Endomyocardial fibrosis in Uganda (Davies' disease). Part II: An epidemiologic, clinical, and pathologic study

Daniel H. Connor, Krishna Somers, Michael S.R. Hutt, William C. Manion, Paul G.D'Arbela

Endomyocardial fibrosis in Uganda (Davies' disease) is a common type of fatal heart disease in the autopsy population at Mulago Hospital. During this study, endomyocardial fibrosis caused 25 per cent (1665) of deaths from intrinsic heart disease; it showed a striking and unexplained predilection for the immigrant Rwandans, but spared the large local indigenous tribe, the Ganda. Clinically the patients had a sudden or insidious onset of failure of one or both ventricles, which proved fatal over a period of days, weeks, months, or years. At necropsy there were mural endocardial lesions at one or more of three sites—the apex of the right ventricle, the posterior wall of the left ventricle, and the apex of the left ventricle. In the early stage the cardiac connective tissues, especially those of the endocardium, were swollen with acid mucopolysaccharide (AMP) and covered by a layer of fibrin. In later lesions, the involved areas had resolved as hard, white scard composed of collagen and elastica. There were peculiar, and we believe characteristic, foci of collagen necrosis in the scar tissue at the endomyocardial junction. The cause of endomyocardial fibrosis remains unknown, but the consistent mucinous swelling of the cardiac “ground substance” and vessels plus the focal nonsuppurative disintegration of collagen suggest to us that hypersensitivity is the underlying mechanism. Although rheumatic heart disease and endomyocardial fibrosis have a number of common features, including diffuse and focal disruption of the cardiac connective tissues, the consistent differences in RHD and EMF have convinced us that the two are different diseases. At the present time there is no microscopic evidence to support the view that endomyocardial fibrosis in Uganda (Davies' disease) has a similar histogenesis to cardiopathies in other parts of the world.