Pericardial effusion and mitral valve involvement in systemic lupus erythematosus

Echocardiographic study

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SUMMARY Echocardiography was used in 30 women and 2 men with systemic lupus erythematosus (SLE) in order to determine the incidence and severity of pericardial effusion and mitral valve involvement. 31 patients showed normal thickness of the mitral valve leaflets, only one patient showed irregular thickening of the leaflets suggesting the presence of vegetations. Mitral valve motions were normal in all patients. These results indicate that myocardial and valvular involvement in SLE is usually not severe enough to result in haemodynamic abnormalities. Pericardial effusion was found in 2 patients who were symptom free, whereas 4 of the patients with a past history suggestive of pericarditis showed no echocardiographic evidence of pericardial effusion. These suggest the transient nature of pericarditis in SLE, and the value of echocardiography as a diagnostic tool in detecting clinically inapparent lupus pericarditis.

Since Libman and Sachs (1924) first described cardiac involvement in systemic lupus erythematosus (SLE), many clinical and pathological studies have shown that different heart tissues may be affected (Taubenhaus *et al.*, 1955; Shearn, 1959; Brigden *et al.*, 1960; Kong *et al.*, 1962; Hejtmancik *et al.*, 1964; James *et al.*, 1965; Estes and Christian, 1971; Cosh, 1972). The verrucous endocarditis which mainly affects the mitral valve rarely results in significant valvular obstruction or regurgitation, and is therefore found more often at autopsy than in life (Harvey *et al.*, 1954; Shearn, 1959; Brigden *et al.*, 1960). The natural history of this kind of lesion is unknown, and no correlation has been found between histological findings of endocarditis and heart murmurs (Jessar *et al.*, 1953; Shearn, 1959; Brigden *et al.*, 1960; Kong *et al.*, 1962). In addition, some evidence of pericarditis is practically always found at autopsy in SLE patients, although it too may pass unrecognized during life (Brigden *et al.*, 1960). Echocardiography has been shown to be a sensitive technique for

detecting pericardial effusion (Feigenbaum, 1970; Horowitz *et al.*, 1974) and assessing abnormalities in movement and form of the mitral valve cusps (Edler 1961; Zaky *et al.*, 1968; Dillon *et al.*, 1973).

In this study echocardiography was used to determine the incidence and severity of pericardial effusion and mitral valve involvement in 32 living cases of SLE.

Patients and methods

Thirty-two patients with SLE (15 to 68 years of age, mean 38 years) were studied. The diagnosis of SLE was established according to criteria defined by the American Rheumatism Association (Cohen and Canoso, 1972). The mean duration of illness in the 30 women and 2 men was 9 years, range 1–27 years. All patients had been treated at some time during the course of their illness with corticosteroids; 22 were on maintenance doses of steroids at the time of study.

A complete history and physical examination were carried out on each patient, with special attention to the cardiovascular system. A standard 12-lead electrocardiogram and chest x-ray were taken on each patient. The echocardiograms were recorded

with an ultrasound device (Unirad Corp. Series 100) using a 7.5 cm focus transducer which measured 12 mm in diameter. Echocardiograms were recorded with rigid adherence to the technique and criteria previously established (Chang, 1976; Feigenbaum, 1972; Horowitz *et al.*, 1974). The echoes were displayed on an oscilloscope in the time motion mode, and were recorded on polaroid film along with a superimposed electrocardiogram. Measurements of the diastolic closure (E-F slope) were made on multiple complexes and the highest value was recorded.

Results

During the study 30 patients of the group were completely free of any cardiac symptoms, 2 patients experienced exertional dyspnoea, one of whom also suffered from anginal pains. 3 patients had a past history of acute transient episodes of congestive heart failure, probably due to myocarditis; 9 had a history of hypertension; 2 complained of occasional palpitations; 3 had a history of chest pain suggestive of pericardial or pleural involvement; and 2 had transient episodes of pericardial friction rub.

Fig. 1 Normal echocardiogram in a 38-year-old woman with SL of 16 years' duration. CW=chest wall; RV=right ventricle; IVS=intraventricular septum; LV=left ventricle; AMV=anterior mitral valve leaflet; PMV=posterior mitral valve leaflet.

The cardiovascular examination was entirely normal in 8 patients, hypertension was found in 6 patients (BP > 160/90), a fourth heart sound was heard in 1 patient, a soft systolic murmur was heard at the apex in 8 patients, and 3 patients had a grade 3/6 apical holosystolic murmur. Another 6 patients had a systolic ejection type murmur at the aortic area and 1 patient had a diastolic murmur grade 2/6 along the left sternal border. Bilateral basilar moist rales were found in one patient. The electrocardiogram was abnormal in 14 patients. The abnormal findings were sinus tachycardia in 2 patients, major left axis deviation ($> -30^\circ$) in 2 patients, low voltage (< 5 mm in all limb leads) in 1 patient, left ventricular hypertrophy in 2 patients, incomplete right bundle-branch block in 1 patient, ventricular premature contractions in 3 patients, and atrial premature contractions in 1 patient. The postero-anterior and lateral chest x-rays were normal in 29 patients, 3 patients showed cardiomegaly, 1 of them also showed dilatation of aorta and emphysema of lungs.

According to the echocardiographic data, 31 patients had a normal thickness of the mitral valve

leaflets (Fig. 1), and 1 patient showed an irregular thickening of the leaflets (Fig. 2). All patients had a normal E-F slope: the range was 82–192 mm/s, average 127 mm/s. The amplitude of the mitral valve opening (C-E) ranged between 20 and 32 mm. No echo-free space was seen between the anterior right ventricular wall and the stationary chest wall in any of the patients. These findings excluded noteworthy anterior pericardial effusion. Echocardiographic evidence of posterior pericardial effusion was seen in 2 patients (Fig. 3). Pericardiocentesis was not performed to determine the exact volume of the pericardial fluid.

Discussion

MITRAL VALVE INVOLVEMENT

It is commonly assumed that the pathological changes occurring in Libman-Sachs endocarditis do not cause distortion of the valvular structure and therefore do not significantly change the haemodynamic properties of the valves (Shearn, 1959; Brigden *et al.*, 1960; Hejtmancik *et al.*, 1960; Kong

Fig. 2 Echocardiogram of the anterior mitral valve (A) showing non-uniform thickening suggesting the presence of Libman-Sachs vegetations (veg). P = posterior mitral valve leaflet.

Fig. 3 Echocardiogram shows a posterior pericardial effusion (PE). Note that as the gain is diminished an echo-free space is visualized between the posterior pericardium (PER) and the epicardium (EP). EN = endocardium.

et al., 1962). Our findings of normal thickness and movement of the mitral valve leaflets in 31 out of 32 SLE patients support this assumption.

Our results disagree, however, with those of a recent study published by Maniscalco *et al.* (1975), in which they found a decreased E-F slope in 9 out of 25 patients with SLE and thickening of the mitral valve in 2 patients. If stenosis of the mitral valve, due to endocardial involvement, was the cause of a decreased E-F slope, one would expect to find additional changes similar to those observed in rheumatic involvement of the mitral valve. Such changes include thickening of the mitral cusps, decreased excursion of the leaflets and abnormal posterior leaflet movement in diastole (Segal *et al.*, 1974). Maniscalco and co-workers did not state whether the 2 patients with thickening of the mitral valve belong to the group of 9 patients with decreased E-F slope, and did not deal with the other changes mentioned above. Review of the literature on involvement of the mitral valve in autopsy specimens in SLE patients showed in the majority of the cases a non-severe verrucous endocarditis which did not interfere significantly with cardiac function (Brigden *et al.*, 1960; Kong *et al.*, 1962; Hejtmancik *et al.*, 1964). Stenosis of the mitral valve was an uncommon lesion in SLE patients (Shearn, 1959; Brigden *et al.*, 1960), and in most cases a history of rheumatic fever was found. The only patient in our group who showed thickening of the mitral valve (suggesting the presence of vegetations; Fig. 2), had combined mitral and aortic valve lesions, and gave a history suggestive of rheumatic fever. It is possible, therefore, that when mitral stenosis is found in a patient with SLE, rheumatic valvular disease may also be present.

Decreased left ventricular compliance due to myocardial involvement of the systemic disease can also be a possible cause for diminished E-F slope (DeMaria *et al.*, 1976). Maniscalco *et al.* (1975) did not mention haemodynamic or clinical findings which could reflect reduced compliance. The significance of the histological abnormality, which has been found in SLE, has to our knowledge not been studied haemodynamically. The clinical impression however is that myocarditis, even when extensive, does not commonly lead to cardiac insufficiency (Griffith and Vural, 1951; Shearn, 1959). The fact that all our patients, including 3 who had a history of acute transient myocarditis, displayed normal E-F slopes, argues against significantly reduced myocardial compliance during the chronic phase of SLE.

PERICARDIAL INVOLVEMENT

Pericardial involvement may be the earliest cardiac occurrence in SLE. Notwithstanding the infrequency of this finding during life, it is often found at autopsy

(Griffith and Vural, 1951; Shearn, 1959; Brigden *et al.*, 1960; Hejtmancik *et al.*, 1964). Pericardial effusion was found in 2 of our 32 patients, a very small percentage in contrast to 44% found by Maniscalco *et al.* (1975).

Pericardial involvement, like any other organ involvement in this disease, will be aggravated by an acute phase. All our cases were outpatients and none showed any signs of an acute phase of the disease. Brigden *et al.* (1960) found evidence of recent or old pericarditis at autopsy in 20 out of 27 cases, in those showing recent effusion the disease was up to 3 years' duration, whereas in those showing chronic adhesions the disease was from 2 to 11 years' duration, and in some of these the pericardial space was totally obliterated. The duration of the disease in our series was more than 3 years in 25 out of 32 patients, which may account for the low incidence of pericardial effusion. This finding may also reflect the increased awareness of this disease, the availability of newer diagnostic tests, and the beneficial effect of corticosteroid therapy in controlling the extensiveness of the pericarditis (Hughes, 1973; Dubois, 1962; Bulkley and Roberts, 1975).

Five of our patients with past history suggestive of pericarditis did not show echocardiographic evidence of pericardial effusion. These results suggest the transient nature of pericarditis in SLE. Echo examinations showed pericardial effusion in 2 other patients who were symptom free, and indicates the value of this technique.

Our results, which are in agreement with the pathological findings described in the literature, show that myocardial and valvular involvement in SLE is usually not severe enough to result in haemodynamic abnormalities. They also show that echocardiography can be an important diagnostic tool when used as a routine procedure in SLE. It may help detect clinically inapparent pericardial involvement. Pericardial fluid accumulation can thus be diagnosed before it leads to cardiac tamponade.

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