Esophageal Nikolsky's Sign in Pemphigus Vulgaris

Nikolsky's sign, the easy separation of the outer layers of the skin caused by rubbing (1), is a valuable diagnostic finding in cutaneous pemphigus vulgaris. It can be elicited by applying friction to the skin with either the fingertips or a cotton swab, which leads to the production of a large and unusual bullous lesion (1). Although the involvement of the esophagus in pemphigus vulgaris has been reported sporadically (2–5), esophageal Nikolsky's sign has not previously been reported. We describe here the case of a patient with pemphigus vulgaris with esophageal involvement, in whom a characteristic Nikolsky's sign appeared in the esophageal mucosa.

A 61-year-old man with pemphigus vulgaris affecting both the skin and the oral mucosa, who was attending the Dermatology Outpatient Clinic, was referred with an eight-month history of dysphagia for solid food and loss of weight. Upper gastrointestinal endoscopy showed a normal esophagus, with no abnormalities with regard to esophageal shape or volume. The esophageal mucosa also appeared to be normal (Figure **1**). However, applying friction to the surface of the esophageal mucosa with a closed biopsy forceps caused the mucosa to detach easily from its inner layers, producing a large, flaccid bullous lesion, which left an exposed surface when removed (Figure **2**). Histopathological examination of an esophageal mucosa biopsy specimen showed characteristic intraepithelial suprabasilar vesicles, associated with acantholysis. The patient was subsequently treated with a combination of steroids and azathioprine, with complete resolution of the symptoms.

Pemphigus vulgaris is an autoimmune disease of unknown cause that may affect the skin and mucosal surfaces (2). However, the involvement of the esophageal mucosa seems to be quite rare (3,4). In a careful review by Goldeberg and Weiss of 11 cases reported in the world literature, it was emphasized that Nikolsky's sign in the esophagus was not reported in any of the cases (5). In the present report, esophageal Nikolsky's sign was easily reproduced, and led to a large esophageal erosion. It is likely that in common circumstances, the passage of solid food in close contact with esophageal mucosa affected by pemphigus vulgaris may cause mucosal detachment,

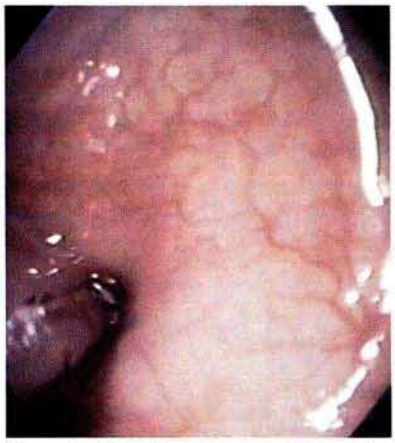

Figure 1: Normal appearance of the esophageal mucosa in a patient with pemphigus vulgaris, just before Nikolsky's sign was elicited.

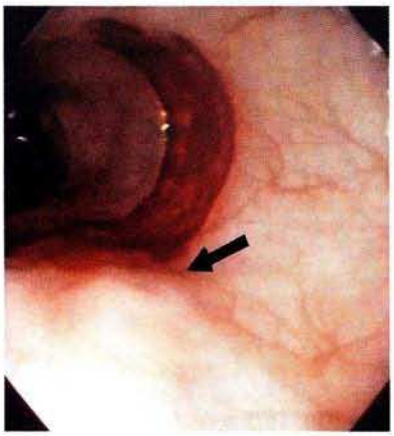

Figure 2: Appearance of the esophageal mucosa after eliciting Nikolsky's sign. Note the detachment of a large area of mucosa, and the bullous lesion (arrow).

thus producing symptoms of dysphagia or odynophagia, or both.

In conclusion, we recommend that an attempt to reproduce Nikolsky's sign in the esophagus should be made in any patient with pemphigus vulgaris and dysphagia or odynophagia, as this may confirm the diagnosis of esophageal involvement.

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