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CASE REPORT | ESOPHAGUS

Diffuse Villous Tumor Arising in Barrett's Esophagus Presenting With Aspiration Pneumonitis

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ABSTRACT

Barrett's esophagus (BE) is associated with an increased incidence of adenocarcinoma. The dysplasia in BE that precedes cancer is usually endoscopically flat. Unlike in the colon, polypoid dysplastic lesions superimposed on BE are uncommon. Furthermore, villous tumors of the esophagus are rare, and few cases have been reported in the literature. We report an 85-year-old man who was found to have a circumferential villiform-appearing esophageal tumor with mucus secretions with recurrent bronchopulmonary aspiration. A diffuse villiform tumor of the entire esophagus with mucin production is rare and, to our knowledge, has not been previously described in the literature.

INTRODUCTION

Barrett's esophagus (BE) is associated with an increased incidence of adenocarcinoma, occurring via the metaplasia-dysplasia-carcinoma sequence.[1,2] Dysplasia in BE typically occurs as flat endoscopically undetectable lesions. The pathological features and clinical progression of flat dysplasia have been studied extensively. Dysplastic polypoid lesions within the esophagus, also described as adenomas due to their resemblance to colonic adenomas, are quite rare, and their clinical features have not been elucidated. Multiple dysplastic polypoid lesions in BE are very uncommon, with only 2 cases being described previously.[3–5] Furthermore, a villiform tumor involving most of the esophagus has not been reported. We report an 85-year-old man with a long circumferential villiform-appearing esophageal tumor with excessive mucin production associated with BE.

CASE REPORT

An 85-year-old Hispanic man was admitted for an acute symptomatic deep vein thrombosis of the left lower extremity. While in the hospital, he was noted to have a cough with copious amounts of phlegm; this productive cough continued even after he was kept nil per os. His medical history included coronary artery disease, dyslipidemia, hypertension, and gout. He had no history of gastroesophageal reflux disease. Chest radiography revealed nodular infiltrates in the left lung base. Chest computed tomography with contrast revealed bilateral multifocal lobulated opacities. There was also a markedly distended esophagus with a large amount of fluid and what appeared to be food debris within the esophagus (Figure 1). Upper endoscopy with endoscopic ultrasonography revealed a long circumferential villiform-appearing esophageal tumor with copious mucoid secretions. The tumor, extending 20 cm from the incisors to the cardia (~30 cm in length), was not causing esophageal obstruction (Figure 2). The mucosa proximal to the tumor appeared normal endoscopically. An endoscopic string sign (≥1 cm string formed in fluid, which lasted for ≥1 second) was noted, suggesting a mucinous type tumor.[6] With endoscopic ultrasonography, the tumor appeared to have a fern-like appearance and involved the mucosa without involvement of the submucosa or muscularis propria. There were no periesophageal lymph nodes seen (Figure 3).

Biopsies of the esophagus proximal to the tumor showed extensive focal low-grade dysplasia. The tumor itself revealed extensive low-grade dysplasia with villous architecture and focal high-grade dysplasia and intramucosal adenocarcinoma (Figure 4). Barrett's

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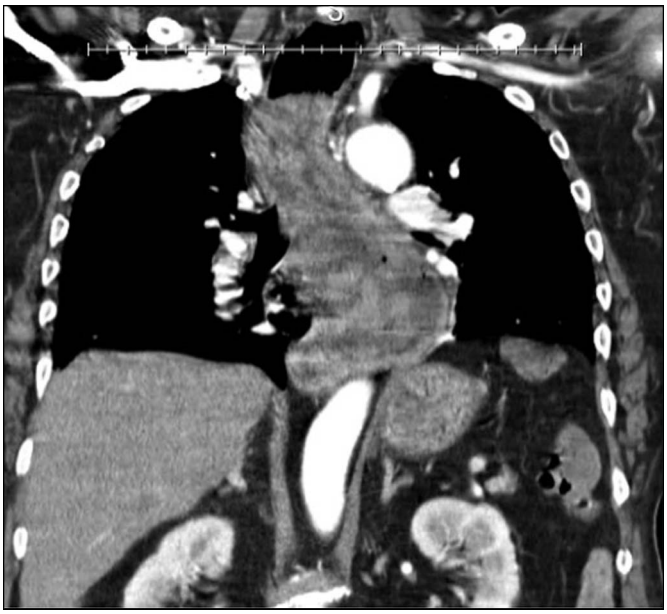

Figure 1. Chest computed tomography showing distended esophagus containing fluid and what appeared to be food material.

metaplasia was identified as well within the tumor. The stomach and duodenum were normal. Esophageal stenting was attempted twice with 18 × 100 mm and 23 × 155 mm Wallflex fully covered stents (Boston Scientific, Marlborough, MA), but these stents migrated shortly after deployment due to the profound dilation of the proximal esophagus and very soft consistency of the tumor. The patient was not considered to be a candidate for radiation therapy, and the tumor was too large for radiofrequency ablation; therefore, a decision was made to proceed with tumor debulking to decrease mucin hypersecretion. He later underwent endoscopic debulking along with endoscopic mucosal resection (EMR). Even with extensive debulking, the patient continued to produce copious amounts of phlegm. He was offered esophagectomy, but elected to proceed with hospice care.

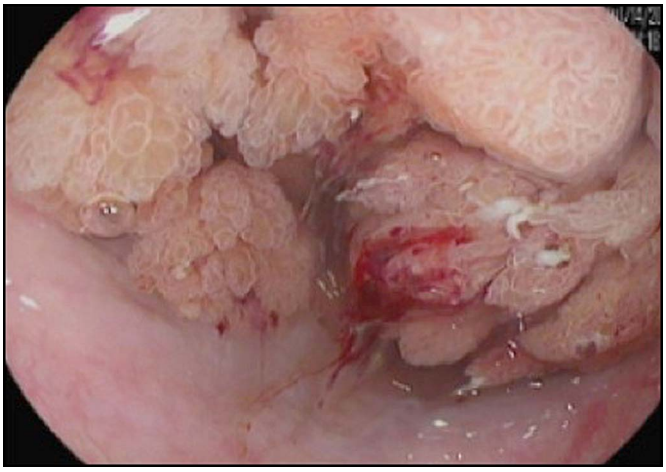

Figure 2. Endoscopy demonstrating a large villiform tumor of the esophagus.

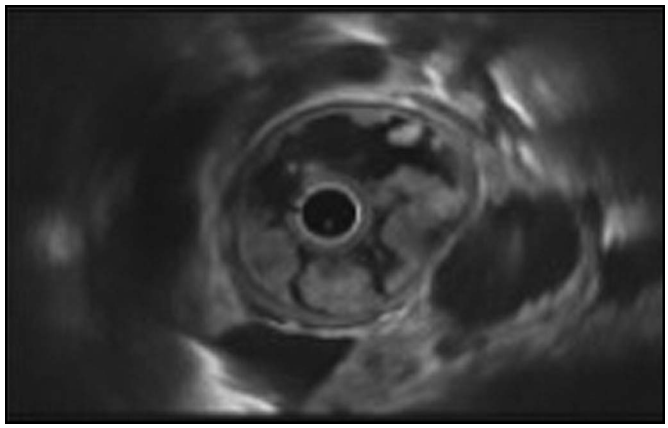

Figure 3. Endoscopic ultrasonography of the large circumferential tumor of the esophagus.

DISCUSSION

Diffuse villiform tumor with mucin production of nearly the entire esophagus has not been previously described in the literature. Dysplastic lesions in BE are mostly flat and are not detectable with endoscopic visualization. Polypoid lesions of the esophagus are rare and most are solitary.[3] A review of the literature by Ahlawat and Ozdemirli, revealed a total of 21 cases that described patients with polypoid dysplasia along with the corresponding clinical characteristics and pathological results.[3] None of these cases showed a diffuse circumferential villiform-appearing esophageal tumor. Most (76%) of the patients were men with an average age of 59 years. Most (88%) of the lesions were located in either the distal esophagus or at the gastroesophageal junction. The length of the lesions ranged from 0.2 to 10 cm, with an average size of 2.3 cm. In our patient, the length of the tumor exceeded 20 cm. All the cases were associated with BE, and 11 of the 21 polyps (52%) had foci of adenocarcinoma.

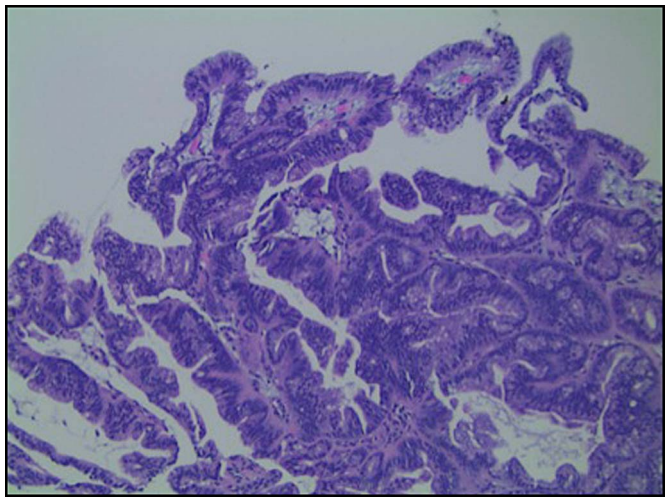

Figure 4. Hematoxylin and eosin stain of esophageal tumor showing extensive low-grade dysplasia with villous architecture and focal high-grade dysplasia and intramucosal adenocarcinoma.

Thurberg et al, compared the immunohistochemical and molecular characteristics of 5 patients with polypoid dysplasia to 5 patients with flat dysplasia used as controls.[7] All 5 patients with polypoid dysplasia had well-defined sessile or pedunculated polypoid lesions seen during endoscopy. Histologically, all the polyps comprised intestinalized epithelium with low- and high-grade dysplasia. In addition, all of the polypoid cases had foci of adenocarcinoma. Immunohistochemistry staining showed that all polyps, as well as the flat dysplasia controls, were positive for surface MiB-1, suggesting increased cell proliferation, as well as positive p53 staining. These findings indicate that flat and polypoid dysplasia share a similar pathogenic pathway, although their visual characteristics during endoscopy are quite different.

Diffuse villiform esophageal tumor involving the nearly entire length of the esophagus has not been reported in the literature. An interesting feature of our case is signified by the mucin production by the tumor to the extent that it caused bronchopulmonary aspiration, which can be explained by both the size of the tumor and the possibility of a change in MUC gene expression. Patients with BE express MUC genes associated with normal gastric and intestinal epithelium.[8] MUC1 has been shown to be upregulated in patients with BE that progress from dysplasia to adenocarcinoma.[8,9] The cases of polypoid dysplasia within the esophagus seen in the review by Thurberg et al showed lesions that involved 1 portion of the esophagus and none involved the entire esophagus. An endoscopic string sign, most commonly used in the diagnosis of mucinous pancreatic cysts, suggested the presence of a mucin-producing tumor.[6] Interestingly, our patient had no symptoms of dysphagia, perhaps due to the soft nature of the tumor.

Options for the management of polypoid dysplasia of the esophagus include endoscopic approaches to remove the adenoma along with ablation of BE, endoscopic resection of BE, and surgical esophagectomy. Radiofrequency ablation with endoscopic resection has shown success in visible dysplasia and early cancer in BE.[10] EMR or endoscopic submucosal dissection allows for excision of potentially malignant tissue and to determine the extent of the tumor.[11] If the lesion is localized, EMR and endoscopic submucosal dissection are effective approaches for excision. Complete debulking of the tumor was not performed in our patient because of the high rate of stricture formation following EMR of circumferential lesions and the significant risk of bleeding or perforation due to the size of the tumor. Esophagectomy allows for complete removal of all neoplastic epithelium and any regional lymph nodes, but it is associated with considerable mortality and morbidity.

To our knowledge, diffuse villiform tumor of the esophagus in the setting of BE has not been previously reported. Our case exhibits the rare presenting complication of bronchopulmonary aspiration with pneumonitis from excessive mucin secretion and endoscopy showing the extent of the villiform tumor that involved almost the entire esophagus.

DISCLOSURES

Author contributions: All authors contributed equally to manuscript creation. V. Kaila is the article guarantor.

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Informed consent was obtained for this case report.

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REFERENCES

1. Haggitt RC. Barrett's esophagus, dysplasia, and adenocarcinoma. *Hum Pathol.* 1994;25(10):982–93.
2. Hameeteman W, Tytgat GN, Houthoff HJ, van den Tweel JG. Barrett's esophagus: Development of dysplasia and adenocarcinoma. *Gastroenterology.* 1989;96(5 pt 1):1249–56.
3. Ahlawat SK, Ozdemirli M. Polypoid dysplasia in Barrett's esophagus: Case report and qualitative systematic review of the literature. *Acta Gastroenterol Belg.* 2012;75(1):49–54.
4. Fang SY, Wu CY, Chen HC, Lin YC, Wang YY, Fang HY. Giant adenoma arising in the lower esophagus with adenocarcinoma. *Dig Dis Sci.* 2007;52(11):3181–3.
5. Wong W, Shek TH, Chan CK, Lai K. Gastrointestinal: Villous adenoma of the esophagus. *J Gastroenterol Hepatol.* 2004;19(10):1213.
6. Bick BL, Enders FT, Levy MJ, et al. The string sign for diagnosis of mucinous pancreatic cysts. *Endoscopy.* 2015;47(07):626–31.
7. Thurberg BL, Duray PH, Odze RD. Polypoid dysplasia in Barrett's esophagus: A clinicopathologic, immunohistochemical, and molecular study of five cases. *Hum Pathol.* 1999;30(7):745–52.
8. Arul GS, Moorghen M, Myerscough N, Alderson DA, Spicer RD, Corfield AP. Mucin gene expression in Barrett's oesophagus: An in situ hybridisation and immunohistochemical study. *Gut.* 2000;47(6):753–61.
9. Burjonrappa S, Reddimasu S, Nawaz Z, Gao X, Sharma P, Loggie B. Mucin expression profile in Barrett's, dysplasia, adenocarcinoma sequence in the esophagus. *Indian J Cancer.* 2007;44(1):1.
10. Phoa KN, Pouw RE, van Vilsteren FGI, et al. Remission of Barrett's esophagus with early neoplasia 5 years after radiofrequency ablation with endoscopic resection: A Netherlands cohort study. *Gastroenterology.* 2013;145(1):96–104.
11. Soetikno RM, Gotoda T, Nakanishi Y, Soehendra N. Endoscopic mucosal resection. *Gastrointest Endosc.* 2003;57(4):567–79.