Primary chylopericardium due to lymphangiectasias: the crucial role of lymphangiography

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A 24-year-old woman was referred to our Cardiology Department because of cardiomegaly at a routine chest X-ray. She was asymptomatic except for some fatigability. Physical examination was unremarkable. Electrocardiogram showed only low QRS voltages. Echocardiography revealed large pericardial effusion with right atrial and ventricular collapse. Pericardiocentesis was performed and 1100 mL of milky fluid were removed, suggesting chylopericardium. The chylous nature of the fluid was confirmed by high content of triglycerides (1200 mg/dL) and by cholesterol/triglyceride ratio < 1. Sudan III stain of the fluid revealed fat globules. Bacterial and tuberculous cultures were negative. Cytology demonstrated abundance of lymphocytes, with no tumour cells. A subxiphoid exterior tube drainage was maintained and alipidic diet was started. After an initial success of this treatment, significant chylopericardium recurred. Thoracic computed tomography was negative. Thus, lymphangiogram was performed by slow injection of ethiodized oil into a cannulated lymphatic vessel of the right foot showing an open thoracic duct with lymphangiectasias at pericardial level (Figure). Left thoracotomy was indicated. The pleural cavity appeared normal; the pericardium appeared thickened with numerous lymphangiectasias. Ligation of the thoracic duct, closure between metallic clips of lymphangiectasias, and pericardial fenestration were performed. Pericardial biopsies showed a chronic inflammatory process with haemorrhagic infiltrate and a large number of lymphangiectasias. After 15 days, the patient was discharged and in 4 weeks she returned to normal diet, full-time work, and full activities. Echocardiography showed no recurrence of chylopericardium at late follow-up.

Panel A. Ascending phase at abdominal level of the lymphangiographic agent after cannulation of a right foot lymphatic vessel.
Panel B and C (detail). Diagnostic lymphangiogram in antero-posterior view showing the presence of lymphangiectasias at pericardial level (arrows).
Panel D. Diagnostic lymphangiogram in lateral view showing the thoracic duct (arrow heads) and the presence of lymphangiectasias at pericardial level (arrows).