Bullous Pemphigoid Presenting with Esophagitis Dissecans Superficialis

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Key words: Bullous pemphigoid, Esophagitis dissecans superficialis

An 87-year-old woman was admitted with a more than 6-month history of an exacerbation of bullae. Edematous erythema and bullae were scattered throughout her extremities and trunk (Picture 1A). Serum autoantibodies against the BP-180 antigen were found. Histopathology confirmed a diagnosis of bullous pemphigoid (Picture 1B). She developed hematemesis on the fourth day after admission. Upper endoscopy revealed a swollen uvula and epithelial exfoliation in the oral cavity (Picture 2A, 2B). The esophageal mucosa showed longitudinal sloughing, indicating esophagitis dissecans superficialis (Picture 3). Exfoliation at the pharyngoesophageal junction at the points of contact with the endoscope suggested positive Nikolsky’s phenomenon. Follow-up endoscopy four weeks after treatment with prednisone revealed evidence of mucosal regeneration in the esophagus without any constriction (Picture 4).

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Received: March 2, 2017; Accepted: May 8, 2017; Advance Publication by J-STAGE: October 16, 2017
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In bullous pemphigoid, skin involvement is predominant, while esophageal involvement is rare, in contrast to pemphigus vulgaris. In this case, systemic corticosteroid therapy was effective for the treatment of both the skin and esophagus.

The authors state that they have no Conflict of Interest (COI).

References