The relative prevalence of dermatomyositis and polymyositis in Europe exhibits a latitudinal gradient

G J D Hengstman, W J van Venrooij, J Vencovsky, H M Moutsopoulos, B G M van Engelen

The idiopathic inflammatory myopathies (IIM) are a heterogeneous group of systemic diseases that include the more familiar disease entities of dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM).¹ The aetiology of IIM is unknown. DM and PM are commonly regarded as autoimmune diseases,¹ while the immune response in IBM is thought to be secondary to degenerative changes in skeletal muscle tissue.² The autoimmune in DM and PM is most probably a result of the interaction of environmental and host factors.¹² Environmental risk factors are suggested by the fact that several viruses (for example, picorna, influenza, paramyxo) have been associated directly or indirectly with IIM, by unconfirmed reports of coxsackie B virus antibodies in children with DM, and by the fact that retroviruses are associated with an inflammatory myopathy pathologically indistinguishable from PM.¹² Host factors include sex and polygenic predisposition.¹²

Methods and Results

As part of a study of environmental and genetic risk factors for DM and PM we evaluated the relative prevalence of these diseases at nine rheumatological and neurological centres in nine European countries. Standardised questionnaires were sent and contained information on clinical signs and symptoms, course of the disease, response to treatment, results of laboratory investigations, electromyographic findings and muscle biopsy characteristics. A total of 381 questionnaires were returned. Incomplete questionnaires and questionnaires that contained insufficient information for an accurate diagnosis according to recently published criteria¹⁴ were excluded from the study. In total 153 patients with DM and 179 with PM were included. Statistical analysis was performed using the Cochran-Armitage trend test. A p value of less than 0.05 was considered statistically significant.

The relative prevalence of DM (DM/DM+PM) increased significantly (p<0.001) with geographical latitude going from northern Europe (Reykjavik, latitude 64, relative prevalence 0.08) to southern Europe (Athens, latitude 38, relative prevalence 0.56) (fig 1).

Discussion

The increase of the relative prevalence of DM may be either attributable to (a) a true increase of the prevalence of DM, (b) a decrease of the prevalence of PM, or (c) a combination of both. The cause of this gradient is unknown, but may include environmental as well as

Figure 1 Relative prevalence of DM (DM/DM+PM) as a function of the geographical latitude. DM: dermatomyositis; PM: polymyositis; GR: Greece; I: Italy; SLO: Slovenia; CZ: Czech Republic; UK: United Kingdom; NL: the Netherlands; S: Sweden; FIN: Finland; ICE: Iceland.
genetic factors. One of the possible environmental explanations is that ultraviolet B, through its various immunomodulatory actions, causes an increase of the prevalence of DM in a population with a particular genetic background.5

Although a selection bias cannot totally be excluded and the number of participating centres is comparatively small, this study on a large group of IIM patients, shows for the first time, a statistical significant latitudinal gradient of the relative prevalence of DM and PM, and may provide further insights in the environmental and genetic risk factors of these enigmatic diseases.

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