

A case of esophageal involvement of Pemphigus vulgaris

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Background: Pemphigus vulgaris [PV] is the most common type of a group of autoimmune disorders called pemphigus blistering disorders. Typically, it presents on the mucous membranes which are found in areas including the mouth, throat, nose, eyes, genitals, and lungs. The involvement of the esophagus is rare. This case report describes a patient who has confirmed the esophageal involvement of PV.

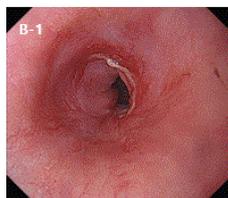
Case: A 74-year-old female with known PV and esophageal ulcer visited gastroenterology department for follow-up Esophagogastroduodenoscopy [EGD]. PV had been pathologically confirmed in this patient 9 years ago through skin biopsy. She had been on maintenance therapy, oral methylprednisolone 6mg and mycophenolate mofetil [MMF] 500 mg daily. She underwent EGD and colonoscopy 6 month ago due to weight loss and CEA elevation (11.3ng/ml). The EGD showed an esophageal ulcer, but the biopsy could not be performed due to the risk of bleeding while she was on steroid for a long time. She was prescribed and maintained proton pump inhibitors [PPI]. At this repeated EGD, the esophageal ulcer did not improve and was slightly aggravated. To rule out malignancy, biopsy was performed cautiously but short-length dissections began to occur from the site of biopsy. Scope was retrieved immediately to prevent further dissection and she was hospitalized with chest CT confirmation and serial monitoring of chest x-rays. After 3 days of fasting, a gastrograffin esophagography confirmed that the damage of the mucosa did not progress to perforation. The pathologic findings of the esophageal ulcer showed surface epithelial detachment and acantholysis of epithelium with necropurulent exudates and reactive atypia. Based on the patient's clinical history and pathologic findings, the esophageal involvement of PV can be diagnosed. She maintains the oral methylprednisolone, MMF and PPI without esophageal symptomatic worsening. A follow-up endoscopy to monitor progress is being carefully considered. This case instructively demonstrated that gastrointestinal involvement in PV can persist for a considerable period during treatment, and the endoscopic biopsy can lead to serious complications.

Fig A. multiple large erosions and desquamation on soft and hard palate (2020-12-01)



Fig B. Esophagus ulcer on UGI endoscopy

B-1) initial finding 6 month ago



B-2) slightly aggravating finding on repeated UGI endoscopy

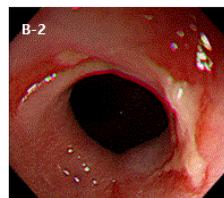


Fig C. Pathologic findings of esophageal ulcer showing surface epithelial detachment and acantholysis with necropurulent exudates and reactive atypia

