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Case Report

A rare case of intra-pericardial teratoma presenting as a mediastinal mass in an infant

Maj Vikram Singh^{a,*}, Brig Sunita Kakkar, vsm^b, Lt Col Anand Arora^c,
Brig Anurag Garg^d, Brig M.M. Harjai^e

^a Resident, Department of Pathology, Armed Forces Medical College, Pune-40, India

^b Consultant (Pathology & Oncopath), Command Hospital (Southern Command), Pune-40, India

^c Graded Specialist (Pathology), Military Hospital (Cardio Thoracic Center), Pune-40, India

^d Professor & HOD, Department of Cardio & Vascular Surgery, Military Hospital (Cardio Thoracic Center), Pune-40, India

^e Commandant, 166 Military Hospital, C/o 56 APO, India

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Introduction

Tumours of the heart are uncommon lesions, their estimated frequency ranges from 0.0017 to 0.33%.¹ They are known to cause a variety of cardiac and systemic symptoms. Clinical symptoms depend on the size and the anatomic location. Small and benign neoplasms may have devastating clinical consequences if present in a critical location. Growth rate, friability and invasiveness are also important factors that determine both the clinical features and outcome. Intrapericardial teratoma is a rare congenital condition, it may be diagnosed in utero or soon after birth due to its association with massive pericardial effusion.²

We report a case of intrapericardial mature teratoma, which was detected in a 6-month-old male child presenting

with respiratory distress due a mediastinal mass. The tumour was excised completely, and histopathological examination of the resected tumour was suggestive of mature teratoma. The patient was asymptomatic three months postoperatively.

Case report

A 6 months old male patient was brought to paediatric outpatient with complaints of respiratory distress. On general physical examination, the child was afebrile and active with a regular heart rate of 138/min. There was no pallor, icterus, cyanosis or lymphadenopathy present.

Systemic examination was within normal limit. All the haematological and biochemical parameters were also within normal limit. Radiograph chest showed a homogeneous opacity in right side of chest with smooth convex lateral margins. Medial margins could not be seen and there was loss of cardiac silhouette over right side (Fig. 1A).

Echocardiography showed a visible mass abutting on the cardiac chambers.

Computerised Tomography (CT) thorax revealed: multicystic anterior mediastinal mass, measuring 6 × 6 × 4 cm, which was well circumscribed, multiloculated and intra-pericardial in location. It was associated with mild pericardial effusion and located in close proximity to aorta, right atrium and right ventricle (Fig. 1B).

* Corresponding author. Tel.: +91 9373648747.

E-mail address: dr_vikramsingh@rediffmail.com (V. Singh).

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Fig. 1 – Imaging findings: (A) Radiograph chest showing loss of cardiac silhouette over right side, (B) Computerised tomography chest showing multicystic anterior mediastinal mass.

Based on these radiological findings possibilities of cystic hygroma, thymic cyst and cystic teratoma were considered.

Surgical excision of the mass was done through a median sternotomy. A cystic, lobulated intrapericardial mass measuring $5 \times 5 \times 3$ cm was seen. The vascular pedicle attached to the aorta was ligated and mass was dissected off the pericardium and other cardiac structures (Fig. 2A and B).

The cut surface of the mass showed a partly solid and partly cystic mass with variegated appearance and hemorrhagic areas. Microscopy showed multiple cysts lined by stratified squamous epithelium, gastric mucosa and respiratory epithelial lining at places. Numerous sebaceous glands, sweat glands, islands of mature hyaline cartilage and mature glial tissue were also present (Fig. 2C and D). No immature neuroepithelium seen. Post op period was uneventful. The patient was asymptomatic three months postoperatively.

Discussion

Primary tumours of the heart in infants are rare. Germ cell tumours are neoplasm of germ cell origin, which are classified histologically into seminoma (dysgerminoma), embryonal carcinomas, yolk sac tumour, choriocarcinoma and teratoma.

Patient age ranges from intrauterine life to 66 years.³ Teratomas generally occur in infants while adults tend to have malignant germ cell tumours. Over 75% of cardiac teratomas occur in children under the age of 15. Main clinical features include respiratory distress, pericardial tamponade, and cyanosis.

Teratomas in newborns usually occur in the sacrococcygeal area, testis and ovaries and less frequently in the neck or intracranium. Intrapericardial teratomas are rare, approximately less than 100 case reports are available in the literature.⁴

Teratomas are embryonal neoplasms containing three germ layers (endoderm, ectoderm, and mesoderm).⁵ Most of these are mature teratomas and very few cases are of immature type.⁶

Today a fair number of intrapericardial teratomas are diagnosed prenatally during routine ultrasound scans, which

provide an opportunity to prevent complications that would otherwise develop before birth in approximately 50% of cases. The foetal ultrasound scan usually shows large pericardial effusion with an intrapericardial mass. The mass appearance can be multilobulated and cystic with calcified areas. Foetal MRI can add information supporting the diagnosis.⁷

Intrauterine pericardiocentesis, which was first performed in 1992, can avert imminent cardiac tamponade and development of hydrops, thus allowing a full-term birth.⁸

Cardiac teratomas may be massive, measuring up to 15 cm.¹ The tumours usually displace the heart and rotate it along its longitudinal axis. Intrapericardial teratomas are usually located on the right side of the heart, displacing the organs to the left and posteriorly; those located on the left side produce the opposite effect.¹

Teratomas are usually attached by a pedicle to one of the great vessels with arterial supply directly from the aorta, similar findings were also present in our patient. The histological structure of an intrapericardial teratoma does not differ from teratomas in other sites. Most of them consist of endodermal (e.g. gastric, intestinal mucosa), mesodermal (e.g. bone, cartilage, fatty or fibrous tissue) and neuroectodermal structures (e.g. neuroglia), in varying proportion.^{9,10}

Interestingly, one report of an intrapericardial teratoma containing pancreatic tissue with endocrine activity has also been published.¹¹

Intrapericardial teratomas are usually benign tumours but may be life-threatening because of large pericardial effusion and cardiac compression.

Surgical excision is the only effective treatment for cardiac teratoma. Once resected, the prognosis is usually good, as most tumours are benign and require no further treatment. Morphology and clinical behaviour of the immature intrapericardial teratoma described by Agozzino et al¹² 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.

Fig. 2 – Intra Op and histopathological examination findings: (A) Intrapericardial mass measuring $5 \times 5 \times 3$ cm (B) Gross image of cut section of resected tumour mass (C) Photomicrograph (Haematoxylin and Eosin stain: 400 $\times$) showing cyst lined by stratified squamous epithelium, sebaceous glands, sweat glands and glial tissue, (D) Photomicrograph (Haematoxylin and Eosin stain: 400 $\times$) showing hyaline cartilage.

To conclude, we have presented a case of an intra-pericardial teratoma, a rare congenital tumour in a 6-month-old baby who presented with respiratory distress and found to have a mediastinal mass. The importance of considering this diagnosis, though uncommon is stressed here, as it has a much better prognosis and surgery is curative.

Conflicts of interest

All authors have none to declare.

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