

Myasthenia Gravis and Pemphigus Vulgaris

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Abstract

An unusual case of Myasthenia gravis, pemphigus vulgaris and hyperpigmentation on the dorsum of the fingers and toes is reported (JPMA 34: 349, 1984).

Case Report

A 25 years old female was admitted in Female skin ward of Mayo Hospital, Lahore on 11-8-1983. Four years prior to admission while working in the kitchen, about 15 days after her father's death, she suddenly felt intense weakness in the body especially the limbs and shoulders. It was a feeling as if she had no energy in her body. She fell and then recovered after 10 to 15 minutes of rest. This happened about once or twice a day and the frequency of such attacks gradually increased. Then the patient learnt that while doing her household work if she took 15 minutes rest after every 10 to 15 minutes, she could continue with her work. There was a history of diplopia in childhood, which was corrected by glasses.



Fig. 1 (b) Drooping of eyelids.



Fig. 1 (a) Drooping of eyelids.

She had drooping of eyelids (Fig. 1, a-b) alongwith loss of energy in the limbs and difficulty in talking with weakness of the tongue during prolonged conversation. There was aggravation of symptoms with excitement, emotional distress, and infection. She was diagnosed as Myasthenia Gravis and markedly improved with oral Neostigmine.

In June, 1983 she developed vesiculo. bulious lesions on the scalp and face and after 2 to 3 days similar lesions appeared on the trunk, especially on the back, chest and abdomen and few on the limbs (Fig.2).



Fig. 2 Vesiculobulious lesions on abdomen.

The bullae were upto one inch in diameter and contained clear fluid. They were flaccid and burst after one day and left a wide raw area more than the size of the original bulla. There was no history of mucous membrane involvement or drug intake.

Dermatological examination revealed bullous lesions and hyperpigmented macular esioris on the above mentioned areas. The bullae, which appeared on normal skin were with 2.5 cms diameter and contained clear fluid. Raw areas more than the size of the bullae covered crusted areas at certain places were also seen (Fig. 2). Nikolsky sign was positive.

Her haemoglobin was 10.2G% and ESR 20 mm 1st hour, total and differential leucocyte count, urinalysis and stool examination were normal. Tzanck test was positive.

Histological examination (Fig. 3 a & b) showed suprabasal bulla.



Fig. 3(a) Acanthocytic cells in the cavity of Bulla.

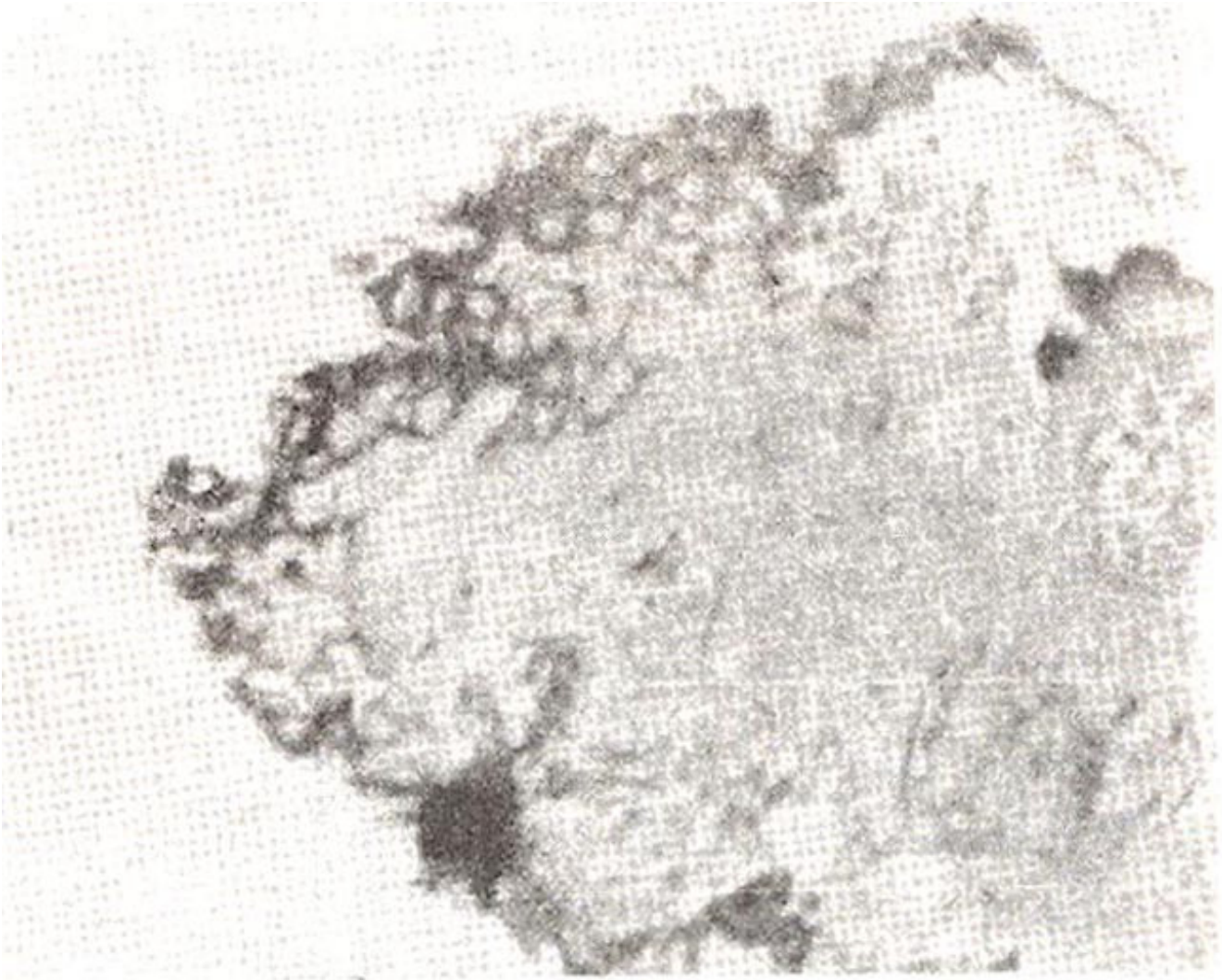


Fig. 3 (b) Bulla covered with epidermis.

The bulla was covered with many layers of epidermis. A few acantholytic cells were seen in the cavity of the bulla.

She was initially given 75 mg of prednisolone and the lesions started subsiding after one week. The dose of steroids was brought down to 15 mg/day over a period of one month.

Comments

Association of pemphigus erythematosus with systemic lupus Erythematosus, Myasthenia

Fig. 3(a) Acanthocytic cells in the cavity of Bulla. gravis, thymoma, Seropositive Rheumatoid arthritis is well known.

Senear and Usher¹ are the pioneers who described this syndrome as having features of both pemphigus and lupus erythematosus. Is this syndrome a localised or an abortive type of pemphigus foliaceus, or can it change to pemphigus vulgaris or pemphigus foliaceus, or is it a separate entity, remains debatable.

Immunological changes characteristic of both pemphigus and Lupus Erythematosus, in patients with senear usher syndrome have been described.^{2,3} Immunoglobulins and complement were found within the intercellular spaces of the epidermis as well as at the dermo-epidermal junction while using the fluorescence antibody technique.

Beutner⁴ described a patient with myasthenia gravis, a malignant thymoma and Pemphigus

Erythematosus. By indirect immunofluorescence, he found antibodies to the cross striations of skeletal muscle, and to intercellular areas of stratified squamous cell epithelium.

Myasthenia gravis is caused by an immunologic disturbance and a close relationship exists between the presence of intercellular antibodies and pemphigus.

The case reported here may be a variant of pemphigus erythematosus which changes into pemphigus vulgaris or this association may be a coincidence.

References

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