Recurrent Cardiac Tamponade: Intrapericardial Teratoma

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Intrapericardial teratoma is a rare entity usually presenting in infancy with a respiratory distress, pericardial effusion and cardiac tamponade. We report a 2-month-old male child with a intrapericardial teratoma with an unusual presentation of recurrent pericardial effusion and tamponade, who was successfully treated by the surgical, resection of the teratoma.

Case Report

A 2-month-old male infant was brought with a history of cough and breathlessness for past 20 days. On physical examination, weight was 3.5 kg, general condition was fair, temperature was normal, pulse rate was 160/min and respiratory rate was 80/min. There was no cyanosis but intercostal and subcostal retractions were present. Jugular venous pressure and cardiac examination was normal except for the cardiomegaly which was revealed on percussion. Liver was just palpable below the costal margin and was not tender. X-ray chest revealed enlarged cardiac shadow with cardio-thoracic ratio of 0.80 suggestive of pericardial effusion. 2D Echocardiography confirmed the presence of pericardial effusion and also revealed a hyperdense shadow in the region of right atrium suggesting a differential diagnosis of either a neoplasm or hematoma either in or around right atrium. Emergency pericardiocentesis was carried out to relieve the cardiac tamponade. The aspirated pericardial fluid was straw colored transudate with proteins of 3 g/dl and absence of cells suspecting an intrapericardial neoplasm. CT Scan guided biopsy (Fig. 1) of the mass was planned to know the histopathological diagnosis and also to rule out malignancy as cardiac tamponade recurred frequently.

After the biopsy, the frequency of tapping required increased further and hence open drainage with surgical excision of the mass was carried out. At surgery a 8 x 7.5 x 4.5 cm pedunculated bossilated mass compressing the right atrium and lying within the pericardium was excised. On histopathology the mass was diagnosed to be a benign intrapericardial teratoma as it showed the cells derived from all three germinal layers including adipose tissue, cartilage, gland cells and bone tissue (Fig. 2).

The patient is asymptomatic for 1 year after the surgery, and X-ray chest has shown a normal heart. The patient has not followed up after that.

Discussion

A teratoma is composed of cells from more than one germinal layer. The tumor is
usually located in midline, an area in the embryo where all three germinal layers may be found. The intrapericardial teratoma usually originate from myocardium or the adventitia of great arteries. Pericardial effusion is found exclusively in those patients in whom intrapericardial teratoma arises from the adventitia of great vessels(1).

Intrapericardial teratoma is a rare but important cause of cardiorespiratory distress in infants and young children(2,7,10). It is a benign tumor and ranks second in the anterior mediastinal tumors presenting during infancy. The cardiorespiratory symptoms may result from (i) compression of the heart or lungs by the intrapericardial mass; (ii) compression of the heart and lungs by a pericardial effusion associated with the mass; or (iii) pulmonary infection secondary to respiratory tract obstruction(l).

Clinical findings among patients with intrapericardial teratoma vary with the patient's age. Only infrequently do the adults with intrapericardial teratoma have associated acute pericardial effusion or tamponade(4). Patients beyond 3 years of age, in fact are usually asymptomatic or have chronic pericardial effusion. In contrast, patients 3 months old or younger invariably have acute pericardial effusion with tamponade. In young infants, the association of acute effusion and tamponade is so consistent that intrapericardial teratoma gets ruled out as a cause of cardiac difficulty if such effusion is absent. If the pericardial fluid is non bloody, sterile and recurrent, intrapericardial teratoma is the most likely cause(2,11).

Intrapericardial teratoma should be suspected in any infant who presents with respiratory distress, cyanosis, superior vena cava obstruction or cardiac tamponade. On
physical examination, apical impulse and other signs of precordial activity are conspicuously absent\(^7\). The heart sounds are muffled. The ECG may show low voltage and non specific T wave changes. The chest X-ray film reveals various degrees of cardiomegaly suggestive of pericardial effusion. The diagnosis can be made by

Fig. 2. Histopathological picture of teratoma: (A) glandular (endodermal tissue), and (B) cartilage (mesodermal tissue).
echocardiography, but cardiac catheterization may be required for determining the extent of intracardiac lesions(10). CT Scan and CT Scan guided biopsy can be helpful as in our case, in determining the nature of the tumor.

The differential diagnosis includes other causes of anterior mediastinal masses, thymic tumors and other causes of pericardial effusion and primary heart lesions(10). Amongst the intrapericardial tumors, intrapericardial teratoma is only second to rhabdomyoma which is the commonest intrapericardial tumor.

Complications usually seen with intrapericardial teratoma are cardiac tamponade, malignant changes, respiratory infections secondary to obstruction and esophageal obstruction(5,6).

Surgical removal is the treatment for intrapericardial teratoma. An important piece of information that the tumor is pedunculated and arises from the aorta or both great arteries is helpful to the surgeons. Since the tumor is usually benign, operation is almost curative and provides prompt relief of symptoms(7,10). Deaths, if at all, occur generally from hemorrhage during operative removal of the tumor, severe cardiac tamponade or malignancy.

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