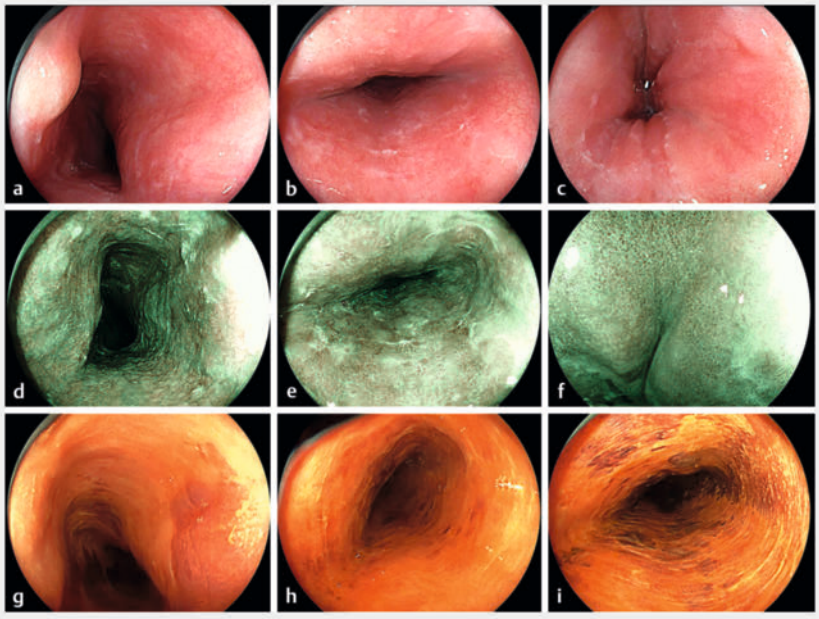
A rare case of primary esophageal Paget's disease with underlying invasive adenocarcinoma

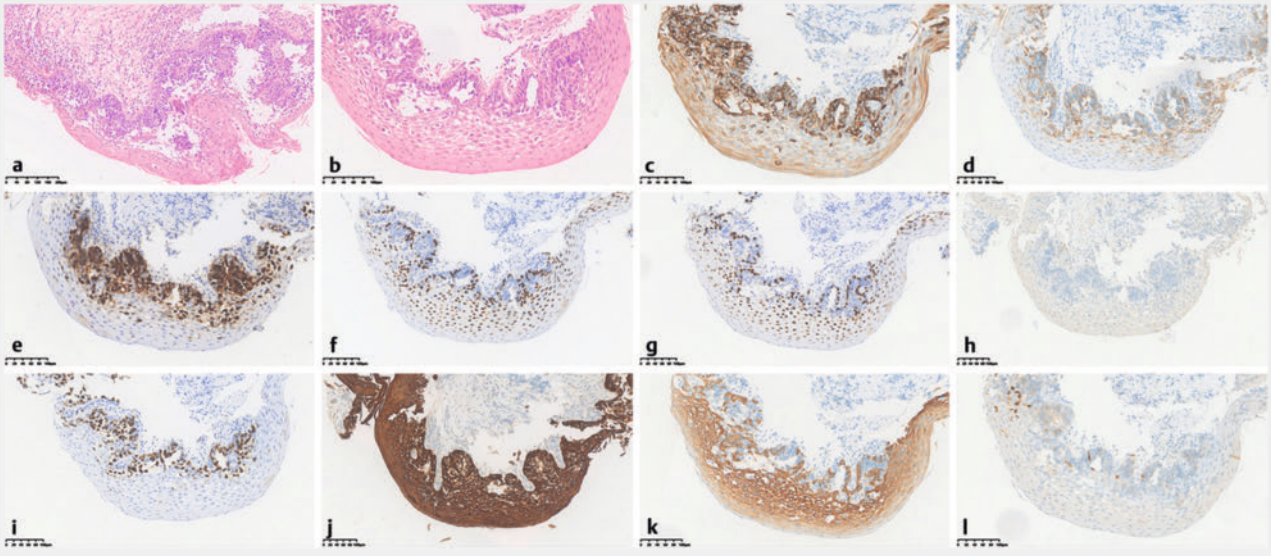
OPEN ACCESS

A 53-year-old man presented to Ruijin Hospital, having been followed up for 9 months with an esophageal lesion that had initially been detected during a routine esophagogastroduodenoscopy (EGD) and had been diagnosed histopathologically as a high grade intraepithelial neoplasia (HGIN).

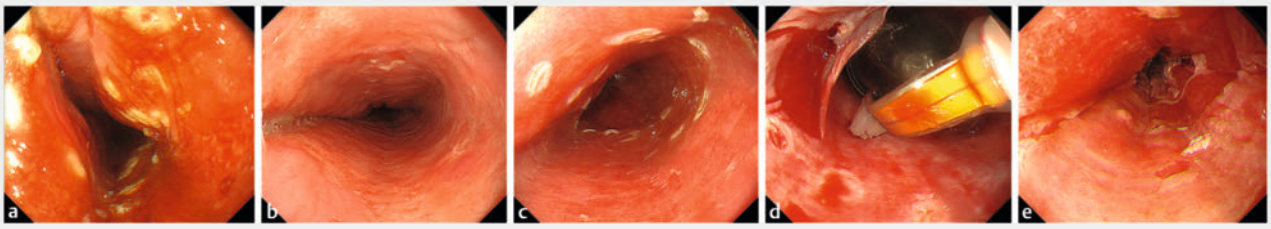
The inpatient EGD revealed a flat (0-IIb) and slightly reddish lesion spanning from 25 to 38 cm of the middle-to-lower esophagus (▶ **Fig. 1**). Magnifying endoscopy with blue-laser imaging (ME-BLI) identified an intrapapillary capillary loop (IPCL) pattern consistent with type B1 (▶ **Fig. 1 d–f**). Notably, a 5 × 5-mm slightly elevated area with mild congestion was observed within the lesion at 26 cm (▶ **Fig. 1 a**). The lesion remained unstained after the application of Lugol's solution and exhibited a partial pink-color sign from 30 to 33 cm of the esophagus, without any signs of deep invasion (▶ **Fig. 1 g–i**). Pathology and immunohis-



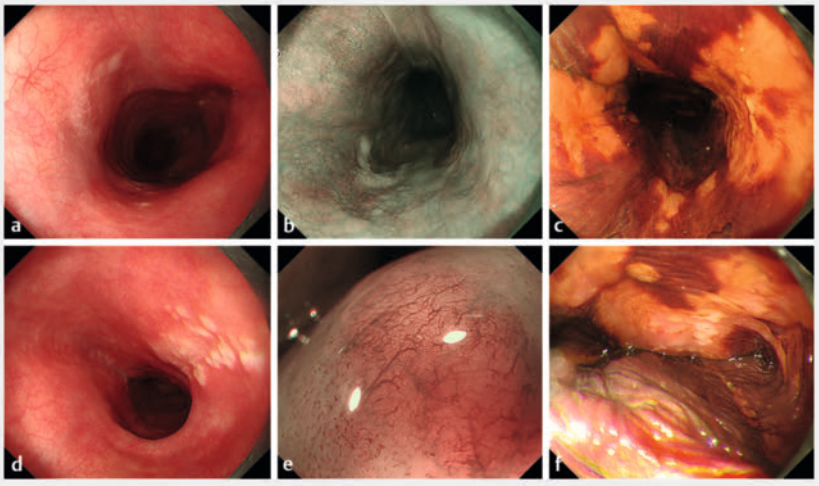
▶ **Fig. 1** Endoscopic features of the lesion before radiofrequency ablation on: **a–c** white-light endoscopy; **d–f** magnifying endoscopy with blue-laser imaging; **g–i** after staining with Lugol's solution.



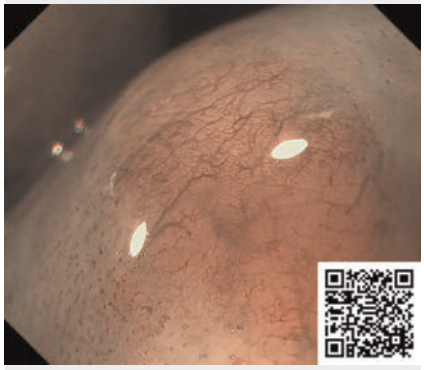
▶ **Fig. 2** Microscopic appearance of the of biopsy specimen on: **a, b** hematoxylin and eosin (H&E) staining; **c–l** immunohistochemical staining with: **c** CK7(+); **d** CK19(+); **e** P53(missense mutation); **f** P63(little+); **g** P40(little+); **h** CK20(−); **i** Ki67(70%+); **j** AE1/AE3(+); **k** CK5/6(partial+); **l** P16(−).



▶ **Fig. 3** Endoscopic images during radiofrequency ablation therapy.



▶ **Fig. 4** Endoscopic images during follow-up esophagogastroduodenoscopy 2 months after treatment on: **a, d** white-light imaging; **b, e** magnifying endoscopy with narrow-band imaging; **c, f** after staining with Lugol's solution.



Video 1 Features of a rare primary Paget's disease of the esophagus under white-light endoscopy and magnifying endoscopy with blue-laser imaging/narrow-band imaging before and after radiofrequency ablation treatment.

tology of the biopsy specimen revealed CK7+, CK19+, P53(missense mutation), P40(little+), P63(little+), CK5/6(partial+), P16(little+), Ki67(70%+), AE1/AE3+, CAM5.2+, CK20(−), SOX-10(−), villin(−), HER2(0), SATB2(−), GCDFP-15(−) (▶ **Fig. 2**), suggesting (i) extramammary Paget disease; (ii) invasive adenocarcinoma with Paget dissemination (M1 for the biopsy, more tissue would be needed for the evidence of invasive adenocarcinoma) [1–4].

Given the size of the lesion and the patient's refusal to undergo surgery, we performed radiofrequency ablation therapy (▶ **Fig. 3**). A follow-up EGD at 2 months post-treatment revealed persistent faintly red, rough mucosa extending from 26 to 38 cm of the esophagus (▶ **Fig. 4**). Notably, brownish areas were observed within the lesion under ME with narrow-band imaging (ME-NBI), with the majority of type B1 IPCL with partial type R vessels (▶ **Fig. 4 b, e**). Subsequent Lugol's solution staining delineated an irregularly geographically distributed lesion with partial circumferential involvement (▶ **Video 1**). A biopsy taken at 30 cm demonstrated a pink-color sign (▶ **Fig. 4 f**), confirming the previous diagnosis.

The patient currently continues on regular EGD follow-up every 3 months at his local hospital. If the lesion were to worsen during follow-up, chemoradiotherapy would be considered.

Endoscopy_UCTN_Code_CCL_1AB_2AC

Conflict of Interest

The authors declare that they have no conflict of interest.

The authors

Xue Chen‡[1], Heng Zhang‡[2], Aihua Qian[1], Xi Chen[1]

1 Department of Gastroenterology, Shanghai Jiao Tong University Medical School Affiliated Ruijin Hospital, Shanghai, China

2 Department of Pathology, Shanghai Jiao Tong University Medical School Affiliated Ruijin Hospital, Shanghai, China

Corresponding author

Xi Chen, MD

Department of Gastroenterology, Shanghai Jiao Tong University Medical School Affiliated Ruijin Hospital, 197 Ruijin Second Road, Shanghai 200025, China

cx11977@rjh.com.cn

‡ These authors contributed equally.

References

[1] Matsukuma S, Aida S, Shima S et al. Paget's disease of the esophagus. A case report with review of the literature. Am J Surg Pathol 1995; 19: 948–955

[2] Liu X, Zhang D, Liao X. Paget cells of the esophagus: A clinicopathologic and immunohistochemical study of 10 cases. Pathol Res Pract 2023; 242: 154345. doi:10.1016/j.prp.2023.154345

[3] Abraham SC, Wang H, Wang KK et al. Paget cells in the esophagus: assessment of their histopathologic features and near-universal association with underlying esophageal adenocarcinoma. Am J Surg Pathol 2008; 32: 1068–1074

[4] Lu C, Jiang N, Shi L et al. Primary Paget's disease with infiltrating adenocarcinoma of esophagus: report of case. Zhonghua Bing Li Xue Za Zhi 2023; 52: 736–738

Bibliography

Endoscopy 2024; 56: E740–E742
DOI 10.1055/a-2378-6256
ISSN 0013-726X

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany