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Localized pericardial inflammation in systemic lupus erythematosus

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ABSTRACT

Regional or localized pericarditis has been infrequently reported. We report a patient with systemic lupus erythematosus (SLE), who presented with retrosternal pleuritic-type chest pain without audible friction rub, electrocardiographic changes or detectable pericardial effusion on echocardiography. Computed tomography, however, revealed a circumscribed area of pericardial inflammation, suggesting a diagnosis of localized lupus-associated pericarditis. This case demonstrates that localized pericarditis may occur in SLE and that chest CT may be required as part of the work-up in the diagnosis of lupus pericarditis.

Introduction

Pericarditis is quite common in systemic lupus erythematosus (SLE), affecting about 30% of patients. The diagnosis of pericarditis relies on the presence of typical pleuritic chest pain (pain aggravated by inspiration), audible friction rub and serial electrocardiographic changes (1,2). These three major diagnostic criteria are often accompanied by additional supporting findings, such as markers of systemic inflammation (fever, acute phase reactants), and echocardiographically visible pericardial effusion (3, 4). It has not been ascertained how many of these criteria are necessary for the clinical diagnosis of acute pericarditis.

In contrast to pericarditis, which affects most of the circumference of the pericardial membrane, localized pericarditis – which is restricted to one section of the pericardial sac – has been only anecdotally reported. In these instances, localized pericarditis was diagnosed by electrocardiographic changes limited to several leads, by various imaging modalities, or at surgery (5-8). Localized pericarditis has never been described in systemic lupus erythematosus (SLE).

We recently cared for a young patient with SLE and pre-cordial pleuritic type chest pain, who despite the absence of the commonly accepted criteria for pericarditis, was diagnosed with localized pericarditis based on computed tomography findings.

Case report

A twenty-four-year old female patient was recovering from a flare-up of systemic lupus erythematosus manifested by arthritis and bilateral pleural effusion without any chest pain. While being treated with prednisone in a tapering regimen, she presented with severe left retrosternal chest pain for the first time in the course of her disease. The pain was sharp and was aggravated by swallowing, deep breathing and lying supine. No other chest-related symptoms were present. On examination, the pulse was 115 per minute, and the temperature 37.8ºC. Auscultation revealed normal heart sounds without audible friction rub. Mild synovitis was evident in the proximal interphalangial joints of the hands. The rest of the physical examination was unremarkable. Chest X-ray revealed residual small right pleural effusion. Serial electrocardiograms showed only sinus tachycardia. A complete blood count, chemistry results and urine analysis were normal. The erythrocyte sedimentation rate was 45 mm in the first hour. The serum anti-DNA level was 72% (normally negative), and the complement components were low, with C4 11 mg/dcl (normal value 16-38 mg/dcl) and C3 59 mg/dcl (NV 79-152). Ventilation-perfusion scan of the lungs was normal. Echocardiography did not show pericardial effusion. Gastroscopy revealed a normal upper GI tract.

The patient's disabling pain persisted for several days, despite her being on prednisone at a dose of 40 mg a day, naproxen 500 mg twice daily and omeprazole 20 mg per day. Computed tomography of the chest revealed focal thickening of the pericardium, with localized effusion between the two layers at the area of the cardiac apex (Fig. 1A). In other areas the pericardium had a normal appearance like a pencil line. On a chest computed tomography done one month earlier, the entire pericardium appeared normal (Fig. 1B). A diagnosis of acute localized pericarditis secondary to lupus erythematosus was made. The prednisone dose was increased, and azathioprine and colchicine were also added. The patient's chest pain subsided within the next days. At the

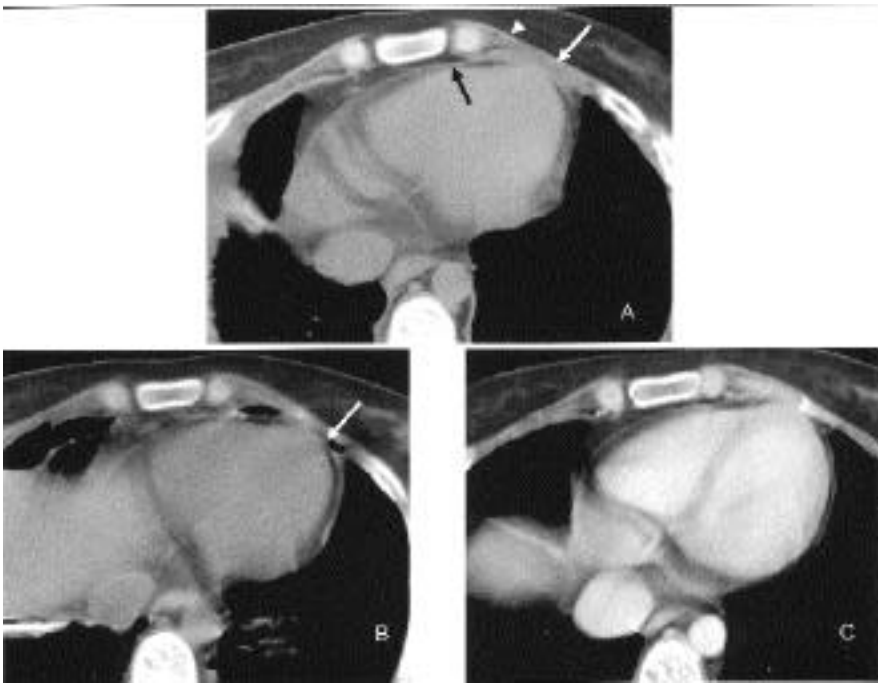

Fig. 1. Localized pericarditis on computed tomography (CT). (**A**) CTat the level of the cardiac apex shows localized effusion between two mildly thickened pericardial layers (**white arrows**), adjacent to an area of normal pericardium (**black arrow**). This area is separated from the chest wall (**white arrowhead**). (**B**) CT at the level of the cardiac apex, one month prior to admission, demonstrating normal pericardium (**white arrow**). (**C**) Contrast enhanced CT scan at the level of the cardiac apex, demonstrating normal pericardium, a year after CT(**A**) was performed.

six-month follow-up the patient was well. A computed tomography performed one year later showed a normal pericardium (Fig. 1C).

Discussion

The patient presented had pleuritic-type substernal pain with features suggestive of pericarditis. Evaluation, making use of classical tools such as EKG and echocardiography, failed to confirm a diagnosis of pericarditis. However, computed tomography demonstrated circumscribed pericardial inflammation (Fig. 1) most likely suggestive of localized pericarditis.

The limited extent of the disease in our patient may have been due to the partial immunosupression provided by a tapering prednisone regimen in a patient who still had active SLE. The 'classical' findings of pericarditis were absent in this patient probably because of the limited extent of the pericardial inflammation.

To the best of our knowledge, this is the first published case of localized pericarditis in SLE. However, rather than being a unique case, we suspect that our patient represents a large group of patients whose work-ups fail to detect 'traditional' evidence of pericarditis, and who therefore receive other diagnoses such as non-specific or atypical chest pain. Most of these patients probably improve because they are treated with steroids or non-steroidal anti-inflammatory drugs to control the other manifestations of SLE.

Yet in certain clinical circumstances a definitive diagnosis of pericarditis may be required, as for example when pericarditis is the sole manifestation or when symptoms are relentless. Under such circumstances, if traditional tests are non-diagnostic, the possibility of localized pericardial disease can be explored using computed tomography, as illustrated by our case and others (9, 10).

In conclusion, in SLE (and probably in other diseases associated with pericarditis), acute pericardial inflammation may present as localized pericarditis, which may fail to produce the accepted diagnostic criteria of pericarditis. Under these circumstances a diagnosis of pericarditis may be reached using computed tomography or other imaging studies of the pericardium.

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