Pericardial involvement in systemic sclerosis

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Abstract

Objective—To determine the frequency and histological characteristics of pericardial involvement in systemic sclerosis.

Method—Necropsy sections of pericardium from 44 patients with systemic sclerosis were studied, together with sections from 19 age/sex matched controls. Sections were stained with haematoxylin and eosin, acid toluidine blue, and elastic van Gieson. Mast cells were counted in 10 random high power fields and the degree of fibrosis was quantified using a Chalkley count.

Results—Chronic pericarditis was seen in 31 (77.5%) of the systemic sclerosis cases, but in only one of the controls. The characteristic changes of uraemic pericarditis were not seen. The degree of fibrosis was greater in those with systemic sclerosis, though numbers of mast cells, thought to be important in fibrogenesis, were similar in both groups. Myocardial fibrosis was seen in 15 (37.5%) of systemic sclerosis cases but in none of the controls. Conclusion—The incidence of pericarditis and myocardial fibrosis is much greater than in controls. The results indicate that pericarditis is a primary disease (rather than secondary to uraemia).

Systemic sclerosis is a multi-system disease characterised by dense fibrosis of visceral organs and skin. It commonly involves the heart, causing myocardial fibrosis and cardiac failure. Necropsy studies report pericardial involvement in between 33% and 72% of cases but symptoms from pericarditis occur in only 7% to 20% of patients. Previous necropsy studies have relied on macroscopic inspection and have not examined the pericardial histologically. We investigated the pericardial changes at the histological level, and, as mast cells have been implicated in the development of fibrosis in systemic sclerosis, also studied the level of mast cell infiltration in the pericardium in systemic sclerosis.

Methods

Forty four postmortem specimens of cardiac tissue (26 female, 18 male), from patients dying in hospital between 1971 and 1991 were obtained from pathology departments throughout the United Kingdom. All patients had a diagnosis of scleroderma recorded on the death certificate and postmortem reports were examined where available. A control group of 19 age/sex matched patients dying without evidence of connective tissue, heart or renal disease was retrieved from the files of Manchester Royal Infirmary. Blocks of cardiac tissue containing overlying pericardium were selected from each case in both groups and sections stained with haematoxylin and eosin, acid toluidine blue, for mast cells, and elastic van Gieson, for collagen and elastic fibres. The histological features were reviewed in each case and the presence of pericardial inflammation and fibrosis and of any myocardial changes noted.

Mast cells, disclosed by the acid toluidine blue stain, were counted in 10 high power fields (hpf) at the pericardial surface in each group. The degree of fibrosis in each group was quantified using a 25 point Chalkley point eyepiece graticule. Points lying over collagen fibres, indicated by the elastic van Gieson stain, were counted in 10 hpf, so that the count in each case was out of a maximum number of 250 points.

Results

The patients ranged in age from 9 to 89 years (mean 54 in males and 41 in females), while the controls (9 males, 10 females) ranged in age from 26 to 84 years (mean 52.5 in males and 49.8 in females). Myocardial fibrosis was seen in 15 (37.5%) of the systemic sclerosis cases but in none of the control group while pericarditis was present in 31 (77.7%) of those with systemic sclerosis, but in only one of the controls. The pericardial infiltrate was composed predominantly of chronic inflammatory cells in most cases (fig 1), though two showed an additional neutrophilic infiltrate. Dense fibrinous deposits, characteristic of uraemic pericarditis, were seen in two cases, one of which had confirmed evidence of renal failure.

The amount of pericardial fibrosis in those with systemic sclerosis was significantly greater than in the controls with a mean Chalkley...
count of 21.1 of 250 compared with 11.8 of 250 (p<0.05). The incidence of mast cells was also greater in those with systemic sclerosis, with a mean of 4.1 mast cells/10 hpf in the index group and 2.9/10 hpf in the controls, but the difference was not significant (p>0.05).

Acute myocardial infarction was seen in three cases. Detailed postmortem reports were available in 16 cases, of which eight had confirmed renal failure, with uremia, at death.

Discussion

Systemic sclerosis causes visceral fibrosis and frequently results in cardiac failure caused by myocardial fibrosis. Pericardial involvement is frequently seen in end stage systemic sclerosis. That antemortem echocardiography has also shown a high incidence of pericardial involvement suggests that this finding is not restricted to those dying with the disorder. The pericardial changes are those of fibrosis and chronic inflammation, while mast cell numbers were not significantly greater compared with normal controls. The pericardial changes are not typical of uremia and we believe they reflect primary involvement in the disease process rather than those secondary to uremia. The lack of increase in mast cells may reflect the late stage of the disease, or degranulation caused by severe illness in these cases.

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