

INTRODUCTION

Cancer of the appendix is an uncommon malignancy rarely diagnosed on colonoscopy. We present a case of incidentally discovered goblet cell adenocarcinoma (GCA) of the appendix and the subsequent management of the malignancy.

CASE PRESENTATION

- An 81-year-old male with past medical history of coronary artery disease, atrial fibrillation, and diverticulosis presented to a GI clinic for persistent epigastric pain and acid reflux.
- Symptoms had been worsening for several years and were refractory to medical therapy.
- He denied unintentional weight loss, nausea, dysphagia, or melena.
- He reported a family history of stomach cancer (grandmother) but denied family history of colorectal cancer.
- Additional history included nine colonoscopies in a span of 17 years for rectal bleeding secondary to hemorrhoids and diverticulosis. A single 2mm tubular adenoma was removed in 2016 and a 5-year follow-up had been advised.
- An upper endoscopy and colonoscopy were performed in early 2021. During the colonoscopy, the appendiceal orifice was noted to have a “heaped-up” appearance and was biopsied with a cold forceps (Figure A)
- Histology from the biopsies showed GCA of the appendix.
- CT scan of the abdomen and pelvis was obtained, showing a mildly enlarged appendix, no lymphadenopathy, and no evidence of metastasis.
- The patient underwent a laparoscopic right hemicolectomy 2 months later.
- Pathologic evaluation showed that carcinoma extended from the appendix to the mesoappendix and cecum with extensive lymphovascular and perineural invasion. (Figures B-E). The resection margins and 19 resected lymph nodes were negative for malignancy. Staging following surgery was pT4aN0 (IIB).
- The patient recovered from surgery uneventfully. Follow-up colonoscopy 1 year later revealed a healthy ileocolonic anastomosis and post-surgical changes with no signs of polyps or other masses (Figure B). Surveillance CT scan showed no active disease.

FIGURES

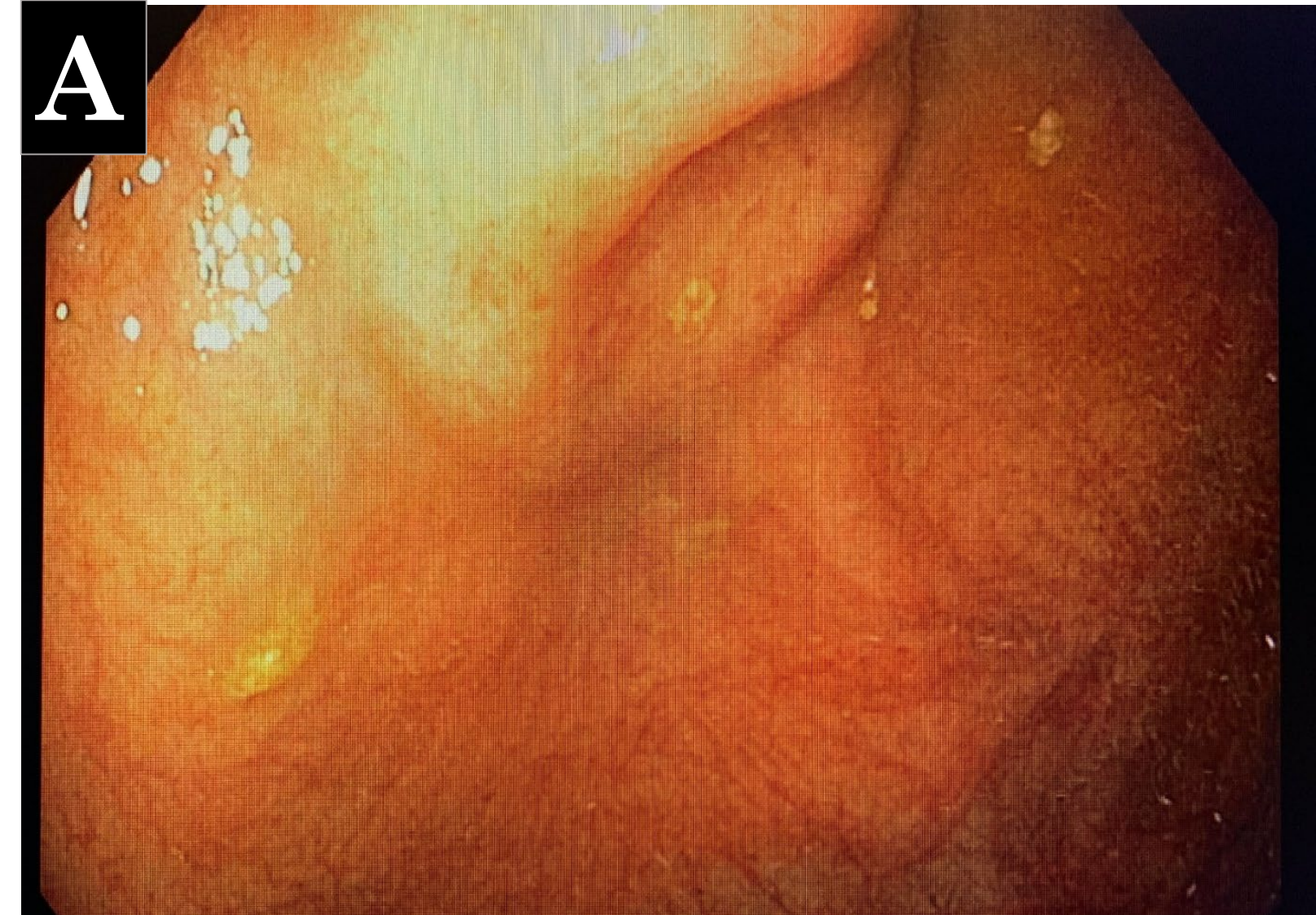


Figure A: Appendiceal orifice with “heaped-up” appearance

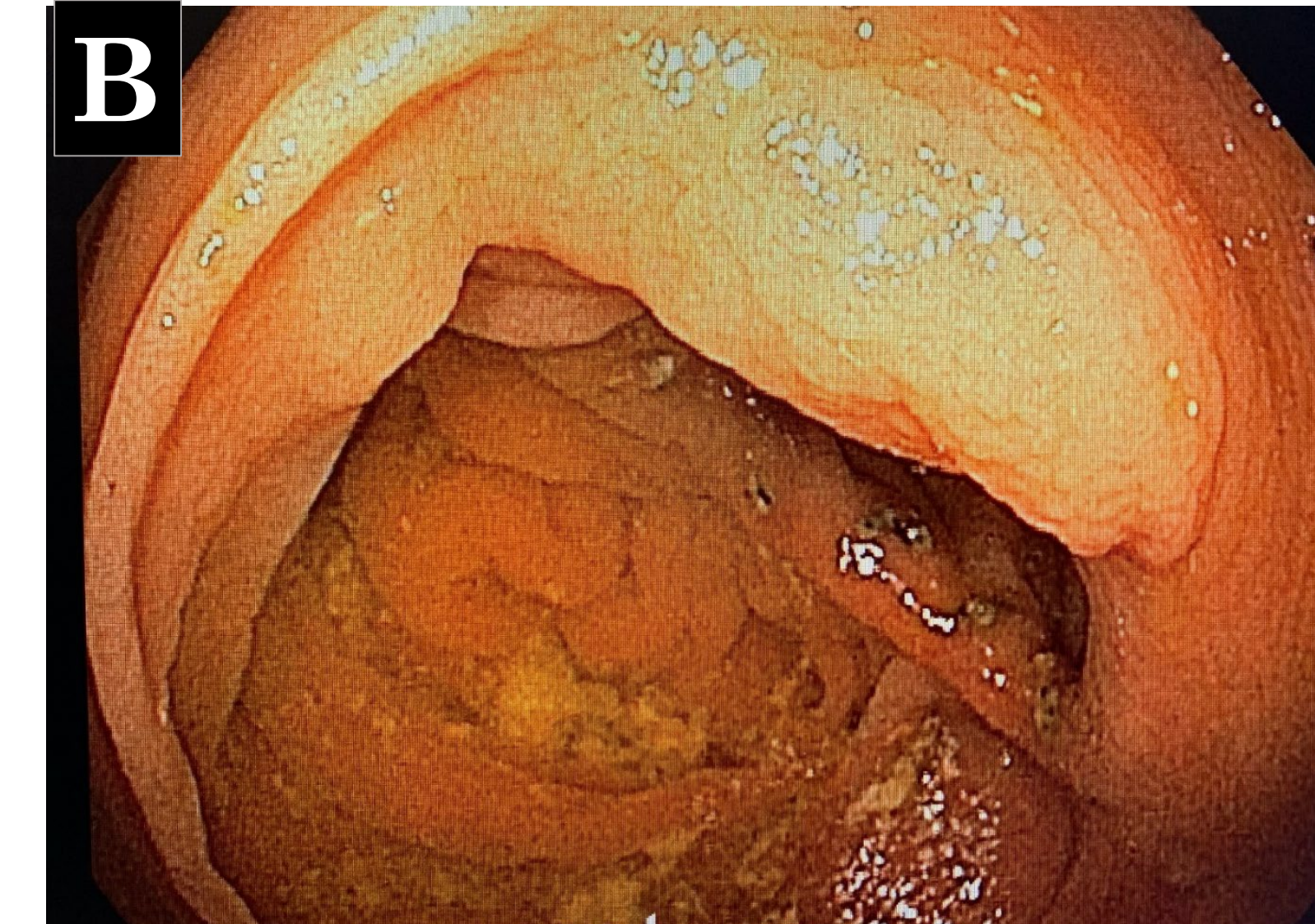


Figure B: Healthy ileocolonic anastomosis

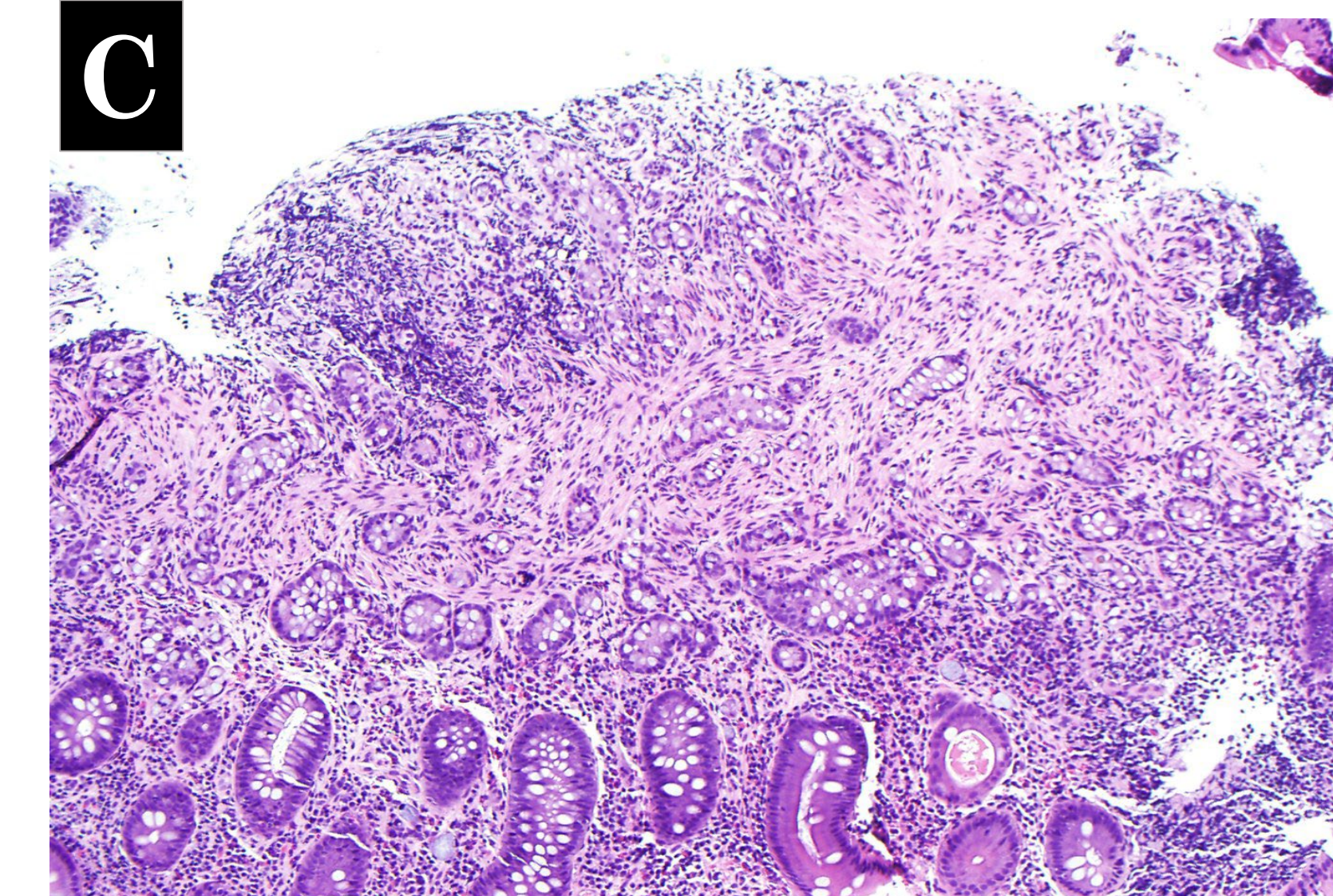


Figure C: Clusters of goblet-like mucinous cells with invasion into and through the lamina propria. 10X

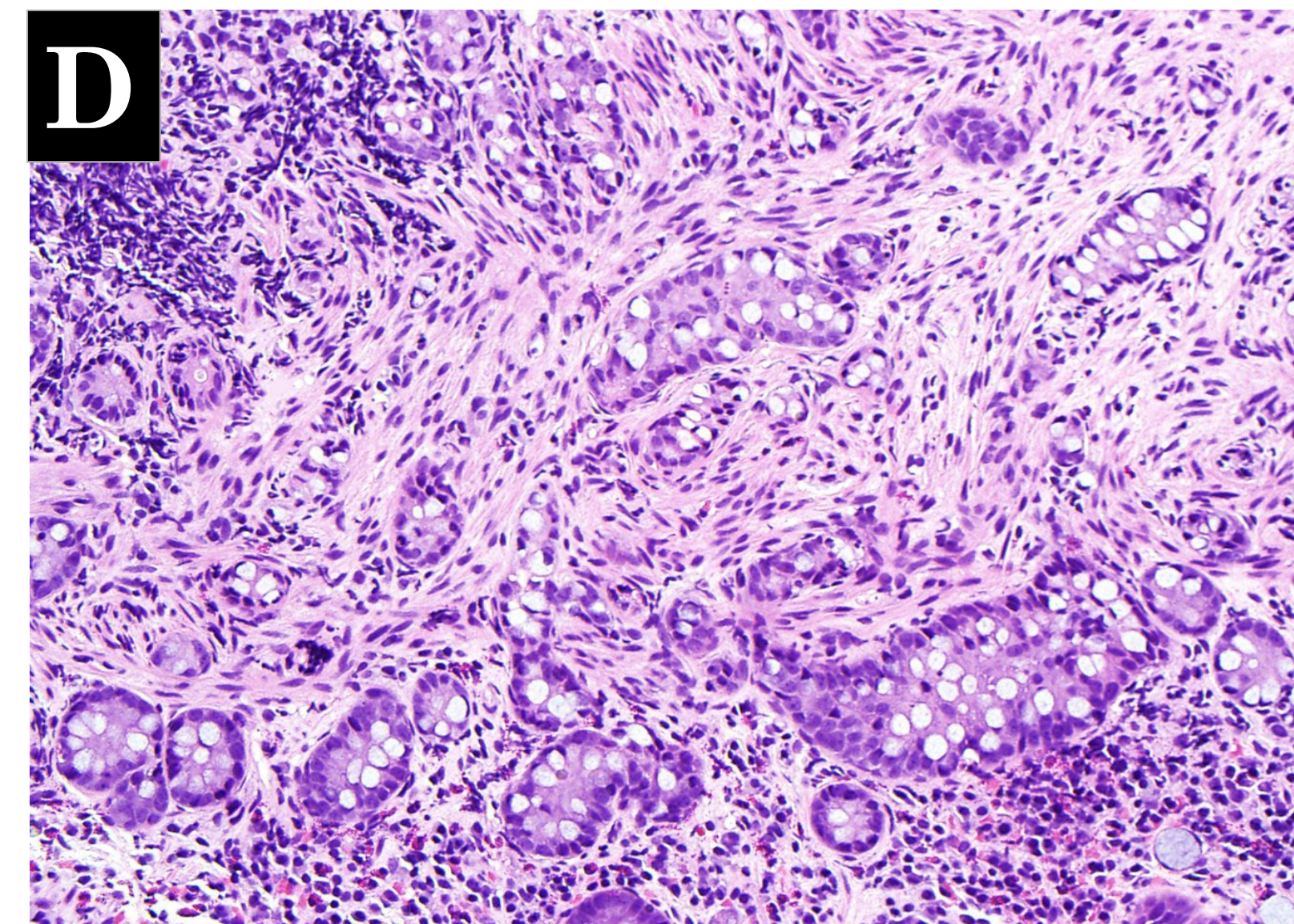


Figure D: High power view (20X) of cohesive groups of goblet-like mucinous cells. Nuclear atypia is mild and mitotic figures are inconspicuous.

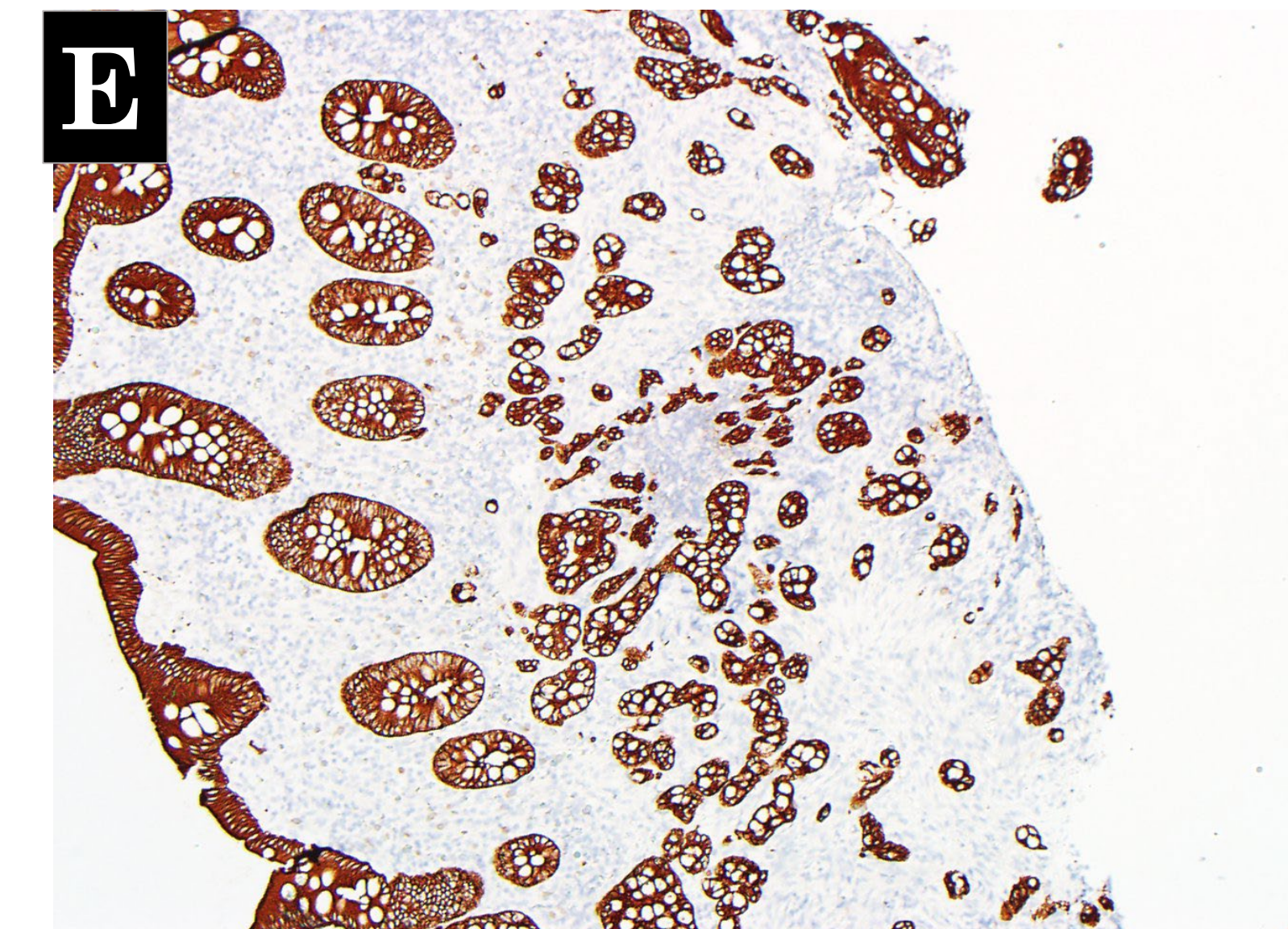


Figure E: Cytokeratin AE1/AE3 highlight the colonic mucosa and the infiltrative goblet-cell tumor. 10X

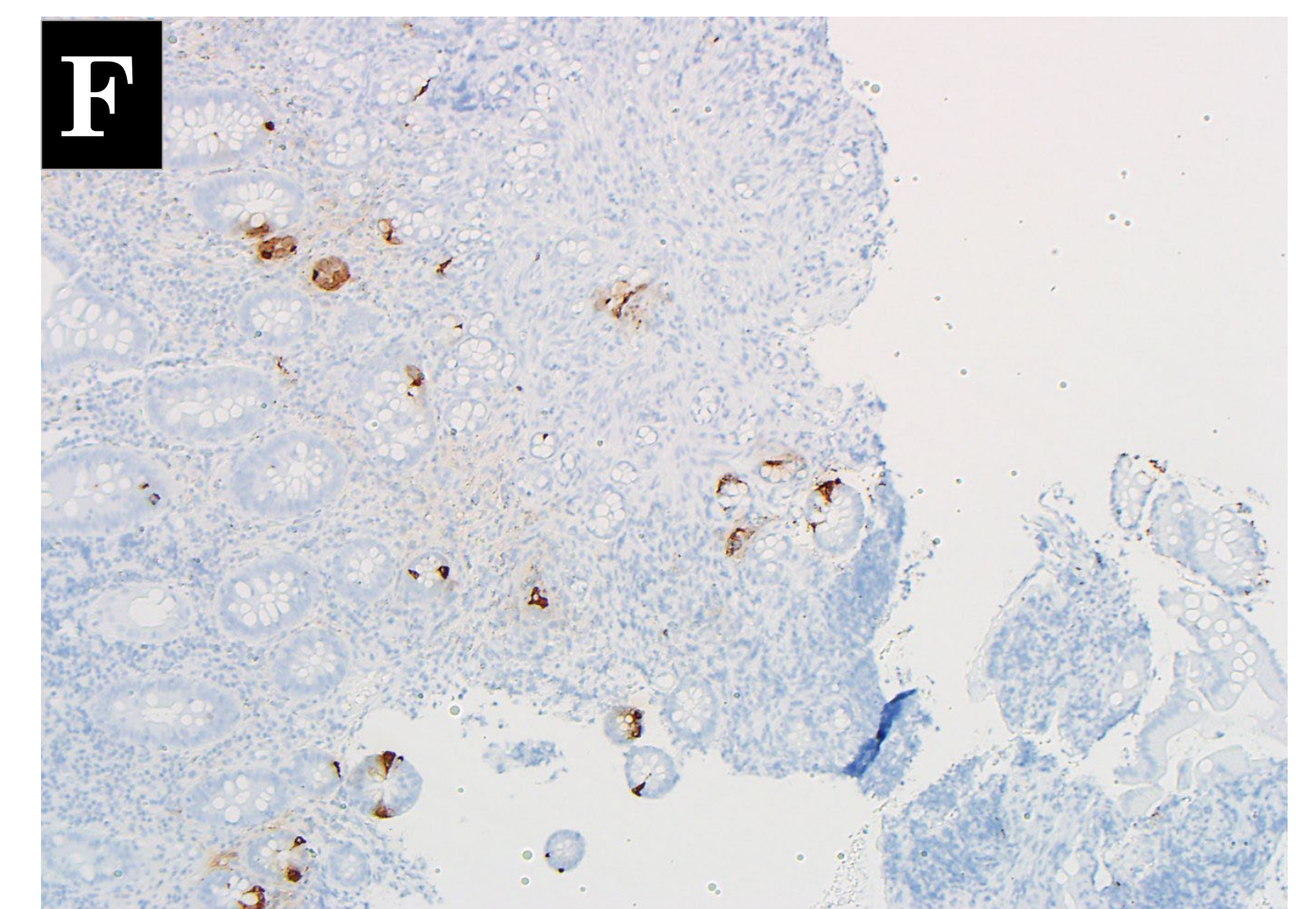


Figure F: Chromogranin highlights scattered endocrine cells. 10X

DISCUSSION

- Appendiceal cancer is categorized into two main types: epithelial and neuroendocrine.
- GCA is an aggressive subtype that histologically has features of both types.
- Incidence is rare, reported to be 0.05 per 100,000 per year.
- While there are no established risk factors for GCA, some cases have shown an association with schistosomiasis and a proclivity for the Caucasian race.
- This tumor is usually an incidental finding following an appendectomy for an acute appendicitis and is uncommonly found via colonoscopy.
- The appendiceal orifice is a key landmark that should be identified on all colonoscopies (unless the patient does not have a cecum) to ensure completeness of the procedure, with low threshold to biopsy abnormal-appearing tissue.
- While GCAs are rare and evidence-based guidelines are not available, current management is similar to that of colon cancer with a right hemicolectomy for localized disease with possible adjuvant chemotherapy based on the surgical pathology, followed by post-treatment surveillance.
- Our hope is that this case highlights the rarity of appendiceal GCA and the need for evidence-based therapy guidelines.