HERPETIFORM PEMPHIGUS

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In a case of pemphigus foliaceus, the initial clinical presentation was like dermatitis herpetiformis. The patient responded to low doses of corticosteroids.

Key words : Herpetiform pemphigus, Pemphigus foliaceus, Dermatitis herpetiformis.

The term herpetiform pemphigus was coined by Jablonska et al[1] in 1975 for those clinical cases in which the clinical picture initially resembles that of dermatitis herpetiformis in the type of eruption, its symmetry and pruritus. Only in the later stages, there is typical clinical and histopathological manifestations of pemphigus. Some of these patients respond to sulphones and others to corticosteroids. The immunofluorescence findings, both direct and indirect, are typical of pemphigus. There have been a few case reports of herpetiform pemphigus in the literature.[2-5] A case of herpetiform pemphigus, recently seen by us is being reported.

Case Report

A 50-year-old man was admitted because of the development of a papulo-vesicular eruption with severe pruritus that had appeared 20 days earlier. Examination revealed a polymorphous rash composed of oedematous papules, 3-4 mm in size, pinkish red in colour, arranged in a herpetiform pattern. Tiny vesicles were also seen, some on an erythematous base and some on an apparently normal skin. Some vesicles were having clear contents while a few were purulent. The lesions were distributed on the back, arms and legs and were symmetrical. The mucosa and nails were normal. Nikolsky's sign was negative. Routine laboratory tests were normal. The vesicular fluid contained 50% eosinophils. A biopsy from a small vesicular lesion on an erythematous base revealed a mild perivascular lymphocytic infiltrate in the upper dermis. Because of the polymorphous nature of the rash, severe pruritus and grouping of lesions, a clinical diagnosis of dermatitis herpetiformis was made. The patient was put on 200 mg of dapsone daily. However, there was no improvement. The lesions persisted with severe itching and new lesions continued to appear. A second biopsy of a vesicular lesion revealed formation of bullae in the granular cell layer and upper part of stratum spinosum. Acantholytic cells were also observed, and within the bullae a small number of neutrophils and occasional eosinophils were present. The dermis showed oedema involving some of the dermal papillae and there was an inflammatory reaction in the dermis with predominance of neutrophils, mononuclear cells and eosinophils. The histopathology was indicative of pemphigus foliaceus. The patient was put on 30 mg prednisolone a day to which he responded dramatically. This was then gradually reduced and tapered off to 10 mg a day. The patient has been on regular follow up since then and there has so far been no relapse.

Comments

Pemphigus and dermatitis herpetiformis are two separate entities with distinct clinical, histopathological and immunofluorescent findings. However, it has been known for years that pemphigus foliaceus in its first stages may be similar in its clinical appearance to dermatitis herpetiformis. Since the initial description

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of such a case as early as 1892 by Brocq,[6] there have been only a few case reports in the world literature[2-5] in which there has been transition from dermatitis herpetiformis to pemphigus foliaceus. Seah et al in 1973[7] reported a case in which the clinical picture and response to sulphapyridine appeared to be typical for DH, but the histopathologic and immunofluorescence findings were diagnostic of pemphigus. They titled this case as pemphigus controlled by sulpapyridine. The term mixed bullous disease as suggested by Barranco and Tulsa[8] for such cases is misleading as it suggests coexistence of two separate entities. Herpetiform pemphigus is perhaps the most apt and suitable term for these cases as suggested by Jablonska et al. Recently Ingber and Feuerman[9] reported 5 patients of herpetiform pemphigus which had been followed up for periods ranging from 5 to 14 years. The disease was benign in nature and required low doses of corticosteroids for control. It is perhaps worthwhile emphasizing here that in all cases of herpetiform pemphigus immunofluorescence findings are typical of pemphigus throughout the course of the disease.[1] However, in centres where such facilities are not available, one has to base the diagnosis on clinical, histopathological and therapeutic aspects as in our case.

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