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Case Report

Intrapericardial Teratoma in a Newborn: A Case Report and Review of Literature

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Abstract

Teratoma arising from the pericardium is an extremely rare tumor.

Intrapericardialor mediastinal teratoma if detected intrauterine in fetal life, the mortality will be less. Tumor size can be large and associated with pericardial effusion, the combination of which leads to progressive cardiac tamponade, fetal hydrops, and death. In the neonatal age group most of these tumors are benign and mediastinal teratomas despite they are benign tumor but must be treated sometimes urgently because they might grow fast.

Histologically teratomas may be classified as mature or immature on the basis of the presence of immature neuroectodermal elements within the tumor. Survival was linked to the degree of immaturity in the teratoma and the risk of recurrence also appears to be related to the degree of immaturity.

We present here our case who is a newly born baby girl diagnosed as having a big mass inside the pericardial sac necessitated urgent surgical excision.

Introduction

Teratoma is a term derived from a Greek word meaning monster [1]. Classically it is composed of tissues from the three embryonic germ layers: ectoderm, mesoderm and endoderm [2]. Teratoma arising from the pericardium is an extremely rare tumor [3].

Intrapericardial and mediastinal teratoma if detected intrauterine in fetal life, the mortality can be less [4]. Tumor size can be large and associated with pericardial effusion, the combination of which leads to progressive cardiac diastolic dysfunction, cardiac tamponade, fetal hydrops and death [5].

The female-to-male predominance is 4:1.

Teratoma occurs in 1/30,000-70,000 live births [6].

In the neonatal age group most of these tumors are benign and teratomas are the 2nd most common benign cardiac tumors. Teratomas are anatomically foundgonadal or extra-gonadal [7]. The neck and mediastinum are the most common extra-gonadal locations. Frequencies of the most common sites are as follows: Sacrococcygeal - 40%, Ovary - 25%, Testicle - 12%, Brain - 5% and other (including the neck and mediastinum) - 18% [8].

Histologically teratomas may be classified as mature or immature on the basis of the presence of immature neuroectodermal elements within the tumor. Mature tumors (grade 0) have no immature elements. In grade 1 tumors, immature elements are limited to one low-power field per slide; in grade 2 tumors, fewer than four fields are present per slide; and in grade 3 tumors, more than four fields are present per slide [9].

Survival was linked to the degree of immaturity in the teratoma and the risk of recurrence also appears to be related to the degree of immaturity. Recurrence in a completely resected mature teratoma is less than 10%; but in an immature teratoma, recurrence may be as high as 33% [9,10].

Also mortality for congenital teratomas depends on gestational age and the size and location of the tumors i.e. the younger gestational age the more risk for mortality and location in cardiac, mediastinum or the neck are more risky due to compression or invasion of adjacent vital structures [9,11].

Fetal intrapericardial teratomas can also be surgically treated in intrauterine life with acceptable results as per Jack and his coauthors who claimed intrauterine surgical excision of fetal pericardial teratomas with more than 50% survival and they concluded that fetal intrapericardial teratoma can be successfully managed utilizing serial surveillance and by treatment prior to the predictable onset of hydrops, determined through increasing tumor size and a declining cardiac output. Surgical resection in

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utero is possible, with good results [12].

We present here our case who is a newly born baby girl diagnosed as having a big mass inside the pericardial sac necessitated urgent surgical excision.

Case Presentation

A girl baby was born full term with a birth weight of 2200 gm by caesarian section delivery at the 38th gestational age from a 35-year old healthy mother who was a farmer and living in a rural area. She was found to have polyhydramniosis on ultrasonography performed before delivery. The baby had severe respiratory distress since birth so was admitted to neonatal intensive care unit, intubated and mechanically ventilated. Chest x-ray was done and revealed query mediastinal mass with wide mediastinum then non-contrast computerized tomography CT- chest revealed a big anterior multi-lobulated mass inside the pericardial sac with a diameter of about 8cm x 6cm with adipose tissue and occasional calcific areas in the middle were observed (figure 1). So after discussing the case between the neonatologist and the cardiothoracic surgery team the consensus was urgent surgical intervention to take out this mass. In cardiothoracic operating room and under general anesthesia, median sternotomy was done then the pericardial sac was opened and the mass was removed completely in the 2nd day of age. The mass was originating from the adventitia of the anterior surface of the proximal part of the ascending aorta and was pedunculated anterior-lateral to the heart with a pedicle.

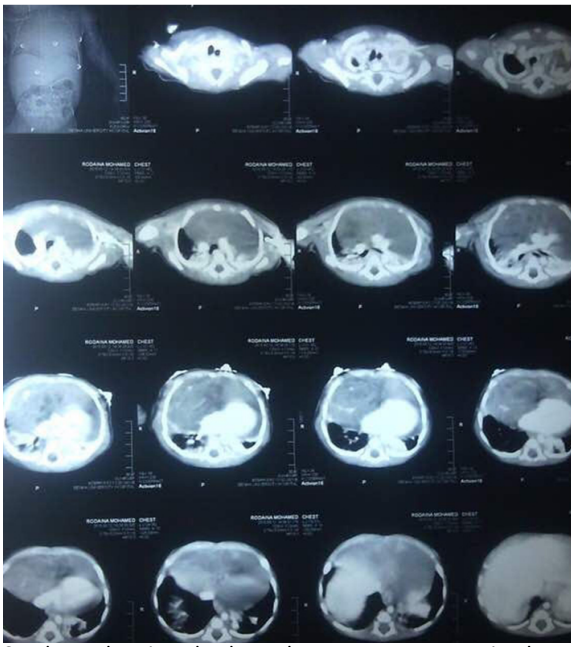

Fig 1: CT chest showing the huge heterogonous mass in the anterior mediastinum with calcification inside

The tumor was successfully separated from the surrounding structures and removed from the thoracic cavity (figure 2).

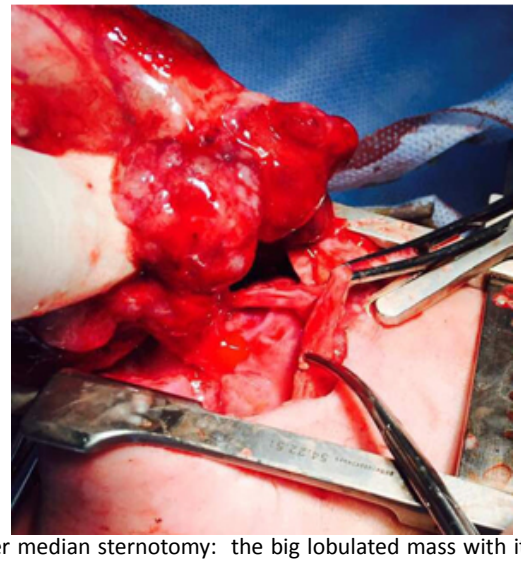

Fig2: after median sternotomy: the big lobulated mass with its pedicle attached to the adventitia of the ascending aorta.

The resected specimen was about 8.5 cm long and 6 cm wide (figure 3).

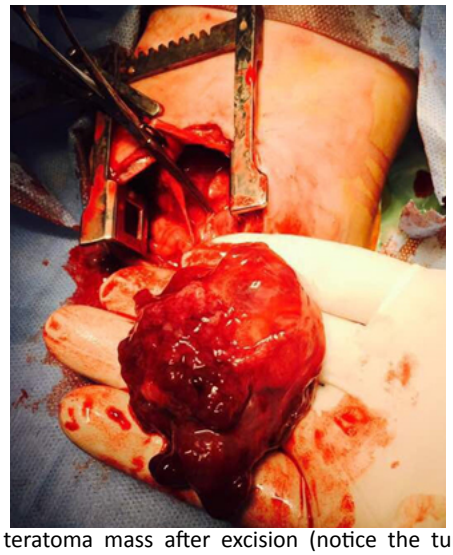

Fig 3: huge teratoma mass after excision (notice the tumour size in comparison to surgeon's hand size and to the bay body size).

The postoperative course was uneventful.

The baby who could be extubated successfully 2nd day after operation was continued to be followed up in the intensive care unit. Respiratory and hemodynamics condition improved dramatically.

The infant was discharged after four days without any complaints.

Histopathology report came out as predominantly solid with areas of cystic changes. Multiple sections studied from tumor showed mature as well as immature elements derived from all 3 germ layers. Mature elements comprised of glands, mature cartilage, and neural tissue. Immature elements included neuroepithelial

elements, neuroectodermal rosettes, and immature cartilage. Final diagnosis was immature mediastinal teratoma free of malignant elements.

Uneventful recovery followed without recurrence.

Discussion

An antero-superior mediastinal mass can be caused by neoplastic and non-neoplastic pathology. Clinical differential diagnoses for an anterior mediastinal mass includes: thymic, thyroid or parathyroid tumours, germ cell tumours including teratomas and teratocarcinoma, mediastinal seminoma, emberyonal cell carcinoma, mediastinal yolk sac tumor, choriocarcinoma or mixed cell type germ cell tumor or vascular origin like aortic aneurysm [11,12].

Primary cardiac tumors in pediatric population are rare with reported incidence of 0.17-0.28% as per echocardiographic or autopsy series [9,13]. Although, majority of such tumors are benign (90%), the frequency and type of cardiac tumors in this age group is different from the adult population [13,14]. Rhabdomyoma is the most common benign cardiac tumor in children, representing more than 60% of primary tumours, followed by teratoma, fibroma and haemangioma [7,15]. Echocardiography, Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) of the thorax are the non-invasive diagnostic tools but histopathology examination remains the conclusive evidence [8,16].

Teratomas are observed most frequently in the sacrococcygeal region. While sacrococcygeal and head-neck teratomas are usually observed in the first two months of life, mediastinal teratomas presents earlier and may lead to complaints at birth, as seen in our case due to compression on the heart and great vessels and airway [7,17].

Teratomas are divided into two groups as mature and immature teratomas [1,3,17], the histopathological examination of our case revealed both mature and immature tissues.

Polyhydramnios and the severity of cardiac or respiratory distress correlate with the size of the teratoma. Large lesions cause cardiac tamponadeor respiratory distress and hemodynamic instability due to possible compression of structures in the medistinum including the heart, great vessels, and trachea and may lead to esophageal obstruction, swallowing disturbance and polyhydramnios [18].

The main therapy of teratoma is complete surgical excision which depends on the site of the tumor. The prognosis is excellent, recurrences are rare, and in our understanding, recurrence may occur due to incomplete surgical resection.

The recurrence can occur in less than 10% of operated patients and can be treated with further surgery or chemotherapy [18,19].

Regarding the treatment of immature teratomas, Marina et al. found in a retrospective study of seventy-three children with extra-cranial immature teratomas that more than 85% of patients can be effectively treated with surgical resection alone and close observation without chemotherapy [19].

Follow-up is based on clinical examination, CT and MRI, especially in case of incomplete excision. Alpha-fetoprotein quantification is recommended by some authors [2,4,5,19].

Conclusion

The prenatal screening should use appropriate imaging tools for early detection of such tumors. Congenital mediastinal teratomas are usually benign. Surgery is the treatment of choice, and should be undertaken on an urgent basis, especially in a patient who presents with signs and symptoms of cardiac tamponade, respiratory distress or airway obstruction.

Complete resection of such tumors is recommended due to malignant transformation, potential rupture, potential recurrence and compression of mediastinal structures.

This case highlights the significant respiratory distress that can occur in newborns with mediastinal teratomas and confirms the need for emergency surgery in this group of patients.

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