Russell Body Barrett’s Esophagus

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ABSTRACT

We report an unusual case of Barrett’s esophagus with prominent intramucosal Russell bodies, also known as Russell body Barrett’s esophagus. We present this case to emphasize the importance of recognizing this unusual entity. It also represents a potential diagnostic pitfall because the distended plasma cells may be mistaken for signet ring cells of gastric adenocarcinoma or low-grade lymphoma. Hence, an awareness of this entity is important to avoid diagnostic confusion.

INTRODUCTION

Russell body gastroenteritis is a rare chronic inflammatory condition characterized by abundant intramucosal polyclonal plasma cells containing intracytoplasmic eosinophilic immunoglobulin globules called Russell bodies (RBs). It has been described as RB esophagitis or RB gastritis or RB enteritis, based on the site of involvement, although it occurs most commonly in the gastric antrum, with rare case reports of esophageal and duodenal involvement. The association of Barrett’s esophagus with RBs is extremely rare and understudied in the literature. We report an unusual case of Barrett’s esophagus with prominent RBs.

CASE REPORT

An 82-year-old man with a medical history of dysphagia underwent upper gastrointestinal endoscopy revealing a 6 cm long Barrett’s mucosa. Microscopic examination of the biopsy revealed specialized columnar cell metaplasia, consistent with Barrett’s esophagus. Lamina propria showed extensive inflammation with numerous monomorphic cells with eccentric nuclei and abundant eosinophilic ground-glass-like cytoplasm (Figure 1). Immunohistochemistry revealed positive staining for CD79a and CD138, confirming the plasma cell phenotype of these cells. These cells were polyclonal and immunoreactive for both kappa and lambda light chains (Figure 2). Cytokeratin AE1/AE3 was negative. The Barrett’s mucosa was negative for dysplasia.

DISCUSSION

First described by a Scottish physician Russell, the eponymously named “Russell bodies” are eosinophilic, large, immunoglobulin-containing inclusions that are commonly found within the cytoplasm of plasma cells.¹ Such plasma cells filled with RBs have also been called Mott cells.² Russell body gastritis (RBG) or gastroenteritis is a form of chronic gastrointestinal mucosal inflammation containing plasma cells with prominent intracytoplasmic RBs. It is believed that RBs are the result of cellular response to overstimulation of plasma cells in chronic inflammation, which results in condensed immunoglobulin in dilated endoplasmic reticulum cisternae.²³ The first case of RBG was described by Tazawa and Tsutsumi in 1998, which was associated with Helicobacter pylori infection.⁴ Since then, several cases of RBG and rare cases of RB duodenitis have been reported.⁵ The first case of RBs with Barrett’s esophagus was described by Rubio in 2005, and it was termed RB esophagitis.⁶ Bhaijee et al reported the second case of RBs associated with Barrett’s esophagus, which expanded the classic description of RBG and enteritis to esophagitis.⁷ The pathogenesis of RBG still remains unknown. An association with H. pylori infection has been suggested.²⁸ It is possible that the chronic infection with H. pylori may stimulate plasma-cell hyperactivation and subsequently lead to hyperproduction of immunoglobulins with
numerous RB formation. The disappearance of RBs after the treatment of *H. pylori* supports such a hypothesis. However, the finding of RBs in the absence of *H. pylori* is not clearly understood. The current case presents a unique situation in which RBs were observed in association with Barrett’s esophagus. A biopsy from the gastric antrum was negative for *H. pylori*. There is clearly no etiologic relationship between Barrett’s esophagus and *H. pylori* infection. Similarly, it is quite reasonable to infer that *H. pylori* infection is unlikely to play an etiologic role in the occurrence of RBs in the setting of Barrett’s esophagus. It has been suggested previously in the literature that immunocompromised status can predispose to the development of RBG. However, the current case was not known to have any associated immunocompromised condition. On the other hand, a chronic inflammatory state appears to be a common setting between both the presence of RBs and intestinal metaplasia.

Chronic inflammation and injury are known to result in mucosal changes such as intestinal metaplasia and gastric mucosal atrophy, among others. It is plausible that plasma cells packed full of immunoglobulin-containing endoplasmic reticulum might have an inflammatory backdrop that can explain both Barrett’s esophagus and the occurrence of RBs. However, this can simply be an incidental association and cannot be absolutely ruled out.

Differential diagnosis remains challenging because clinically and microscopically it can be confused with a neoplastic process. The possibility of hematological malignancy, including plasmacytoma and mucosa-associated lymphoid tissue lymphoma, should be ruled out. Signet ring cell carcinoma is another important diagnostic consideration, which can be ruled out by the absence of nuclear atypia, cytomorphic characteristics, and lack cytokeratin expression. The periodic acid-Schiff reaction can help identify RBs by conferring a dense, glassy stain to intracytoplasmic immunoglobulins. Plasma cell markers, such as CD138 and CD79a, are helpful, and coexpression of kappa and lambda light chain will demonstrate the polyclonal nature of the plasma cell infiltrate. Associated gastric carcinoma and infectious agents, such as *Helicobacter* and *Candida*, may alter patient management and clinical outcome and, therefore, should be excluded.

The treatment of RBs associated with Barrett’s esophagus is not well defined. Previous studies on RBG had suggested treating *H. pylori* when the organism was found to be associated. For RB Barrett’s esophagus, where *H. pylori* is an unlikely association, treatment should be aimed at managing Barrett’s esophagus, per recommended guidelines.

We present this case to emphasize the importance of recognizing this unusual entity. It also represents a potential diagnostic pitfall because the distended plasma cells may be mistaken for signet ring cells of gastric adenocarcinoma or low-grade lymphoma. Hence, an awareness of this entity is important to avoid diagnostic confusion.

**DISCLOSURES**

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