Rare Primary Esophageal Paget’s Disease Diagnosed on a Large Bloc Specimen Obtained by Endoscopic Mucosal Resection

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ABSTRACT

Primary esophageal Paget's disease is rare. Only a few case reports have described the intraepithelial papillary capillary loop (IPCL) pattern obtained by magnified Narrow Band Imaging (M-NBI) endoscopy in this rare pathology. This report highlights the usefulness of M-NBI and the successful diagnosis using a large bloc specimen obtained by endoscopic mucosal resection with the cap method (EMR−c). A 53-year-old man was referred to endoscopic examination for dysphagia. The endoscopic image revealed a ring-shaped scarring of the esophagus suggestive for eosinophilic esophagitis. The IPCL pattern by M-NBI endoscopy showed an inflammatory pattern, and the entire epithelium of the esophagus was not stained by Lugol iodine spraying. Based on six biopsies randomly performed, a poorly differentiated adenocarcinoma was diagnosed. Since the M-NBI pattern and the histology were completely different, EMR−c was performed to obtain large bloc specimens for a more detailed diagnosis. The pathological findings revealed extensive Paget's cells infiltration into the epithelium and multifocal invasion from the mucosa to the submucosal layer with adenocarcinoma. In conclusion, a large bloc specimen by EMR-c might be more useful than a small biopsy for an accurate diagnosis of the rare esophageal Paget's disease.

Key words: Primary Paget's disease – large bloc specimen – endoscopic mucosal resection.

INTRODUCTION

Primary Paget's disease of the esophagus is very rare; only a few cases have been reported to date [1, 2]. Conventional Lugol chromoendoscopy is used to diagnose the malignant potential lesion of the esophagus. This technique has some limitations for the correct assessment of Lugol-voiding lesions (LVLs) [3]. Magnified Narrow Band Imaging (M-NBI) endoscopy might be used for a better characterization of LVLs [4]. This technique cannot accurately diagnose Paget's disease in the esophagus. Endoscopic mucosal resection with the suction and cap method (EMR−c) allows the pathologist to obtain a large en bloc specimen for the accurate diagnosis of primary Paget's disease of the esophagus.

CASE REPORT

A 53-year-old man, a heavy drinker, was referred for endoscopic examination having dysphagia. The esophageal mucosa presented on the whole circumference a ring-shaped scarring, suggestive for eosinophilic esophagitis, without erosive or ulcerative lesions (Fig. 1). The intraepithelial papillary capillary loop (IPCL) pattern described based on the M-NBI endoscopy showed an inflammatory pattern (Fig. 2). Not all the esophageal epithelium was stained by the Lugol iodine spraying, suggesting malignancy (Fig. 3). Based on six biopsies randomly performed, the diagnosis was of poorly differentiated adenocarcinoma. The macroscopic appearance of the mucosa, the M-NBI endoscopy pattern and histology were discordant. Hence, EMR−c was performed in order to obtain a larger en bloc specimen. The histopathological examination, hematoxylin and eosin (H&E) stain showed intraepithelial large and round-shaped tumor cells spreading...
within the entire layer, characteristic for Paget’s disease (Fig. 4). The periodic acid-Schiff with diastase (PAS-D) staining was partially positive (Fig. 5). The immunohistochemistry staining for mucins, cytokeratin (CK) 18 (Fig. 6) and E-cadherin was positive (Fig. 7), differentiating between malignant melanoma, poorly differentiated adenocarcinoma, poorly differentiated squamous cell carcinoma, undifferentiated carcinoma, metastatic carcinoma. The Paget’s cells infiltrated the epithelium in the esophagus extensively; these cells multifocally invaded the mucosal and submucosal ducts, indicating adenocarcinomatous differentiation. There was no primary lesion such as squamous cell carcinoma, or adenocarcinoma based on Barrett’s esophagus. The final histological diagnosis was a primary Paget’s disease of the esophagus. Moreover, Positron-Emission Tomography and Computed Tomography (PET-CT) revealed the up-take of 18-fluoro deoxy glucose (18FDG) only in the esophagus. Therefore, an esophagectomy with partial gastrectomy was performed.

**DISCUSSION**

According to several reports of primary esophageal Paget’s disease, the patients’ main complaints were dysphagia or chest discomfort similar to reflux esophagitis [2]. As Paget’s cancer cells grow extensively superficially and multifocally into the mucosal and submucosal ducts without any protruded lesions,
This report is of the first patient in whom an accurate diagnosis was established by EMR–c for rare primary Paget’s disease of the esophagus.

**CONCLUSION**

A large bloc specimen by EMR-c might be more effective than a simple biopsy to obtain sufficient sample for the accurate diagnosis of esophageal Paget's disease.

**Conflict of interest.** The authors declare no conflict of interest and no financial arrangement with any company.

**Authors’ contributions:** M. H. and A. M. were responsible for devising the research and writing the manuscript. K. H., G. Y., N. N. participated equally in the work. Prof. M. T. provided a critical revision of the manuscript for intellectual content and was responsible for the final approval of the manuscript.

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