Clinical and Genetic Characteristics of Late-Onset Behçet’s Disease

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1. INTRODUCTION

In approximately 80% of patients with Behçet’s disease (BD), the first symptom occurs between the age of 16 to 35 years, and in 97% of patients before the age of 40 (Shizumi, unpublished data). Onset of the disease after age 40 is relatively rare. We evaluated the clinical spectrum of patients with BD in whom the disease manifested after age 40 or more, as well as the genetic factors that may be associated with this late expression.

2. PATIENTS AND METHODS

BD was defined according to the ISG criteria. Data from medical files and from patient interviews were collected. The results were analyzed separately for adults and late-onset patients with BD. Adult BD was defined if the first disease manifestation appeared between the ages of 18 and 40 years. Late-onset disease was defined if onset of BD symptoms occurred after the age of 40. Severity score was calculated as the sum of 1 point for each of mild symptoms, 2 points for each of moderate symptoms and 3 points for each of severe disease manifestations, according to Krause et al 1. The HLA-A, B and C tissue typing were performed as previously described 2. Statistical analysis was performed employing Student’s t-test for mean values and chi square ($\chi^2$) for table analysis.
3. RESULTS

Our study included 67 BD patients: 26 males (39%) and 41 females (61%). Thirteen patients (19.4% of all our BD patients) were identified as having late-onset BD.

3.1 Clinical manifestations of adult-onset BD

Oral aphthosis was the most common initial disease manifestation. All 13 patients presented first with recurrent aphthous stomatitis. Genital ulcers appeared in all 13 patients. Major eye involvement occurred in nine patients with adult-onset BD. Typical skin lesions occurred in three males and four females. A positive pathergy test was found at a similar rate in males and females, totaling five of 10 tested patients (50%). Nine patients (69.2%) had recurrent arthralgia, six patients (46.1%) had arthritis. Non-specific mild gastrointestinal symptoms occurred in seven patients (36.8%). None of the patients was suspected of having Familial Mediterranean Fever, which was also not reported in close family members. Pleuropulmonary manifestations appeared in one patient. Frequent headaches were reported by seven patients (36.8%). Other expressions of neuro-Behçet were diagnosed in another five patients. Vascular involvement, in the form of deep or superficial vein thrombosis of the lower limbs, was found in seven patients. A positive family history of oral aphthosis was reported in three patients (23%).

3.2 Adult-onset vs. adult BD

Major disease manifestations: The frequency of major manifestations in adult-onset BD and adult BD is presented in Fig. 1.

Minor disease manifestations: Similar prevalences were found in adult-onset and adult BD concerning overall joint disease, vascular involvement and recurrent headache.

The response to treatment was moderate in the two groups of patients. The curve of severity score increases from the age of 18 till the age of 35 to 40 at which time it starts to decline. Mean severity score of adult BD was $7.46 \pm 3.4$ and of the adult onset patients $7.77 \pm 2.61$ ($p = 0.7$).

Of the adult-onset patients, 85% were found to carry HLA-B5 whereas 66% of patients with adult BD carried HLA-B5.