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Case Reports

A FAMILIAL CASE OF PEMPHIGUS VULGARIS

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ABSTRACT

A family in which two members are affected by pemphigus vulgaris is presented from Gilan. Up to now only 25 families in which more than one member was affected have been reported.[1]
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INTRODUCTION

Pemphigus is a chronic autoimmune bullous disease with unknown etiology. The bullae frequently affect skin and mucous membrane. In light microscopy pemphigus can be recognized by demonstrating acantholytic cells and bullae formation between epidermal cells.

Although pemphigus vulgaris mainly appears sporadically, but up to date more than 25 families have been reported with pemphigus.

In this case report two patients (aunt and nephew) are presented both suffering from pemphigus vulgaris.

CASE REPORTS

Patient 1

A 28 year old woman from Foman was admitted in 1982 with erosions on buccal mucosa and intertriginous areas.

Biopsy from skin lesions showed intraepidermal bullae with acantholytic cells compatible with pemphigus vulgaris. Routine tests all were normal and her blood group was O^+.

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She was initially treated with prednisolone 60 mg and azathioprine 150 mg daily. 4 weeks later while the lesions were improved, prednisolone was tapered to 20 mg/day.

Patient 2

A 60 year old woman from Foman, the aunt of our first patient, was admitted to hospital with skin and mucous membrane lesions. Physical examination revealed erosions and flaccid bullae over most parts of her body, mainly chest and intertriginous areas and oral mucosa. Nikolsky sign was positive.

Skin biopsy showed a suprabasal bullae with acantholytic cells. Routine tests were all normal. Her blood group was O^+.

She was treated initially with 120 mg prednisolone and 150 mg azathioprine.

DISCUSSION

Pemphigus is a rare autoimmune disease with antibodies against intercellular antigens. Bullae with acantholytic cells is formed within the epidermis.

According to our knowledge, the genetic basis of pemphigus vulgaris is not known but high prevalence of the disease in Jews and its correlation with some types of HLA such as HLA DR4, HLA A10, HLA DRW4,

HLA B7, HLA A13[3-5], and blood groups A and B[6-7] indicate the possibility of a genetic basis in pemphigus.

Here we present the first case of familial pemphigus in Iran. Our patients were 0^+, but unfortunately we could not do HLA typing.

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