Scleroderma heart: pericardial effusion with echocardiographic signs of tamponade during pregnancy

Sirs,

A 33-year old woman with a 2-year history of diffuse systemic sclerosis (dSSc) and no evidence of internal organs involvement apart from gastroesophageal reflux was followed up in our department, for a small pericardial effusion without compression signs. The patient had previously been treated with penicillamine along with a channel calcium blocker. Inflammatory indexes and renal function were normal. Immunology tests were positive for anti-topoisomerase antibodies. There was no serological evidence of bacterial or viral infection. The diagnosis was mild pericardial effusion related to dSSc. The patient became pregnant after IVF procedure. At the 34th week of gestation, she complained of exertional dyspnea and mild ankle oedema. The echocardiogram showed a mild to moderate pericardial effusion, without hemodynamic compromise, but with signs of tamponade, *i.e.* paradoxical movement of the anterior right ventricular wall and right atrial free wall (Fig. 1, panels A and B, respectively). Because of the posterior localisation and the absence of foetal maturity, the therapeutic decision was to follow-up in the intensive care unit. At the 36th week of pregnancy, while a repeat echocardiogram revealed an increase in pericardial effusion, she underwent successfully elective caesarean section. The patient was then successfully treated with 0.5mg per day of colchicine and 50mg per day of captopril. The duration of colchicine treatment was 3 months. At this time a repeat echocardiogram was performed and the medication was stopped. The patient continues on 50mg per os of captopril and she is under follow-up with echocardiography study every 6 months.

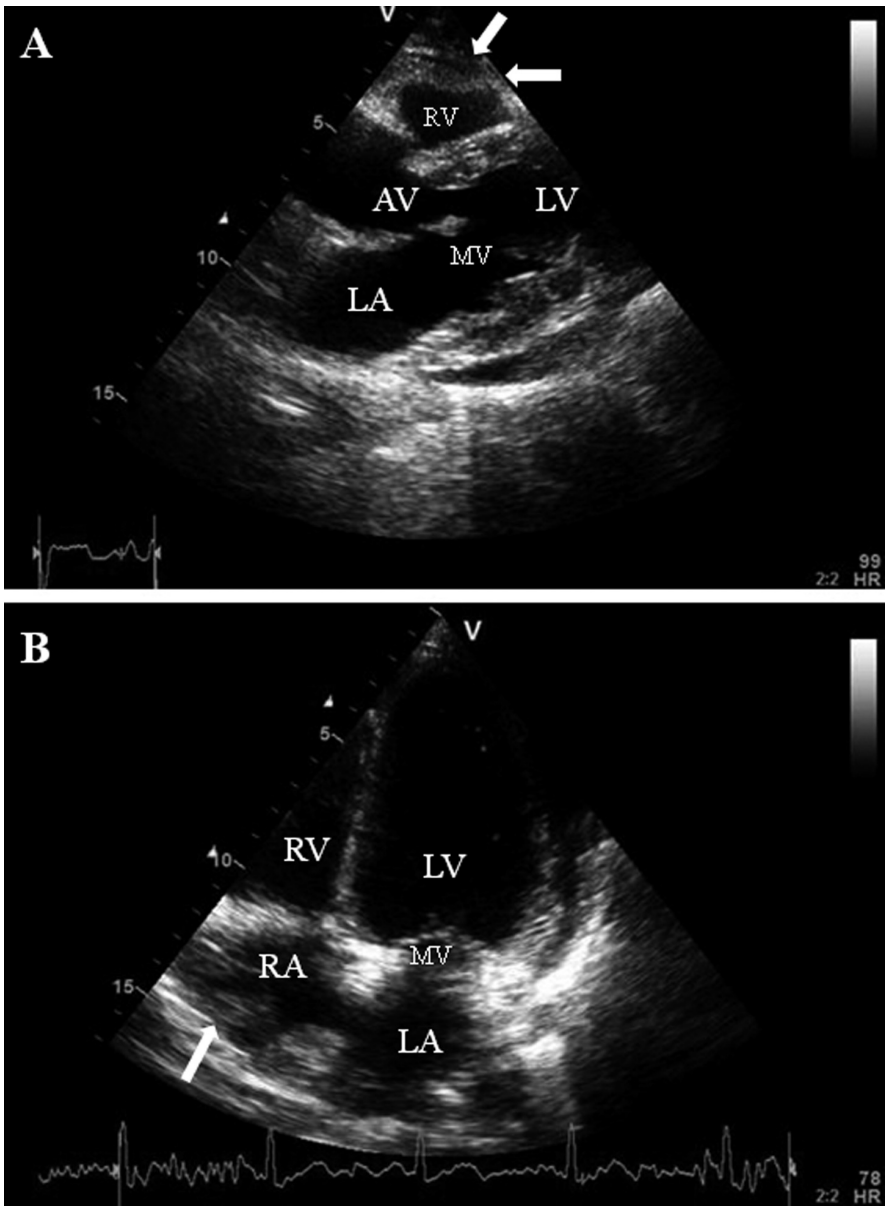

Fig. 1. Echocardiography (parasternal view and 4-chamber view) showing mild to moderate pericardial effusion (arrowhead). Note the diastolic collapse of the free wall of the right ventricle (parasternal view, arrow) and the paradoxical movement of the right atrial free wall (4-chamber view, arrow).
RV: right ventricle; LV: left ventricle; LA: left atrium; RA: right atrium; MV: mitral valve; AV: aortic valve.

Systemic sclerosis (SSc) is a chronic autoimmune disease characterised by widespread microvascular damage and fibrosis of the skin and various internal organs, including the heart (1, 2). Necropsy studies report pericardial involvement in 33% to 72% of cases, although symptoms from pericarditis occur in 7% to 20% of patients only (3). Pericardial tamponade is exceptional and the treatment of choice usually is pericardiocentesis (4-8). The majority of the reported cases of pericardial tamponade had a poor outcome due to severe pulmonary hypertension, renal or heart failure, with infrequent exceptions (4). The described case is the second case report of scleroderma complicated with pericardial tamponade during pregnancy (9). The decompensation of a chronic silent pericardial effusion related to SSc during the pregnancy may be explained by several factors: the impairment of scleroderma by pregnancy, the increase of pericardial effusion due to haemodynamic changes during pregnancy, the natural evolution of the disease, or interruption of the treatment with angiotensin converting enzyme (ACE) inhibitors, and other factors including bacterial or viral infection may contribute to the increase of pericardial effusion (10). In our patient the renal function and the arterial blood pressure were normal.

The reported effects of pregnancy in a patient with SSc have varied (10-11). Retrospective studies clearly show an increased frequency of pre-term births and small full-term infants, but the frequency of miscarriage and neonatal survival rate did not differ from healthy controls (11). The worst life-threatening complication of a pregnancy is scleroderma renal crisis. Despite the fact that ACE inhibitors are associated with congenital abnormalities and are relatively contraindicated in pregnancy, in this case their use is recommended (10). The therapeutic attitude in pregnancy with tamponade remains a challenge. The pericardiocentesis or the surgical drainage and the discontinuation of pregnancy in cases with haemodynamic intolerance are necessary. In our patient the normal renal function and the stable haemodynamic condition allowed

a more conservative therapeutic approach. Pregnancy in SSc patients remains a challenge for the physician in order to protect the health of both the mother and the foetus (12). In order to avoid complications, pregnancies in SSc should be planned when the disease is stable, and should be avoided in rapidly progressing diffuse SSc as such patients are at a greater risk for developing serious cardiopulmonary and renal problems early in the disease.

Monitoring for signs of prematurity and intrauterine women with SSc can successfully complete pregnancy. Tamponade in SSc is rare, and difficult to treat. The therapeutic attitude depends on the location of effusion, haemodynamic tolerance and the foetal maturity.

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Competing interests: none declared.

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