Scarring mucous membrane pemphigoid presenting as double stenosis of the larynx and esophagus: precautions during therapy can avoid complications

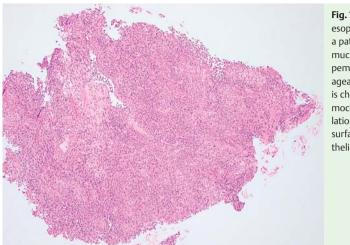
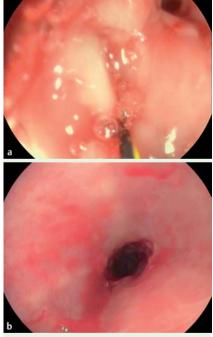


Fig. 1 Severely altered esophageal mucosa in a patient with scarring mucous membrane pemphigoid. An esophageal biopsy specimen is characterized by plasmocyte-enriched granulation tissue and lacks surface squamous epithelium (× 100).



**Fig.2** a Esophageal stenosis before development of the complication (no residual lumen, wire-guided esophageal entrance, laryngeal stenosis at top right). b Esophageal stenosis after endoscopic therapy and before percutaneous endoscopic gastrostomy placement.



**Fig. 3** Laryngeal stenosis due to mucous membrane pemphigoid involvement. (A perforation was not clearly visible during or after development of the complication.)

Mucous membrane pemphigoid (MMP) is a chronic immune bullous disease involving predominantly the mucous membranes. It is characterized by the formation of autoantibodies against components of the dermal-epidermal junction. The incidence of aerodigestive involvement is approximately 5% to 15% [1-3]. A 41-year-old woman with MMP-associated esophageal strictures was admitted periodically to our hospital for dilation treatments (seven endoscopic dilation treatments in 30 months). At the time of an endoscopic examination, which we report here, the patient was experiencing third-degree dysphagia in addition to progressive stridor due to laryngeal involvement.

The pathologic and endoscopic findings included severely altered esophageal mucosa (**•** Fig.1) with an impassable stricture from 17 to 25 cm (**•** Fig.2). In addition, laryngeal involvement could be detected, with altered mucosa and loss of mobility of the vocal cords (**•** Fig.3).

Because of the stridor, a bronchoscope (diameter of 5.5 mm) was first placed for translaryngeal oxygenation during the dilation therapy. The larynx was restricted by inflammatory alterations, but the bronchoscope could easily pass the stenotic area. Subsequently, esophagoscopy was performed with a thin-caliber gastroscope (8 mm).

While the patient was being prepared for the esophageal dilation procedure, emphysema of her whole body suddenly developed, with severe compromise of her respiratory and circulatory function, and short-term resuscitation was required. Orotracheal intubation was performed immediately, and the patient was transferred to our intensive care unit.

Computed tomography showed generalized emphysema and pneumothoraces (**•** Fig. 4). Thoracic drains were placed bilaterally, and broad-spectrum antibiotics were started. After 5 days, a percutaneous endoscopic gastrostomy was placed via direct puncture technique. After 6 days, extubation was possible, and the patient could breathe without stridor or impairment of her respiratory function.

In conclusion, MMP can involve the upper respiratory and digestive tracts. The therapy of symptomatic laryngotracheal stenosis should be the first step taken to avoid life-threatening complications during the endoscopic treatment of esophageal strictures.

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Fig. 4 a, b Computed tomography demonstrating generalized emphysema and bilateral pneumothoraces.

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## Bibliography

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