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Discussion

DR E. G. J. OLSEN: I am delighted to learn that Prof. Hutt and I agree that the two conditions are pathologically and clinically the same. I, like him, mentioned additional aetiological factors, inflammatory, parasitic, genetic or immunological. He mentioned endomyocardial scars in other conditions but I cannot think of one single condition that gives a scar which mimics, even vaguely, endomyocardial fibrosis.

PROF. HUTT: I agree, given that you have a reasonable view of the actual heart. The reason I suggested this difficulty is that when I first went to Uganda, I sent some blocks of EMF to Dr Manion who thought that the changes were non-specific until he saw pictures of the heart. There are problems in diagnosis based on small samples of tissue.

DR OLSEN: In my experience, the diagnosis can be made on endomyocardial biopsy, that is on tissue 2 mm in diameter.

DR P. J. RICHARDSON: As an analogy, the streptococcus has been identified as the aetiological agent in 50% of cases of rheumatic fever. I am not denigrating, in any way, the observation about the degranulated eosinophil, a pathological observation, but the whole process could be initiated by something else—a bacterial or viral agent.

PROF. HUTT: It is very difficult to fit it in say with a single parasitic disease as we are all trying to do. I have a feeling that somewhere in the equation right at the beginning of the process there is a missing factor, which perhaps leads to degranulated eosinophils, which would fit with the extraordinary geographical distribution. There are, of course, many diseases with very peculiar distributions which are unexplained. Why, for example, does Kaposi's sarcoma have such an extraordinary distribution in Africa?

DR C. J. F. SPRY: Many problems would be resolved if early cases of EMF could be diagnosed during life or at post-mortem. Why have they not been found in Uganda or elsewhere? Is it that these patients are clinically well until they reach the late fibrotic stage? How would you recommend that we set about finding early disease?

PROF. HUTT: We have failed to find any by chance at necroscopy. Clinically in geographical areas with a high incidence of infectious diseases resulting in eosinophilia, it is difficult to pick out of thousands of people who are ill, the early stage of a comparatively uncommon disease such as EMF. In terms of those interested in heart disease it is very important, but in terms of the population it is not very important.

DR SPRY: Are there any family studies on endomyocardial disease?

PROF. HUTT: There was one group described from Uganda by Patel *et al.* (1971). This was a Rwandan family in which there was also associated tropical splenomegaly. I think that we should search for a localised group where the incidence is abnormally high and I would be very interested to know whether, in South India, there are any groups that seem to be unduly susceptible. The most economical way to screen for something is to identify the susceptible group, because you cannot screen whole populations for a rare disease. In Uganda, I would choose the Rwandans.

DR SPRY: Can you tell us more about patients with the thrombotic stage of the disease in East Africa?

PROF. HUTT: We saw several cases where, as described by Dr Olsen, there was a large thrombus with a curious greenish colour in the left ventricle. We used to call these cases 'acute', but in fact when you look at the histology, it was clear that the process had been going on for some time.

DR J. J. PUIGBO: Venezuela, is in the same latitude as Nigeria and Uganda. We have the same typical clinical manifestations, the same pathology and we have about one third of cases with eosinophilia. We should look first at the population with eosinophilia. When the disease reaches the fibrotic stage, a very late stage, eosinophilia may no longer be present.

PROF. HUTT: Absolute eosinophil levels may not be the marker as we once thought. We should be looking for degranulated eosinophils. Such studies have not been done in Uganda.

DR OLSEN: We have not seen the earlier stages in a large number of African or other tropical countries. I think that it is on the latter that we should direct our attention. It is also a matter of tuition, particularly for those working in the bush. This was one of our aims in the Year of Tropical Cardiology. Dr Falase, who is one of our most active Council members, is in fact, concentrating on instruction of paramedical staff.

DR H. ACQUATELLA: The most common presentation is advanced heart failure, but we also have in our clinical material early cases. Two patients had typical chest pain and they underwent coronary arteriography which was normal, but the ventriculogram showed typical angiographic findings of endomyocardial fibrosis.

CHAIRMAN: On the point of early diagnosis. I think it is clinically very difficult because the disease may not come to light for a long time. Systolic ventricular function is well maintained until the late stage. Unless you get atrioventricular valve regurgitation or a myocardial infarction, the clinical diagnosis may be extremely difficult, even with angiography and isotope studies. With regard to a viral agent, is the distribution of EMF similar to Burkitt's lymphoma?

DR OLSEN: Several years ago the distribution of EMF was considered to be confined to the Burkitt's lymphoma belt. Subsequently, it was found that the condition is much more widely distributed.

DR SPRY: My view is that there must be potent environmental factors involved in the development of endomyocardial disease in the tropics, but a genetic factor cannot be ruled out. A genetic disorder which produces somewhat similar features to endomyocardial disease is carnitine

deficiency. We measured carnitine levels in sera from several of our patients and found they were normal. We must continue to look for other possible genetic factors. However, environmental factors are easier to pursue at the moment. Many different parasites can produce hypereosinophilia which could cause endocardial disease in the tropics. These parasites of course vary from region to region, but if this is the principal cause for endomyocardial disease, why has it not been described in many parts of the world where there is a very high incidence of parasitic diseases, such as Egypt and most of South East Asia and the Far East?

PROF. HUTT: I would like to say one more thing about the Rwandans. Shaper emphasized what he called the tropical immunological syndrome (Shaper, 1968). These people come from an area which is non-malarious due to its high altitude so that there is little malarial transmission. In addition to showing a much higher prevalence of EMF than other Ugandans, they also show a very much higher proportion of the tropical splenomegaly syndrome which is an abnormal immunological response to repeated attacks of malaria. We think that this may be genetically determined due to their origins from a non-malarial region. If we consider the aetiology of Burkitt's lymphoma, it is now postulated that the geographical distribution is related to endemic malaria in childhood. This alters their immune response so that EB virus infection becomes oncogenic. It seems possible that Rwandans react differently to malaria from most Ugandans and that this affects their immunological reactions. Subsequently, parasitic infections which may induce eosinophilia may then initiate cardiac lesions. It is always nice to have a single cause of a disease, but I do not think life is like that.

Reference

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