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Case Report

Cystic lymphangioma of pericardium presenting as isolated chylopericardium – A case report

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ABSTRACT

Isolated chylopericardium due to cystic lymphangioma of pericardium is a rare entity. We report a case of asymptomatic chylopericardium in a young male who presented with cardiomegaly. Echocardiography revealed massive pericardial effusion without tamponade. Pericardiocentesis yielded 1.25 L of tea-colored fluid which showed triglyceride level of 1723 mg/dL and cholesterol of 1021 mg/dL with a cholesterol to triglyceride ratio of <1, characteristic of chylous fluid. Lymphoscintigraphy using 99Tc demonstrated lymphatic leak around the heart region. Fusion of MRI images with lymphoscintigraphy was taken with a view of localizing the leak site; it demonstrated enhancement in the pericardial space. Surgery was done via right lateral thoracotomy. Thoracic duct was ligated above diaphragm and pericardial window created by anterior pericardiectomy. The patient had an uneventful recovery and was well after 6 months of follow up. Pericardial biopsy showed cystic lymphangioma of pericardium.

An 18-year-old male was referred to our hospital for asymptomatic cardiomegaly (Fig. 1A). There was no history of trauma, thoracic surgery or neoplasm. Clinical examination was unremarkable except for distant heart sounds. Electrocardiography showed low voltage complexes. Echocardiography revealed massive pericardial effusion without tamponade (Fig. 1B). Pericardiocentesis yielded 1.25 L of tea-colored fluid (Fig. 1C). Aspirated pericardial fluid had lymphocytic predominance with numerous RBCs; fluid protein was 7 gm/dL, sugar 116 gm/dL and ADA 21U /L; there were no malignant cells, bacterial growth on culture or trophozoite or bacilli on gram stain. Mantoux test and antinuclear antibody were negative. Laboratory tests demonstrated normal blood counts, serum electrolytes, serum lipid profile, liver function, serum urea, creatinine, calcium and phosphate. Pericardial aspirate also showed triglyceride level of 1723 mg/dL and cholesterol of 1021 mg/dL with a cholesterol to triglyceride ratio of <1, characteristic of chylous fluid. High resolution computed tomography did not show any mediastinal mass. Lymphoscintigraphy using $_{99}$Tc demonstrated lymphatic leak around the heart region (Fig. 2A). Fusion of MRI images with lymphoscintigraphy was taken with a view of localizing the leak site; it demonstrated enhancement in the pericardial space (Fig. 2B). Patient was kept on low fat medium-chain triglyceride diet. Since there was no reduction in the daily aspirate, surgery was done via right lateral thoracotomy. Thoracic duct was ligated above diaphragm and pericardial window created by anterior pericardiectomy. The patient had an uneventful recovery and was well after 6 months of follow up. Pericardial biopsy showed cystic lymphangioma of pericardium (Fig. 2C).

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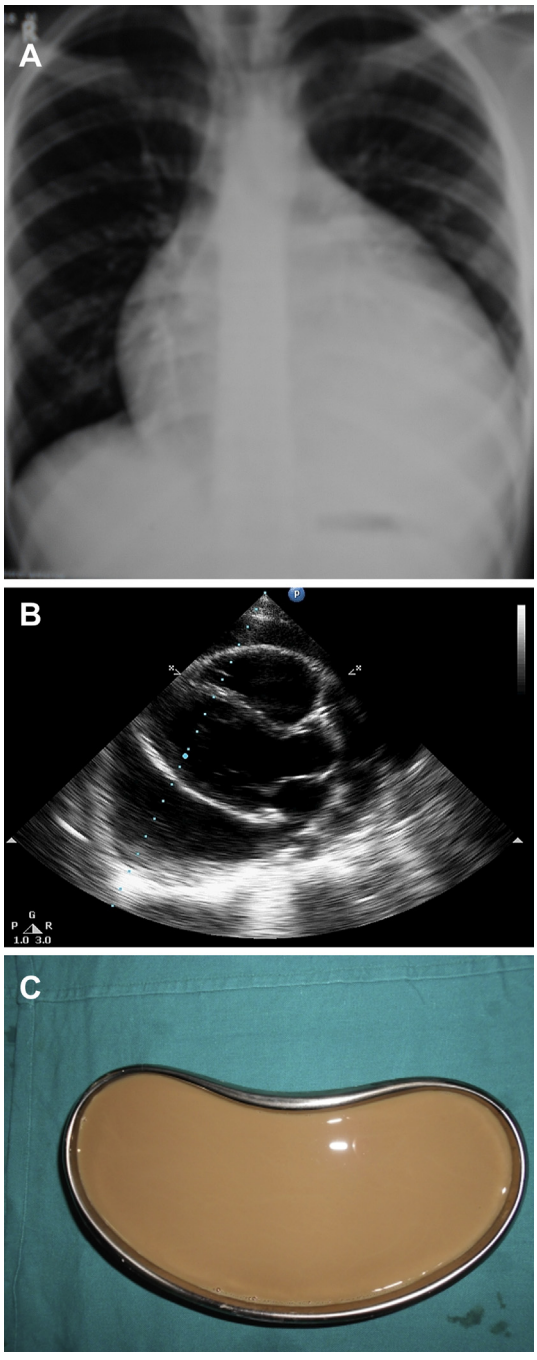

Fig. 1 – (A) X-ray of the patient showing cardiomegaly, (B) Echocardiogram showing pericardial fluid, (C) Tea-colored pericardial aspirate.

Chylopericardium, first described by Hasebrock in 1886, is a rare entity. It may be a consequence of thoracic and cardiac surgery or as a result of chest trauma, mediastinal tuberculosis, mediastinal neoplasm, mediastinal radiotherapy or thrombosis of subclavian vein. The term primary isolated chylopericardium was first reported by Groves and Effler in 1954.[1] Abnormalities of the lymphatic system and mediastinal lymphangiectasia causing chylopericardial effusions are referred to as idiopathic chylopericardium. Age at diagnosis ranges from 18 to 68 years.

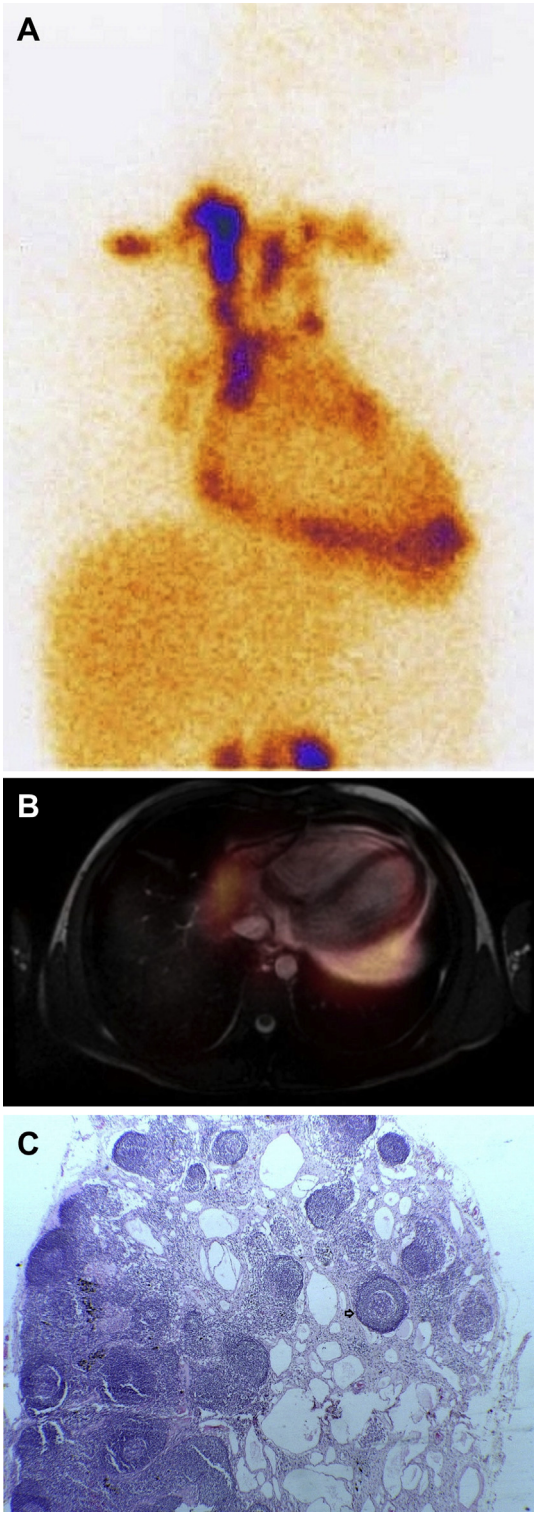

Fig. 2 – (A) Lymphoscintigraphy showing Tc_{99} radioactivity around the heart, (B) Fusion MRI and scintigraphy image showing seepage of Tc_{99} in the pericardial space, (C) Photomicrograph of the histology of lymphangioma pericardium showing lymphoid follicles (arrow) and cystic spaces.

Clinical presentation may vary from incidental detection of cardiomegaly (as in our case) to those presenting with dyspnea, fatigue, or cardiac tamponade.[2] Characteristics of chylous fluid include a milky yellowish appearance, triglyceride level >500 mg/dL, cholesterol-triglyceride ratio of <1 and lymphocyte predominant fluid with negative cultures. In our patient, the fluid was tea-colored, probably owing to mixing of red blood cells (RBC). Diagnosis of chylopericardium can be made non-invasively by precordial imaging using $_{99}$Tc-labeled RBC or oral administration of $_{131}$I-triolein. Lymphangiography may be helpful in identifying fistulous communications as well as anatomy of thoracic duct which is well known for its variations.[3] In our case we used $_{99}$Tc-sulfur colloid for lymphoscintigraphy which showed uptake of the radiopharmaceutical around the heart region suggesting chylopericardium. MRI fusion lymphoscintigraphy confirmed the leakage of chyle into pericardial cavity. Our case is the first case where fusion imaging was used for localization of chylous fluid leakage into pericardial space.

Cystic lymphangioma is a childhood tumor usually confined to head and neck. Cardiac lymphangiomas are exceptionally uncommon. First described by Armstrong and Monkeberg in 1911, most of them occur in the pericardial space.[4] To the best of our knowledge only ten cases have been reported in medical literature. Most often detected incidentally on chest X-ray as asymptomatic masses, cardiac lymphangiomas may cause congestive heart failure, syncope, arrhythmia or cardiac tamponade. Absence of RBC in lymphatic spaces differentiates it from hemangioma or lymphohemangioma. In the case reported by Zakaria et al[4] pericardial lymphangioma detected in a 1-year-old child presented with recurrent episodes of cough & worsening respiratory distress and was not associated with chylopericardium. In another case by Naz et al pericardial lymphangioma was found as a large mass presented with dyspnea on exertion, productive cough and heaviness of chest in a 32-year-old female.[5]

In contrast to post-traumatic cases, conservative treatment is rarely helpful in primary chylopericardium. Operative procedure consists of ligature and excision of thoracic duct just above diaphragm combined with partial pericardiectomy. Pericardiectomy is done to ensure complete drainage and to prevent development of constrictive pericarditis.[6]

We report this case in view of its rarity and being the first Indian case report of cardiac lymphangioma presenting as chylopericardium. We believe that fusion imaging may be of help in accurately localizing the site of leak and identifying the site of collection of chylous fluid.

Conflicts of interest

All authors have none to declare.

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