

Epidemiology aspects of endomyocardial fibrosis

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Summary

Endomyocardial fibrosis (EMF) is a specific clinicopathological entity. It occurs sporadically throughout the world but is endemic in parts of Africa, India and South America. In sporadic cases, blood eosinophilia is a common feature; this is a variable finding in endemic cases. The curious geographical distribution of EMF in the tropics remains unexplained.

KEY WORDS: epidemiology, endomyocardial fibrosis, eosinophilia.

Endomyocardial fibrosis (EMF) is a specific clinicopathological entity which was first described by Löffler (1936) in a patient with an associated eosinophilia. Sporadic cases with similar clinical and pathological features have since been reported from many parts of the world. In most of these cases, blood eosinophilia has been a feature, though the causes of the rise in eosinophils have been varied.

Endomyocardial fibrosis (EMF) in Uganda

Bedford and Konstam (1946) described a form of heart disease in 40 West African soldiers serving in the Middle East. Several of these patients died and post-mortem revealed subendocardial fibrosis with features that are now associated with EMF. Davies (1947) working in Uganda, encountered in quick succession 3 necropsies in patients dying of cardiac failure each of which was characterised by endocardial fibrosis of the ventricular inflow tracts. In subsequent studies, (Davies and Ball, 1955; Williams, Ball and Davies, 1954; Ball, Williams and Davies, 1954) the clinical and pathological features of EMF as seen in Uganda were defined and attention was drawn to its frequency as a cause of organic heart disease, particularly in the Rwanda-Burundi ethnic groups. Davies (1961) recorded 82 cases of EMF studied at necropsy. Later post-mortem studies in Uganda confirmed the high frequency of EMF (Connor *et al.*, 1967, 1968; Shaper, Hutt and Coles, 1968). During the period 1950-1965, 172 cases were diagnosed at necropsy, of which 63% occurred in Rwandan immigrants who, as a group, only account

for 24% of all necropsies (Shaper *et al.*, 1968). During the period 1965-68, when facilities, patient turnover and necropsy rates were high in Kampala, Uganda, 77 cases were diagnosed at post-mortem, EMF was the third most common form of organic heart disease accounting for 13% of all cardiac deaths and the predominance in Rwandans was still evident (Hutt, 1970).

Endomyocardial fibrosis elsewhere in Africa

Although cases of EMF have been reported in Kenya (Turner and Manson-Bahr, 1960) and from Tanzania (Makene, 1970), it is a comparatively uncommon condition in these countries, and does not occur in the highland areas. During 1980 and 1981 no cases were seen at post-mortem in Nairobi. Individual cases have been described in Zambia (Lowenthal and Fine, 1968), Zimbabwe (Gelfand, 1957) and Mozambique (Biylsma, 1979), but the condition does not appear to occur in black South Africans. EMF was first reported in Nigeria from Enugu (Nwokolo, 1955), but most of the cases have been described from Ibadan (Parry and Abrahams, 1965; Edington and Jackson, 1963; Brockington and Edington, 1972). Brockington and Edington (1972) in an analysis of 6,817 necropsies studied between 1958 and 1966, described 41 cases of EMF, the largest post-mortem series outside Uganda. EMF appears to be rare in Zaria in Northern Nigeria. EMF has been diagnosed clinically and confirmed at necropsy in a number of cases in the Ivory Coast (Bertrand *et al.*, 1975, 1976; Parmentier and Collin, 1972; Batzenschlager, Reville and Finicker, 1961), Ghana (Edington, 1954), Zaire (Coelho and Pimental, 1963), and the Sudan (O'Brien, 1954). However, it is difficult to compare prevalence within the countries of sub-Saharan Africa because information from large areas of the continent is still negligible.

Endomyocardial fibrosis in Asia and the New World

In the Asian subcontinent, EMF is endemic in South India and Sri Lanka (Nagaratnam and Dissanayake, 1959; Gopi, 1968). Some cases have also

been seen in Chandigarh, North India (Datta *et al.*, 1977). EMF has been described in Malaya (Brody, 1957), but has apparently not been seen in Papua, New Guinea, which has a very similar environment and disease pattern to sub-Saharan Africa.

In the New World, cases of EMF have been described in Brazil (Fagundes, 1963; Andrade and Guimaraes, 1964), Venezuela (Suarez and Suarez, 1967) and Colombia (Correa *et al.*, 1963). No cases have been reported from Jamaica where other forms of cardiomyopathy, such as congestive cardiomyopathy are common.

The epidemiology and aetiology of endomyocardial fibrosis

In any search for the aetiology of a disease attention must be paid to its epidemiological features. EMF occurs sporadically, usually associated with eosinophilia, throughout the world. Scattered throughout the tropical belt, but particularly in some parts of sub-Saharan Africa, Southern India and South America, EMF is endemic. However, this endemicity appears to have localized areas of high prevalence as is evident in East Africa. The very high prevalence in Rwandans living alongside other Africans in Uganda and the occurrence in families, suggests that genetic factors may play a role in the aetiology (Patel *et al.*, 1971) possibly by determining immunological susceptibility to some environmental influence.

It is apparent from available evidence that the prevalence of EMF shows wide variations throughout the world, with a high endemicity in many tropical regions. Unfortunately, to obtain reliable data on the prevalence of EMF, we need a combination of good facilities and the availability of cardiac diagnostic services and necropsy confirmation of the lesion. Such evidence is lacking in many tropical countries, including Rwanda and Burundi.

There is now evidence that the initial heart lesion in EMF may be associated with abnormalities of eosinophils (Brockington and Olsen, 1973; Andy, Bishara and Soyinka, 1981) though eosinophilia is common in many tropical regions where EMF does not appear to be prevalent. This suggests that other factors, possibly immunological in nature, are also necessary to determine the prevalence of the disease in a particular location. If we are to understand the aetiology of EMF as well as its pathogenesis, it is essential that more comprehensive epidemiological information on the prevalence and natural history of this condition throughout the world is obtained.

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