LETTER TO THE EDITOR

Epidermolysis Bullosa Acquisita
Versus Pemphigoid

I would like to comment on the interesting patient with epidermolysis bullosa acquisita (EBA) described by Livden et al. (3) recently. The authors acknowledge that their immunological findings resembled those seen in bullous pemphigoid. However, they then dismiss this diagnosis for dubious reasons. They argue that their failure to detect circulating antibasement membrane zone antibodies tells against the diagnosis. However, in a survey of many publications Alexander (2) concluded that 22% of patients with active pemphigoid did not have demonstrable circulating antibodies and this was the case in 7 out of 36 (19%) patients in a recent series (1). Livden et al. also suggest that the finding of immunoglobulin and complement in non-lesional skin (as in their patient) is not typical of pemphigoid. However, this is not the case, and the reference which they cite (4) in support of their views does in fact state the opposite.

Finally they claim that the basement membrane deposits of immunoglobulin were granular as opposed to the typical linear pattern in pemphigoid. In my own experience the classical description of a "linear or tubular" pattern of immunofluorescence in pemphigoid rather oversimplifies the usual situation. The typical pattern is often interrupted by areas which are much more irregular. Inspection of many published photomicrographs of pemphigoid immunofluorescence will confirm this. Factors which appear to contribute towards an irregular staining pattern include very intense fluorescence, thick sections and sections in which the basement membrane zone is cut obliquely. This latter phenomenon is frequently seen at the tips of papillae and rete pegs and is beautifully illustrated in figure 4 of their paper in which the arrow appears to show not an early bulla as claimed, but an area where the basement membrane is obliquely sectioned. Moreover the classical linear pattern of pemphigoid is well illustrated in the left hand part of this photomicrograph.

I believe that this patient has cicatricial pemphigoid and the finding of linear basement membrane IgA (in addition to IgG) in the oral mucosa as well as the absence of circulating antibodies are all consistent with this diagnosis (5).

I have recently noted blisters occurring following minor trauma (excoriation) in 2 patients with otherwise typical bullous pemphigoid and in a child with juvenile pemphigoid (to be published). In retrospect, I think that this is quite a common, but usually minor and overlooked feature of the disease. The situation is analogous to the Koebner phenomenon in psoriasis in that lesions induced by trauma rarely dominate the clinical picture, although occasionally they may. I believe that at least some cases of so-called epidermolysis bullosa acquisita represent an unusual variant of pemphigoid in which trauma is the dominant localizing trigger factor. I would suggest that patients considered to have EBA should be carefully and, if necessary, repeatedly investigated by direct immunofluorescence. Studies on such patients by immunoelectronmicroscopy should prove extremely interesting.

REFERENCES

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