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Case Report

A rare case of cardiac lymphangioma

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ABSTRACT

Introduction: Primary cardiac tumors are rare, their incidence ranging from 0.0017% to .33% at autopsy.[1] Cystic lymphangioma, usually confined to head and neck, is a well-recognized benign tumor that occurs during childhood.[2] However, a cardiac lymphangioma is exceptionally uncommon and a particularly rare form of cardiac disease.

Case report: A 22-year-old girl admitted with accidental trauma and dyspnea on exertion since childhood. Chest X-ray showed right hydropneumothorax and mild cardiomegaly. MDCT chest showed right hydropneumothorax and a cystic mass which was present on the left side anterior to the heart. Then Echo was done which showed normal left ventricular function, normal great vessels, chambers and valves. A 12 cm × 8.5 cm mass was seen in the pericardium over right ventricle (RV) compressing it with no features of obstruction. Cardiac MRI revealed a large hyperintense lesion of 14 cm × 8 cm × 12 cm in anterior mediastinum with multiple septations which was present anteriorly to right atrium (RA) and right ventricle, compressing RA and RV. MDCT coronary angiogram RCA running through the mass. During surgery, after opening the pericardium a cystic mass of 20 cm × 15 cm × 15 cm was found adherent to RA and RV moving with the heart. About 700 ml of fluid was aspirated and mass was opened up which showed multiple strands and trabeculations. Mass was completely excised except the wall adherent to RV. RCA was transected and end to end anastomosed with saphenous venous graft. Histopathology of biopsied mass confirmed it as cardiac lymphangioma containing lymphatic spaces lined by single layer of endothelium.

Discussion: Cardiac lymphangiomas are very rare. Cardiac lymphangioma are most often diagnosed incidentally by chest X-ray presenting as asymptomatic masses. However, they may cause congestive heart failure, syncopal or embolic pathology, arrhythmias, palpitations, or cardiac tamponade. Majority of symptoms are accredited to their size and location. These tumors most commonly occur in the pericardial space, but other unusual primary sites include the myocardium, the posterior wall of the left atrium, and AV node regions.[3–5] At MRI, these tumors may have a high signal intensity on T1 weighted images, perhaps due to the presence the proteinaceous material in the lymph within the stroma and cystic spaces.[6] Cystic lymphangioma is typically a multiloculated lesion with cystic cavities divided by the septa of variable thickness. Cardiac lymphangiomas have a possible risk of recurrence, especially if there has been incomplete resection. Therefore, extensive resection is recommended.[7]

Conclusion: Imaging with MDCT and MRI aid in the proper diagnosis of pericardial lymphangioma. A meticulous planning is required for surgical excision of this lesion. We report this case because of its rarity and to highlight the role of imaging and the surgical challenge in view of its location and proximity to coronary vessels.

1. Introduction

Primary cardiac tumors are rare, their incidence ranging from 0.0017% to 0.33% at autopsy. Benign tumors account for 75% of all primary cardiac tumors, of which 75% are myxomas.[1] Cystic lymphangioma, usually confined to head and neck, is a well-recognized benign tumor that occurs during childhood.[2] However,

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a cardiac lymphangioma is exceptionally uncommon and a particularly rare form of cardiac disease, and is considered to be a malformation that arises from sequestration of lymphatic tissue that fails to communicate normally with the rest of the lymphatic system. Cardiac lyphangiomas most frequently occur in the pericardial space, sometimes compressing adjacent structures. This case highlights its atypical clinical picture and diagnostic uncertainty until surgical exploration.

2. Case report

A 22-year-old girl admitted with accidental trauma in casualty department. She had some stab injury in her chest following which a Chest X-ray was taken which showed right hydropneumothorax and mild cardiomegaly. ECG showed sinus bradycardia 58/min and T inversion in leads II, III & avf & V1-V6 (Fig. 1). Then immediately an intercostal tube drain was put inside right chest. On next day a MDCT chest was taken which showed right hydropneumothorax and a cystic mass which was present on the left side anterior to the heart (Fig. 2). She had history of dyspnea on exertion since childhood. She was treated for hydropneumothorax and 4 months later another MDCT chest was taken which showed resolution of hydropneumothorax however that mass persisted. She was advised further investigations but she lost on follow up. After one and half year she admitted in obstetrics department with second month gestation for her check-up but her prior records led her back to cardiology for further evaluation. Then Echo was done which showed normal left ventricular function, normal great vessels, chambers and valves. A 12 cm × 8.5 cm mass was seen in

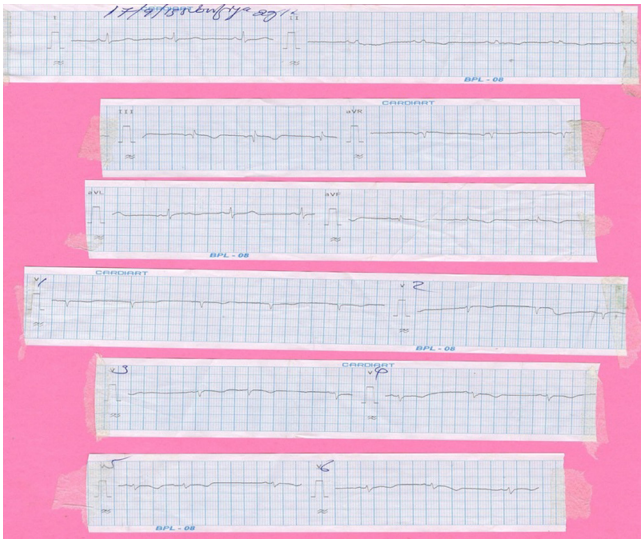

Fig. 1. Electrocardiogram of the patient.

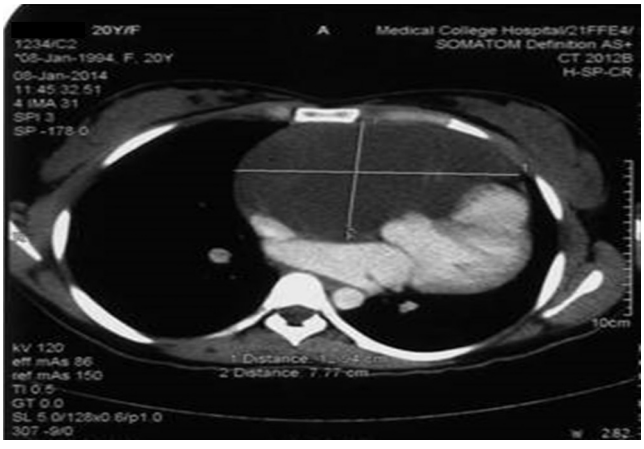

Fig. 2. MDCT chest showing large cystic mass present in anterior mediastinum.

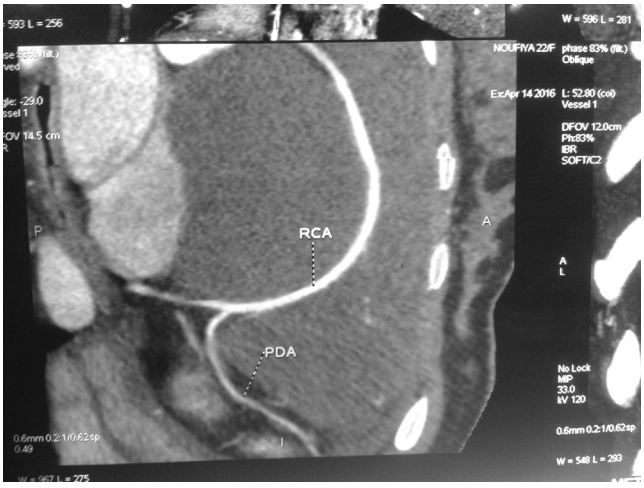

Fig. 3. CT coronary angiogram showing course of right coronary artery through the tumor.

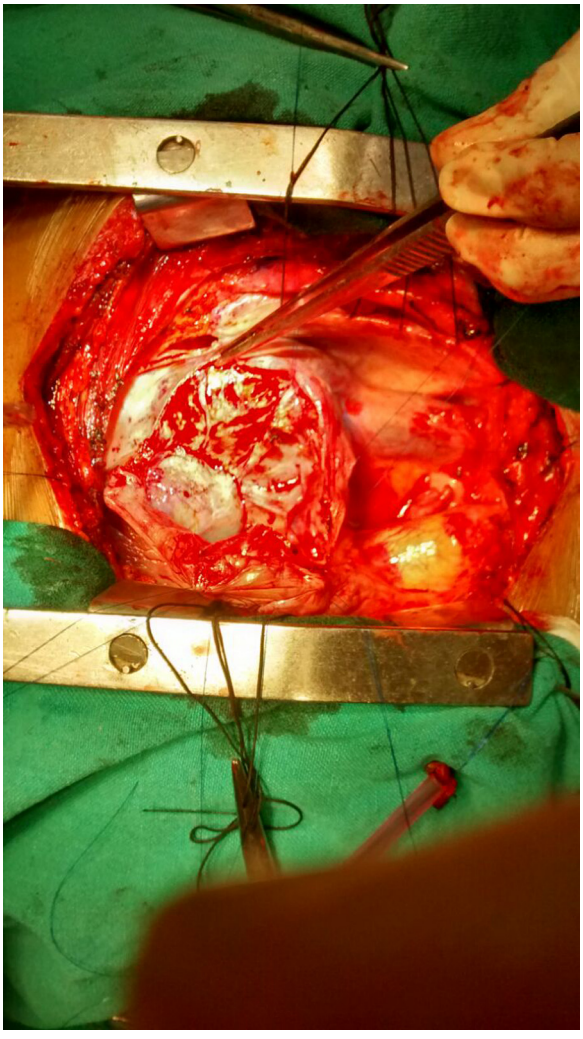

Fig. 4. Opened mass showing multiple strands and trabeculations.