Pemphigus Vulgaris in Two Sisters

Sir,

Pemphigus is a chronic bullous dermatosis with autoimmune etiology. It is a relatively rare disease that has been shown to occur with increased frequency in certain ethnic groups (1–2). Recently, we had the opportunity to see two sisters suffering from pemphigus vulgaris.

CASE REPORTS

The first patient, a 52-year-old Greek woman, was admitted to hospital with a great number of small, flaccid vesicles on the trunk and numerous erosions with intense oozing on the scalp. Skin biopsy showed suprabasal clefting with acantholytic cells, characteristic of pemphigus vulgaris. Both direct and indirect immunofluorescence tests gave positive results for pemphigus vulgaris. Human leukocyte antigen (HLA) typing showed the following: $A_{9(24)}$, $A_{19(32)}$, $B_{5(51)}$, B_{35}, C_{W4}, $DQ_{1(6)}$, $DQ_{3(8)}$, DR_4, $DR_{6(13)}$, DR_{W52}, DR_{W53}. The patient was treated with prednisolone 75 mg/day. As the lesions regressed, the steroid dose was gradually decreased and now the maintenance dose is 15 mg prednisolone daily.

Her younger sister, a 45-year-old Greek woman, had also suffered from pemphigus vulgaris for the last 7 years. When recalled, the clinical examination showed few, small erosions on her back. Skin biopsy and immunofluorescence tests showed features consistent with pemphigus vulgaris. HLA-typing showed the following: A_{28}, $A_{19(32)}$, $B_{16(38)}$, B_{35}, C_{W4}, $DQ_{3(7)}$, DQx, DR_4, $DR_{5(11)}$, DR_{W52}, DR_{W53}. She has been treated with prednisolone 40 mg/day, the dose being tapered following clinical improvement. Currently, the patient is on a maintenance dose of 10 mg prednisolone daily.

Sera, of both patients, were examined by the ELISA technique in order to ascertain the antigens of pemphigus vulgaris. The ELISA detected only desmoglein-3 in both cases.

DISCUSSION

Human leukocyte antigen typing in pemphigus has been determined in a wide variety of ethnic groups (3). From HLA typing in Greek Caucasians, an increased frequency of HLA-BW_{22} and HLA-B_7 was found (4). In another study of familial pemphigus vulgaris with oral manifestations in Greece, HLA typing revealed DR_4 antigen associated with all members, except one, of the families (5). The literature indicates an association between pemphigus vulgaris and HLA serotype DR_4 (6, 7). In our report, DR_4 haplotype was found in both sisters. The current review of the literature reinforces the importance of genetics in the pathogenesis of pemphigus (8), although the mode of inheritance of pemphigus is still unclear. Further familial studies are required before we can reach a definite conclusion.

REFERENCES

1. Park MS, Terasaki Pl, Ahmed AR, Tiwari JL. HLA-DR_{W4} in 91% of Jewish pemphigus vulgaris patients. Lancet 1979; 2: 441–442.
2. Spinowitz AL, Fiedler-Weiss VC, Fu T, Solomon LM. Pemphigus vulgaris in sisters. J Am Acad Dermatol 1986; 15: 115–116.
3. Feinstein A, Yorav S, Movshovitz M, Schewach-Millet M. Pemphigus in families. Int J Dermatol 1991; 30: 347–351.
4. Zervas J, Tosca A, Apostolakis I, Varelzidis A. HLA and pemphigus. Br J Dermatol 1979; 101: 357–358.
5. Laskaris G, Sklavounou A, Stavrou A, Stavropoulou K. Familial pemphigus vulgaris with oral manifestations affecting two Greek families. J Oral Pathol 1989; 18: 49–53.
6. Reohr PB, Mangklabruks A, Janiga AM, De Groot LJ, Banjasuratwong Y, Soltani K. Pemphigus vulgaris in siblings: HLA-DR_4 and HLA-DQ_{W3} and susceptibility to pemphigus. J Am Acad Dermatol 1992; 27: 189–193.
7. Katzenelson V, David M, Zamir R, Mellibovsky J, Idises C, Sandbank M. Familial pemphigus vulgaris. Dermatologica 1990; 181: 48–50.
8. Starzycki Z, Chorzelski TP, Jablonska S. Familial pemphigus vulgaris in mother and daughter. Int J Dermatol 1998; 37: 211–214.

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