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Evaluation of Serine α1-antitrypsin and Polymorphonuclear Leukocyte Elastase Contents and Their Immunogenetic Correlation in Psoriasis

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The purpose of our study was to quantify the serum content of α1-antitrypsin (α1-AT) and polymorphonuclear leukocyte elastase (PMN-E) in of 21 patients affected by active and stationary psoriasis, and 12 normal controls. HLA typing was also performed to identify a correlation among HLA antigens, age at onset of psoriasis and biochemical results. α1-AT levels were within the normal range in all patients, even in those with active, extensive, inherited and juvenile psoriasis, and in the controls. These data allow us to exclude, in our patients, the presence of rare or defective phenotypes, frequently associated with reduced serine levels of α1-AT. The PMN-E serine content was greatly increased in 3 patients, increased in 2, and slightly modified in 6 cases. All patients with the highest PMN-E levels reported a positive family history and absence of pulmonary, hepatic and atopic diseases. An increased psoriatic inheritance has been observed in the CW6-positive subjects (7/20), comparing B13 and DR6 antigen frequency. No correlation among HLA antigens, age at onset, clinical phase, or biochemical results could be established. ***Key words: psoriasis; α1-antitrypsin; elastase; HLA***

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Since proteolytic processes are prominent features in psoriasis, sera of 21 non-pustular and non-arthropathic psoriatic patients, genetically investigated for HLA antigens, were examined for α1-antitrypsin (α1-AT) and peripheral blood polymorphonuclear leukocyte elastase (PMN-E) contents.

It is well known that neutral serine proteinases such as PMN-E and their inhibitors can contribute to the pathogenesis of psoriasis because of their role as inflammatory agents causing damage to matrix components.

α1-AT, the most important serum inhibitor of PMN-E, protects the skin and pulmonar and hepatic tissues from the increased enzymatic activity of PMN-E. A decrease in serum concentration of neutral serine proteinase inhibitors, especially during flare and in patients with early onset of psoriasis, has been described (1), whereas increased elastase activity in neutrophils has been found in patients with active plaque lesions (2). These data may indicate that a protease-antiprotease imbalance could be related to the proteolytic tissue degeneration as the basal keratinocyte herniations observed in psoriatic skin (5).

The purpose of our study was to quantify the α1-AT and PMN-E serum contents in patients with active and stationary psoriasis and in normal controls. Human lymphocyte antigen (HLA) typing was also performed to identify a correlation among HLA antigens, age at onset of psoriasis and biochemical results, as proposed by several authors (4, 6).

PATIENTS AND METHODS

A total of 21 non-pustular and non-arthropathic psoriatic patients with active or stationary plaque lesions, randomly selected, and 12 normal healthy controls, have been studied for α1-AT and PMN-E serum concentrations. Pulmonar, hepatic, infective and joint diseases were absent in both groups.

In the performance of α1-AT test, a quantitative determination of antigen-antibody reaction was evaluated by rate nephelometry (Beckman Immunochemistry System, Calif., USA).

PMN-Elastase IMAC assay (Merck Diagnostica, Darmstadt, Germany) for elastase determination was employed.

Tissue typing for human histocompatibility antigens was applied in all patients studied.

RESULTS

α1-AT serum content was within the normal range for our control group (90–295 mg/dl) in all patients tested (Table I). PMN-E serum level was increased in patients nos. 18, 19, 20

Table I. *Total psoriatic patients: age of onset, clinical phases, α1-AT, PMN-E serum contents*

Patient	Age at onset	Clinical phase	α1-AT (mg/dl)	PMN-E (μg/l)
1. CA, f.	20 yrs	Active	141	43
2. NE, f.	25 yrs	Stationary	208	40
3. RM, m.	5 yrs	Stationary	151	59
4. DA, m.	31 yrs	Active	162	35
5. SA, m.	7 mo	Active	145	72
6. ML, m.	30 yrs	Stationary	163	60
7. TM, m.	4 yrs	Active	144	58
8. FR, f.	10 yrs	Stationary	175	124
9. VA, f.	30 yrs	Stationary	152	40
10. OE, f.	50 yrs	Stationary	160	50
11. BR, m.	22 yrs	Stationary	126	52
12. TN, f.	13 yrs	Active	145	41
13. RC, f.	30 yrs	Active	168	53
14. PA, m.	12 yrs	Stationary	128	50
15. PG, f.	20 yrs	Stationary	169	37
16. DG, m.	5 yrs	Stationary	129	50
17. BP, m.	54 yrs	Active	164	47
18. ZG, m.	40 yrs	Active	202	1967
19. DB, f.	30 yrs	Stationary	166	1504
20. MG, m.	3 yrs	Active	145	1596
21. GE, m.	40 yrs	Active	191	377

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Table II. *HLA antigens, age of onset, family history of psoriasis*

Patient	HLA antigens	Age at onset	Family history
1. CA	A2 A30 B13 B49 CW6	20 yrs	Negative
2. NE	A1 A26 B7 B37 CW6 DR2 DR5 DQ1 DQ3	25 yrs	Mother brother
3. RN	A24 A29 B17 B44 CW5 DR2 DR6 DQ1	5 yrs	Grandmother
4. DA	A1 A33 B14 B57 CW6 DR1 DR7	31 yrs	Mother
5. SA	A1 A2 B18 B35 CW4	7 mo	Mother
6. ML	A24 A30 B13 B44 CW6 DR1 DR2 DQ1	30 yrs	Grandmother
7. TM	A3 A23 B7 B44 CW4 DR2 DR5	4 yrs	Negative
8. FR	A24 A30 B7 B44 CW2 DR1 DR4 DQ1 DQ3	10 yrs	Mother
9. VA	A2 A24 B7 B18 CW2 DR4 DR8 DQ3	30 yrs	Negative
10. OE	A9 A30 B13 B18	50 yrs	Negative
11. BR	A13 A30 DR3	22 yrs	Negative
12. TN	A9 A30 B13 CW6	13 yrs	Sister
13. RC	A2 B13 CW4 DR6	30 yrs	Negative
14. PA	A24 A31 B51 B53 CW4 DR5 DR6	12 yrs	Negative
15. PG	A29 B44 CW5 DR2	20 yrs	Negative
16. DG	A3 A24 B18 CW6 CW7 DR1 DR5	5 yrs	Negative
17. BP	A2 B13 CW4 DR5	54 yrs	Negative
18. ZG	A2 B13 CW4 DR6	40 yrs	Brother
19. DB	A1 B7 CW5 DR1	30 yrs	Daughter
20. MG	A24 B44 CW6 DR2	3 yrs	Mother
21. GE	not done	40 yrs	Negative

Table III. *PMN-E level, inheritance, clinical phase*

Patient	PMN-E	Inheritance	Clinical phase
18, 19, 20	++++	Positive	Active: nos. 18, 20 Stationary: no. 19
8, 21	++	Positive: no. 8 Negative: no. 21	Active: no. 21 Stationary: no. 8
3, 5, 6, 7, 11, 13	+	Positive: nos. 3, 5, 6 Negative: nos. 7, 11, 13	Active: nos. 5, 7, 13 Stationary: nos. 3, 6, 11

(++++), 8, 21 (++), 3, 5, 6, 7, 11, 13 (+), as shown in Table I vis-à-vis the normal control value (2–42 μg/l).

All these serine data have been correlated to the age at onset (early onset = 0–20 yrs) and the different clinical phases (active – stationary) of the disease (Table I).

HLA antigen typing results, correlated to the age at onset and the family-history of psoriasis, are reported in Table II.

DISCUSSION

α1-AT levels were within the normal range in all patients, even in those with active, extensive, inherited or juvenile psoriasis, and in controls.

These data allow us to exclude the presence of the rare or defective phenotypes with very low levels of this enzyme, described in patients affected by inherited psoriasis, especially with early onset and during flare-up of the disease (1, 3).

PMN-E content was greatly increased in 3 patients (nos. 18, 19, 20), elevated in 2 cases (nos. 8, 21), and slightly modified in 6 patients (nos. 3, 5, 6, 7, 11, 13). All patients with high PMN-E levels reported a positive family history for psoriasis, but absence of pulmonary, hepatic and atopic diseases (Table III). These data need to be further investigated. We underline the results in cases 19 and 20, mother and daughter, respectively, in whom the biochemical values were similar, normal α1-AT content and very high PMN-E level, but the age at onset and the clinical phase of the disease were different.

We may suggest a genetic influence leading to an elevated PMN-E level despite different clinical features. All the patients investigated reported absence of either atopic dermatitis or contact dermatitis.

Regarding the HLA psoriatic antigens, CW6 was found in one-third of psoriatic patients (7/20), 4/10 cases with early onset and 3/10 cases with late onset of the disease. B13 was present in 7/20 patients, mostly with late onset of psoriasis; DR6 was revealed in 4/20 cases.

An increased positive family history in CW6 positive patients (5/7) can be highlighted by comparing B13 (3/7) with DR6 (2/4) antigen frequency, considered as genetic markers of inherited psoriasis. No correlation between HLA antigens and biochemical results could be established.

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