About 10–15 ml of hemorrhagic pericardial fluid was aspirated from pericardial sac. The tumor was excised and sent to us for histopathological examination.

On gross examination, the tumor was well circumscribed, well encapsulated with grey brown smooth outer surface and was measuring 5 × 3.5 × 2 cm. Cut surface was variegated showing gray-brown, gray-white and tan-brown areas with multiple minute cysts, some of which are filled with mucinous material.

Histopathological examination revealed multiple cysts lined by pseudo-stratified columnar epithelium, stratified squamous epithelium, intestinal epithelium, mucinous epithelium and flattened epithelium. Foci of pancreatic tissue, smooth muscle bundles, blood vessels and lymphatic channels were seen. Neural tissue in the form of glia, choroid plexus tissue and ependymal tissue was appreciated. Multiple foci of immature cartilage as well as immature neuroepithelium were seen in the form of neuro-rosettes and neuro-tubules [Figure 2] in more than one low power field (40×). A diagnosis of Grade II intrapericardial immature teratoma was given.

DISCUSSION

Secondary tumors are more common in pericardium than the primary tumors. The incidence of primary cardiac tumors varies between 0.0017 and 0.027% of all live births.

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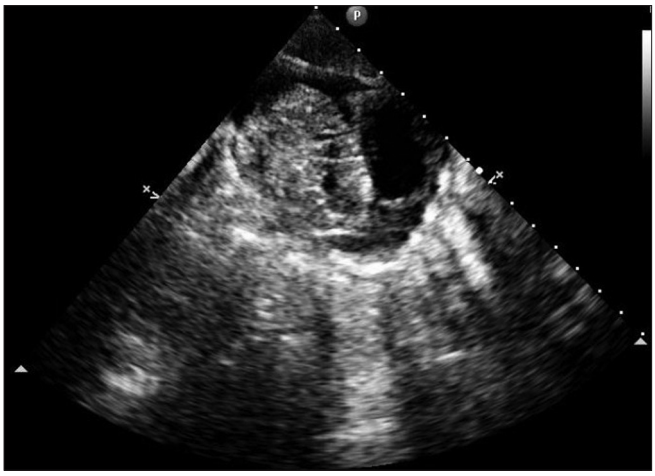

Figure 1: Echocardiogram revealed an intrapericardial, predominantly solid mass with several small cystic areas associated with pericardial effusion

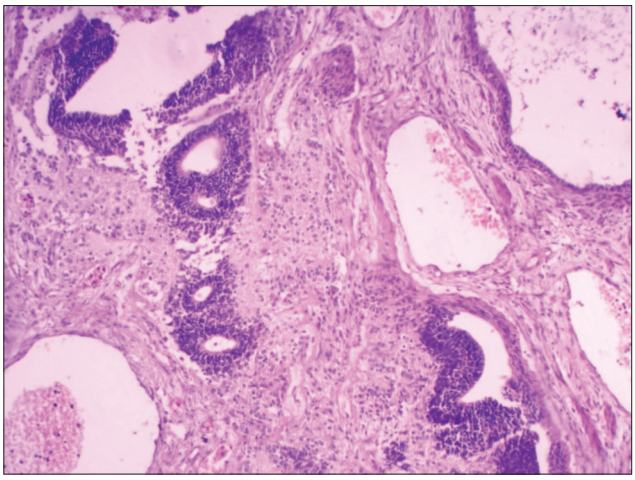

Figure 2: Photomicrograph showing immature neuroepithelium, arranged in the form of neuro-rosettes and neuro-tubules (H and E, 20×)

Teratomas correspond to 19 to 25%, of these tumors. Intrapericardial teratomas are rare lesions accounting for approximately 10% of all mediastinal masses in children.[1] Intrapericardial teratomas were first described by Joel *et al*.[2] in 1890, and to date there have been approximately 62 cases reported in the literature. Most of these are mature teratomas and very few cases are of immature type.[3]

These tumors are usually detected in the neonatal period or in utero and have exceptionally been diagnosed in older children.[4] Intrapericardial teratomas arise from the base of the heart and are attached to the root of the pulmonary artery and aorta. In fetal life, the effusion or mass effect may cause extrinsic compression on the heart and cardiac tamponade. It can also impede venous return with resultant hydrops fetalis and fetal death. After delivery, ventilation may be compromised by extrinsic airway compression or pulmonary hypoplasia and cardiac output may be compromised by mass effect or tamponade. There may be severe respiratory distress and hemodynamic compromise.[5-7] In the neonates, it can also cause cardiomegaly, congestive heart failure, neonatal hydrops and sudden death. Prenatal echocardiography offers a new approach to this problem. Diagnosis may be possible in utero. In an infant with intrapericardial teratoma, prenatal echocardiography permitted early diagnosis and immediate postnatal treatment with complete recovery.[8] Echocardiogram typically shows the pedunculated mass arising from the aorta projecting anterior and to the right side of the heart. The use of computed tomography and magnetic resonance imaging is advocated to define the tumor and its relation with the heart, and to assess the mediastinum.

A rapidly growing mass can develop from 20 to 40 weeks of gestational age and this represents the time of detection for approximately one third of the cases reported. A fetus presenting early in the second trimester with an effusion and hydrops is a complex problem because of prematurity; some centers have successfully preformed transabdominal pericardiocentesis, and open fetal surgery has been suggested[7] in the case of a severely compromised fetus.[6,9] In most cases presenting in the third trimester, a cesarean section is performed preoperatively and surgery takes place on a semi-urgent basis. Cases diagnosed near term without signs of hydrops fetalis can be followed closely with surgery after delivery. When an intrapericardial teratoma is diagnosed with a large pericardial effusion that causes impending cardiac tamponade, in utero pericardiocentesis has been successful in salvaging the fetus.

The surgical procedure for removal of an intrapericardial teratoma is usually uncomplicated. The mass is rarely associated with an intracardiac condition and removal does not necessitate cardiopulmonary bypass. The mediastinum should be explored to ascertain the absence of another teratoma.[6] Once resected, the prognosis is usually good, as most tumors are benign and require no further treatment. Morphology and clinical behavior of the immature intrapericardial teratoma described by Agozzino *et al*.[10] suggest that, as in ovarian and sacrococcygeal teratomas, the presence of immature neuroepithelium carries a poor prognosis. In such cases radio- or chemotherapy should be performed.

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