

A contradictory phenomenon of thicken pericardium and cardiac compression without inferior vena cava dilation: sign of IVC escape

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A 77-year-old female presented with shortness of breath and tightness of chest was admitted. Her past medical history included hypertension and she has been taking nifedipine regularly. Two years before, she was diagnosed with pericardial effusion (Figure 1) and had pericardiocentesis drainage. On physical examination, her blood pressure was 151/100 mmHg and her pulse rate was 91 beats/min. Physical examination revealed both lower extremities edema. Electrocardiogram revealed V2-V6 T wave inversion. Cardiac Troponin I was in normal range, less than 0.05 ng/mL (normal range: 0-0.0268 ng/mL), Brain Natriuretic Peptide (BNP) was 670.7 pg/mL (normal range: 0-125 ng/L), Plasma D-Dimer was 1.03 mg/L (normal range: 0-0.5 mg/L). T-SPOT.TB

assay result was negative. Coronary angiography revealed 60% stenosis in the mid right coronary artery, 40% stenosis in the mid anterior descending artery and diffuse atherosclerotic plaque distally. Right heart catheterization examination revealed mean pulmonary artery pressure was 25 mmHg, right ventricular pressure was 26 mmHg, right atrial pressure was 16 mmHg, and superior vena cava pressure was 16 mmHg. Echocardiography showed that the pericardium in front of the anterior wall of the right ventricle was thickened and the thickness is about 14.8 mm, the right ventricular wall and right ventricular outflow tract were compressed, the pericardium behind the posterior wall of the left ventricle was thickened and the thickness is about 7 mm, combined with a small amount of pericardi-

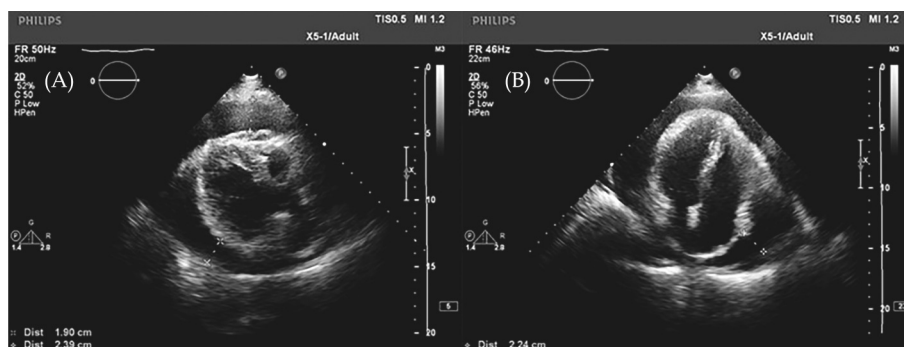


Figure 1 Echocardiography images two years ago. (A): Transthoracic echocardiography showed pericardial effusion in long-axis view; (B): transthoracic echocardiography apical four-chamber view demonstrated pericardial effusion.

al effusion, cardiac contraction was restriction. Contradictory, the inferior vena cava (IVC) was not dilated, about 20 mm wide, and the variability with respiration was more than 50% (Figure 2). Combined with CT result (Figure 3), she was diagnosed with constrictive pericarditis. Then pericardial peel was performed under general anesthesia. The pathological result of pericardial revealed pericardial epithelioid malignant mesothelioma (Figure 4). Programmed cell death ligand 1 (PD-L1) 22C3 staining results showed that Tumor Proportion Score (TPS) was 50% and Combined Positive Score (CPS) was 52. Genetic testing detected five somatic muta-

tions and one embryonic mutation. Subsequently, she was given chemotherapy with pemetrexed and oral olaparib. After two rounds of chemotherapy, her symptoms improved, and her chest CT and echocardiogram results were also better than before (Figure 5).

Pericardial mesothelioma is a rare malignant cardiac neoplasm with early metastasis and poor prognosis.^[1] It may present as congestive heart failure, pericardial effusion or constrictive pericarditis.^[2] A definitive diagnosis of pericardial mesothelioma is often delayed because the disease lacks characteristic findings.^[3] Constrictive pericarditis usually mani-

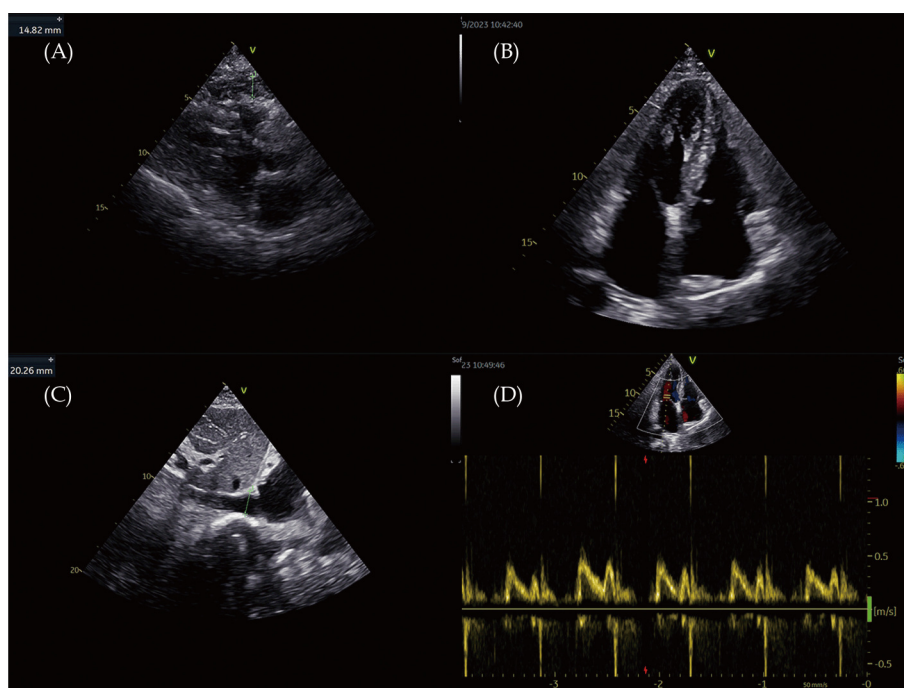


Figure 2 Echocardiography images when the patient was admitted. (A): Transthoracic echocardiography showed pericardial thickening; (B): transthoracic echocardiography apical four-chamber view demonstrated constriction of the pericardium (C) the inferior vena cava was not widened (D) mitral flow changed with respiration.

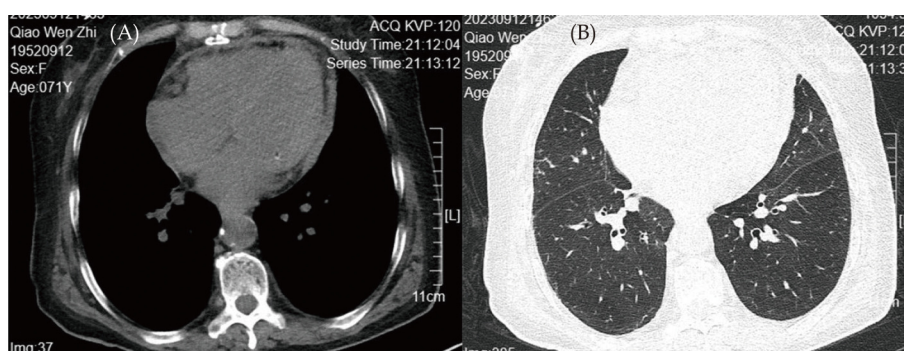


Figure 3 CT images. (A): Initial CT demonstrated thickening of the pericardium; (B): contrast-enhanced CT image.

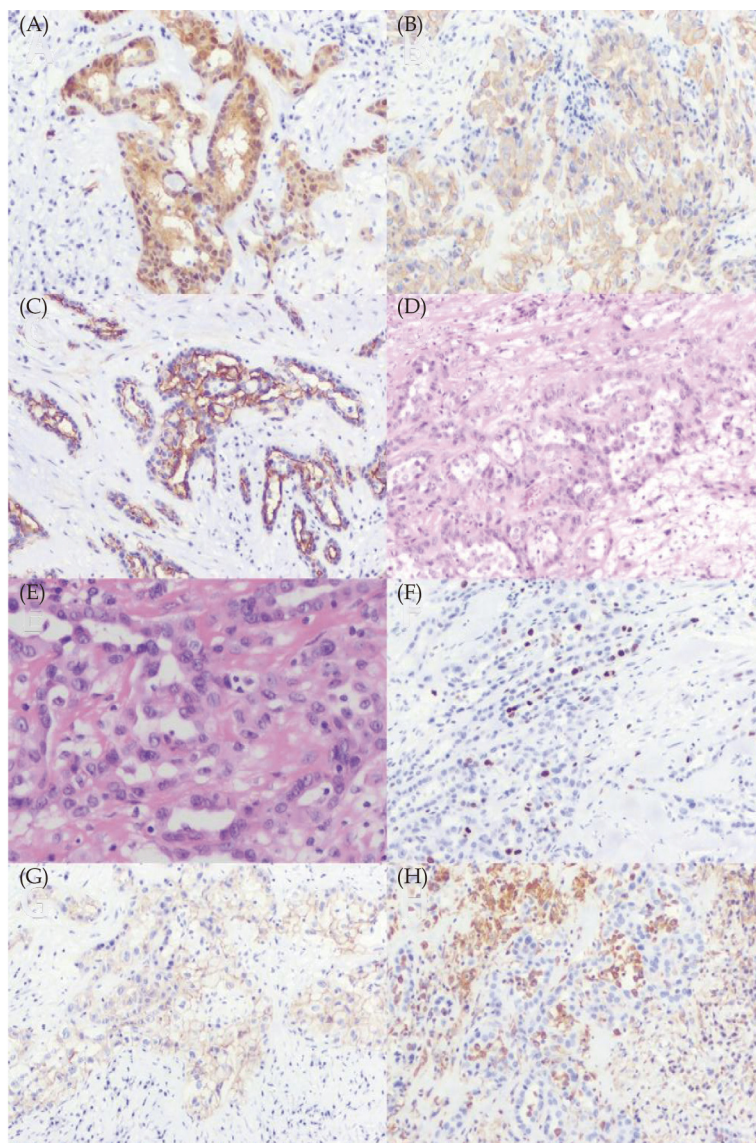


Figure 4 Histopathological images. (A): Immunohistochemical markers for mesothelioma (calretinin); (B): broad-spectrum cytokeratin positive-abnormal proliferation and differentiation of epithelial tissues, and tumors originate from the epithelium (CKpan(+)); (C): characteristic markers of lymphatic endothelial cells (D2-40(++)); (D & E): the tumor tissue was adenoid and small nest-like, the cells were mild, the cytoplasm was red stained, the nucleoli were obvious, and the nuclear division image was occasionally seen (HE stain). The interstitial fibrous tissue is significantly proliferated, collagenized, and a large number of lymphatic and plasma cells are infiltrated; (F): markers of cell proliferation (Ki67(15%+)); (G): adenocarcinoma markers (MOC31(60%)); (H): markers of mesenchymoma (vimentin).

feats as pericardial thickening and highly reflective echo, combined with or without a large amount of pleural effusion, abnormal inspiratory wall movement and markedly dilated hepatic veins.^[4, 5] The IVC is often dilated, and its diameter does not change much in the respiratory system. Patients with constrictive pericarditis have higher mitral diastolic flow velocity when expiratory and lower mitral flow velocity during inspiration. The speed of blood

flow also was found to vary with breathing through the tricuspid valve.^[6] However, the present patient presented with these features of previous pericardial effusion and followed constrictive pericarditis misled the diagnosis. She presented with chest pain and suffocation two years ago. Echocardiography had shown large pericardial effusion, with an ejection fraction of 51%. The tumor marker results were all within the normal range. The patient had been

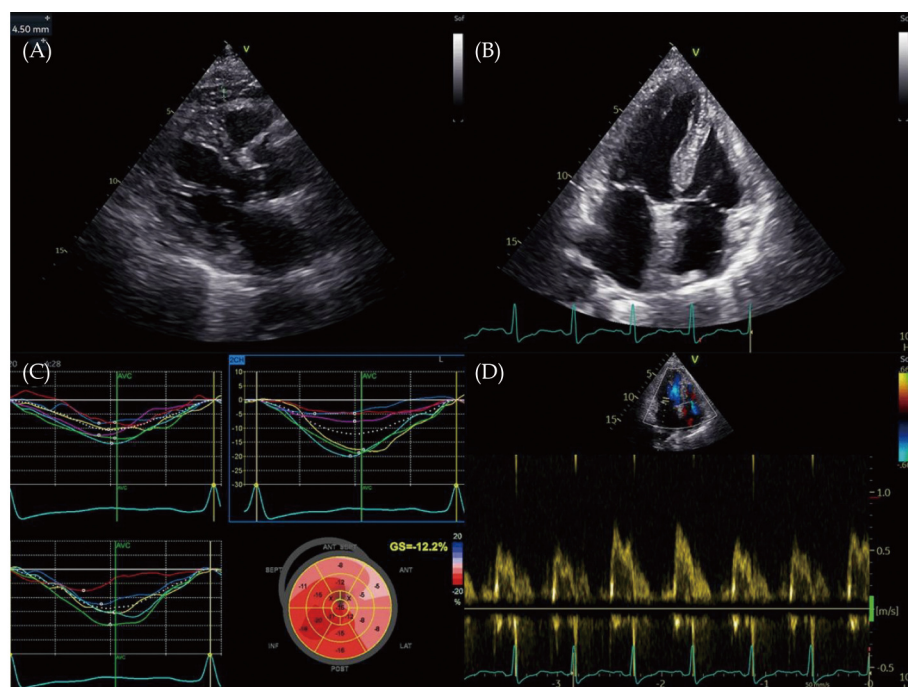


Figure 5 Echocardiographic images after two chemotherapy treatments. (A): Transthoracic echocardiography long-axis view of the left ventricle showed a small amount of pericardial effusion; (B): transthoracic echocardiography showed an apical four-chamber view, the constriction of the pericardium was relief; (C): global longitudinal strain revealed improvement of cardiac function; (D): increased spectral integration of mitral valve inflow blood.

treated with pericardiocentesis. The drainage fluid from the pericardial puncture was yellow and turbid, with a positive result on the Leishman's test and an anti-nuclear antibody (1: 320 nuclear granular type). IgG was elevated, and tuberculosis and bacterial pericarditis were excluded. During the current admission, echocardiography showed a small amount of encapsulated pericardial effusion, pericardial thickening, and cardiac systolic and diastolic restriction, suggesting constrictive pericarditis.^[7] Contradictory, the IVC was not dilated. It was speculated that pericardial adhesions were widely presented in constrictive pericarditis, while tumor compression was mostly localized. We named the echocardiographic sign-- IVC escape.

Pericardial mesothelioma has a low incidence and atypical symptoms and signs, making early definitive diagnosis difficult.^[8] Echocardiography can detect pericardial effusion and other abnormalities in the early stage.^[9] It should be alerted the possibility of tumor compression when the contradictory phenomenon of thicken pericardium and cardiac compressed without dilated IVC or the sign IVC escape was found.

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