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Endomyocardial fibrosis and Löffler's endocarditis parietalis fibroplastica

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Summary

The morphological features of endomyocardial fibrosis and Löffler's endocarditis are described. Sixteen cases of the chronic stage of Löffler's endocarditis were compared with pathological material of thirty-two patients with endomyocardial fibrosis. No morphological differences were noted. It is suggested that endomyocardial fibrosis and Löffler's endocarditis are part of the same disease spectrum, the origin of which can be traced back to the presence of eosinophils in the myocardium.

Introduction

Both these entities represent examples of the obliterative type of cardiomyopathy, using the clinical classification of Goodwin (1974) and Oakley (1974, 1975).

Endomyocardial fibrosis (EMF)

The earliest clinical report of the disease appeared in the literature in 1946 by Bedford and Konstam, and the description of the pathology by Davies (1948). Although it does not represent a major problem in Venezuela, cases have been reported (Hernández-Pieretti, 1977), in other parts of North and South America, for example Fagundes (1963), Bishop *et al.* (1968) and Guimarães *et al.* (1971). For a long time it was considered to be a disease of tropical and subtropical regions, often affecting Europeans resident in Africa (Brockington, Olsen and Goodwin, 1967). Recent evidence suggests that it is by no means confined to these regions.

Clinically the disease may be preceded by a febrile illness. Patients tend to be wasted and are often cyanosed and may present with features similar to those of constrictive pericarditis, or with mitral insufficiency accompanied by pulmonary hypertension, which may be severe (Davies, Deuchar and Missen, 1965). Embolic phenomena occur and characteristically angiography reveals a ventricular chamber which is reduced in size.

Pathology

Macroscopically, the distribution of the thickened endocardium, which may be up to 5 mm thick (normal left ventricular inflow tract 10 μm) (Okada, 1961) may vary (Shaper, Hutt and Coles, 1968), but when the left ventricle is affected, the posterior mitral valve leaflet, the inflow tract and papillary muscle are involved. The thick endocardium often ends abruptly in a thick rolled edge (Fig. 1) (Davies, 1968). Thrombus is superimposed in more than 50% of patients. Fibrous septa, extending into the underlying myocardium for a short distance, are also characteristic appearances. Most commonly both ventricles are involved, but when one ventricle is affected only, the left is involved three times more commonly than the right.

FIG. 1. The left ventricular cavity has been opened showing the severely thickened endocardium, involving the inflow tract, papillary muscle and apex. Note the thick rolled edge as the outflow tract beneath the anterior mitral valve leaflet is approached.

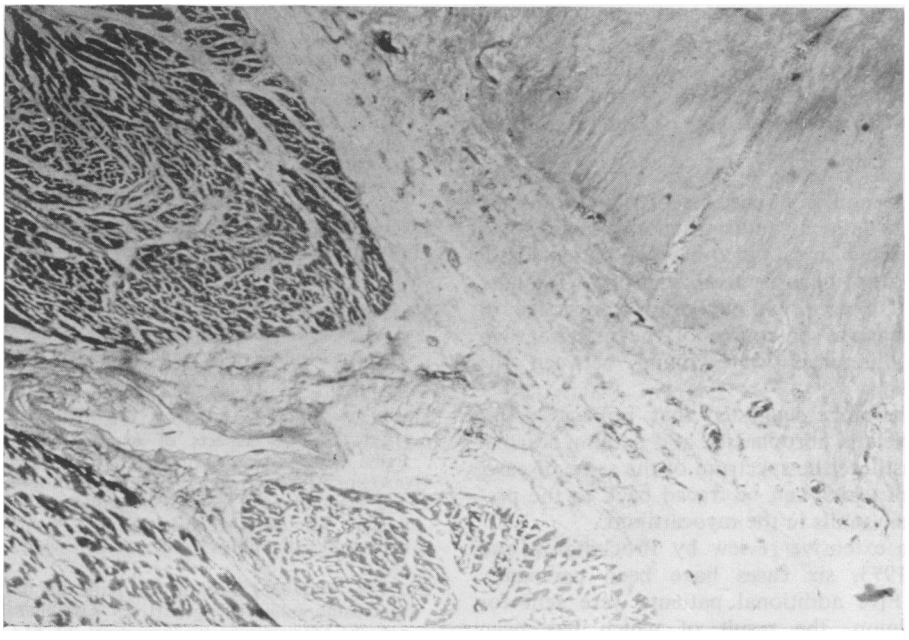

FIG. 2. Photomicrograph of a case of endomyocardial fibrosis showing part of the thickened endocardium. Note the collagen tissue to the right of the photograph and the granulation tissue layer extending also into the underlying endocardium. Haematoxylin and eosin ×25.

Histologically, the thick endocardium shows zonal layering, superficial hyaline collagen in which foci of calcification may be present. The middle zone consists of fibrous tissue and the deepest layer shows dilated blood vessels, varying degrees of chronic inflammatory cells and a variable number of eosinophils. From this layer the septa extend into the underlying myocardium (Fig. 2).

Löffler's endocarditis parietalis fibroplastica (Löffler 1936)

This constitutes the syndrome of eosinophilia and chronic heart failure. Clinically it resembles EMF. In its chronic form endocardial fibrosis is prominent. Because of the striking similarities between EMF and the chronic form of Löffler's endocarditis, Brockington and Olsen (1973) reviewed the literature of approximately 90 endocarditic cases which had been documented. The cause of eosinophilia varied and may be categorized in three groups: firstly idiopathic (approximately 50%), secondly reactive (25%), and patients in this group suffer from conditions such as polyarteritis nodosa, asthma, sensitivity to antiarrhythmic drugs, Hodgkin's disease or carcinoma, and thirdly, eosinophilic 'leukaemia' (25%).

From thirty of these patients pathological material was obtained and three major histological patterns may be recognized.

(1) *Necrotic stage*

(Ten patients, average duration of illness 5·5 weeks.)

An eosinophilic myocarditis particularly prominent in the inner layers of the myocardium, was the characteristic feature of this stage. Arteritis of small intramural vessels was commonly found. Areas of myocardial necrosis were present.

(2) *Thrombotic stage*

(Eight patients, average duration of illness 10 months.)

In this stage thrombus was prominent; in four of the patients it was mild; in the remaining four, prominent fibrous endocardial thickening was present. The thickened endocardium was, in some areas, continuous with the necrotic foci—noted in Stage 1—which had now become fibrous. Arteritis still persisted in this stage.

(3) *Fibrotic stage*

(Twelve patients, average duration of illness 24·5 months.)

Thick fibrous endocardium, with some thrombus superimposed in ten cases, characterized this stage. The thickened endocardium was arranged in layers and was identical to the changes described in EMF.

Arteritis was no longer present and eosinophils were either scanty or absent.

Macroscopically, three hearts were identical in the appearances of the distribution of the thickened endocardium to that described in endomyocardial fibrosis.

Histological material of twelve patients of the fibrotic stage and four patients of the thrombotic stage with prominent endocardial thickening, was then compared with that of thirty-two patients with endomyocardial fibrosis from Europe, Uganda, Nigeria and Brazil, without prior knowledge of previous diagnosis or source of the material. No significant differences were found between the various groups.

It was therefore suggested that Löffler's endocarditis parietalis fibroplastica and endomyocardial fibrosis constitute the spectrum of the same disease, the origin of which can be traced back to the presence of eosinophils in the myocardium.

Since the extensive review by Brockington and Olsen in 1973, six cases have been personally examined. Five additional patients were sent for second opinion, the result of which has been published (Scott and Bruce, 1975). One additional case was diagnosed in life from endomyocardial biopsy material obtained by bioptome. The subject has recently been reviewed by Oakley and Olsen (1977).

References

Bedford, D.E. & Konstam, G.L.S. (1946) Heart failure of unknown aetiology in Africans. *British Heart Journal*, **8**, 236.

Bishop, M.B., Bousvaros, G., Cunningham, T.J., Jain, A.C. & Davies, J.N.P. (1968) Endomyocardial fibrosis in a North American Negro. Probable familial incidence. *Lancet*, **ii**, 750.

Brockington, I.F. & Olsen, E.G.J. (1973) Löffler's endocarditis and Davies' endomyocardial fibrosis. *American Heart Journal*, **85**, 308.

Brockington, I.F., Olsen, E.G.J. & Goodwin, J.F. (1967) Endomyocardial fibrosis in Europeans resident in tropical Africa. *Lancet*, **i**, 583.

Davies, H., Deuchar, D.D. & Missen, G.A.K. (1965) Endomyocardial fibrosis. Two cases with severe pulmonary hypertension presented in England. *Guy's Hospital Reports*, **114**, 1965.

Davies, J.N.P. (1948) Endocardial fibrosis in Africans. *East African Medical Journal*, **25**, 10.

Davies, J.N.P. (1968) The ridge in endomyocardial fibrosis. *Lancet*, **i**, 631.

Fagundes, L.A. (1963) Endomyocardial fibrosis. Report of three cases in Southern Brazil. *Review of the Institute of Tropical Medicine, São Paulo*. **5**, 198.

Goodwin, J.F. (1974) Prospects and predictions for the cardiomyopathies. *Circulation*, **50**, 210.

Guimaraes, A.C., Esteves, J.P., Filho, A.S. & Macedo, V. (1971) Clinical aspects of endomyocardial fibrosis in Bahia, Brazil. *American Heart Journal*, **81**, 7.

Hernández-Pieretti, O. (1977) Echocardiographic diagnosis and evaluation of cardiomyopathies: idiopathic hypertrophic subaortic stenosis, Chagas' heart disease and endomyocardial fibrosis. *Postgraduate Medical Journal*, **53**, 533.

Löffler, W. (1936) Endocarditis parietalis fibroplastica mit Bluteosinophilie ein eigenartiges Krankheitsbild. *Schweizerische Medizinische Wochenschrift*, **17**, 817.

Oakley, C.M. (1974) Clinical recognition of cardiomyopathies. *Circulation Research*, **35** (Suppl. 2), 152.

Oakley, C.M. (1975) The relation between function and causation in cardiomyopathy. *Postgraduate Medical Journal*, **51**, 271.

Oakley, C.M. & Olsen, E.G.J. (1977) Eosinophilia and heart disease (Editorial). *British Heart Journal*, **39**, 233.

Okada, R. (1961) Clinicopathological study on the thickening of parietal endocardium in the adult heart. *Japanese Heart Journal*, **2**, 220.

Scott, M.E. & Bruce, J.H. (1975) Löffler's endocarditis. *British Heart Journal*, **37**, 534.

Shaper, A.G., Hutt, M.S.R. & Coles, R.M. (1968) Necropsy study of endomyocardial fibrosis and rheumatic heart disease in Uganda, 1950–65. *British Heart Journal*, **30**, 391.