

## Case Report

# Polypoid adenomyoma of the gastric fundus: an unusual endoscopic and pathologic presentation

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**Abstract:** Gastric adenomyoma (GA) is a rare benign lesion, most frequently found in the pyloric/antral region of the stomach. It consists of a mixture of smooth muscle and glandular structures, occasionally cystically dilated, lined by tall columnar epithelium, Brunner's glands, and sometimes pancreatic parenchyma. In this report, we describe an unusual case of adenomyoma of the fundus of the stomach, which presented as a polyp on endoscopy, and was found to be a polypoid, cystic lesion pathologically; both of these features are rare for a gastric adenomyoma.

**Keywords:** Gastric adenomyoma, polypoid, fundus

### Introduction

Gastric adenomyoma (GA) is a rare benign lesion, most frequently found in the pyloric/antral region of the stomach. The first case was reported in 1903 by Magnus-Alsleben [1]. It is considered hamartomatous in nature and consists of a mixture of smooth muscle and glandular structures, occasionally cystically dilated, lined by tall columnar epithelium, Brunner's glands, and sometimes pancreatic parenchyma [2]. GA occurs mainly in adults, but newborns and children are not spared, with ages ranging from 1 week old to 81 years old. Rarely malignant transformation has also been reported [3, 4]. In this report, we describe an unusual case of adenomyoma of the fundus of the stomach, which presented as a polyp on endoscopy, and was found to be a polypoid, cystic lesion pathologically; both of these features are rare for a gastric adenomyoma.

### Case report

A 59-year old woman presented with retrosternal pain and a physical examination did not reveal significant abnormalities. All laboratory tests were within normal range and she underwent gastroesophageal endoscopy for heartburn.

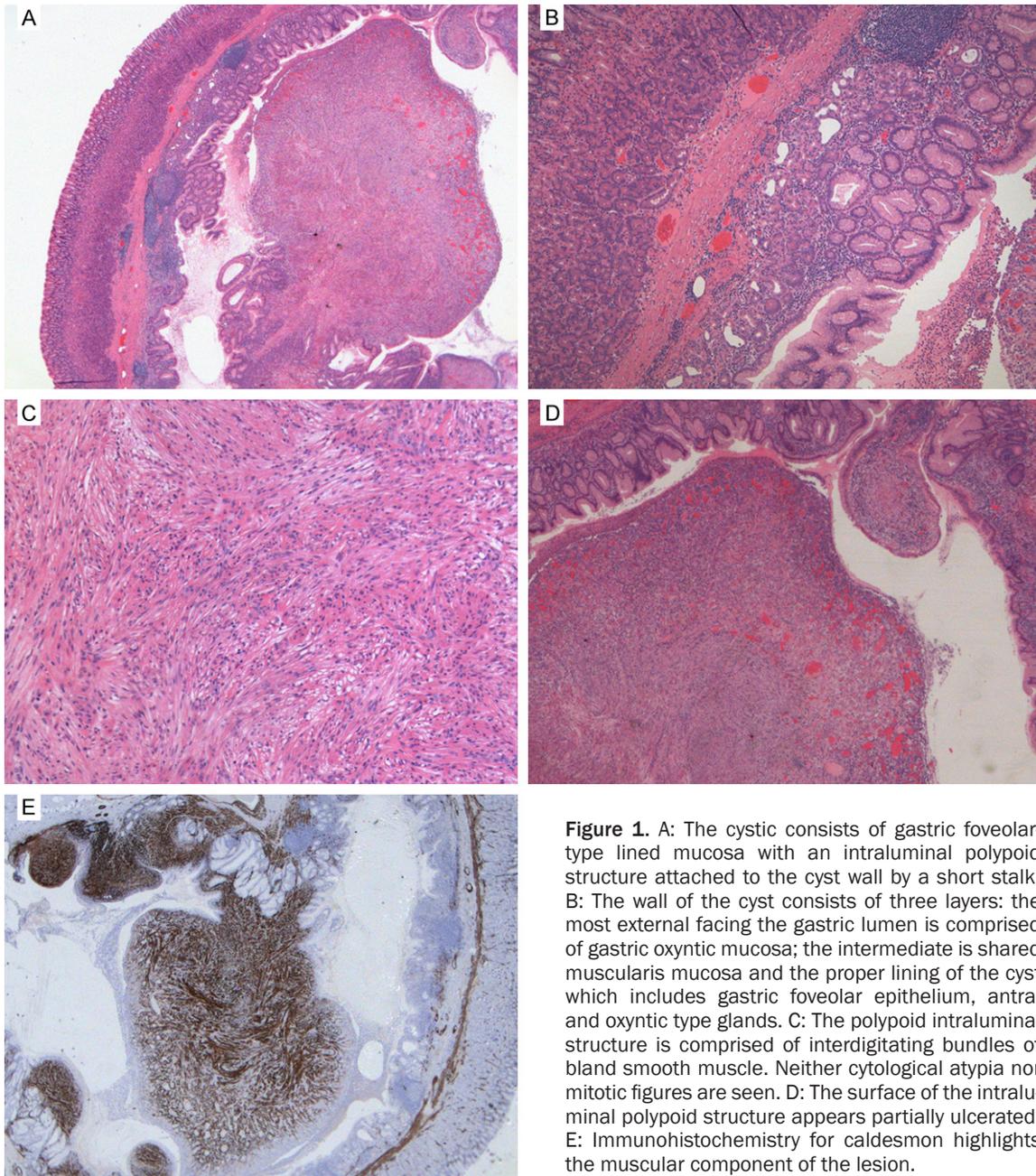
At endoscopy, the esophagus and duodenum were unremarkable. The stomach showed a small amount of bile but on retroflexed view, a large submucosal, circular mass covered by normal gastric mucosa was seen in the gastric fundus near the gastroesophageal junction. The rest of the stomach was unremarkable. Biopsies of the fundic lesion and of the antrum were taken in an attempt to characterize the lesion and to exclude the presence of *H. pylori* organisms. The biopsy showed only unremarkable oxyntic mucosa and *H. pylori* organisms were absent.

The patient underwent a CT scan of the abdomen and pelvis, which failed to identify the gastric lesion. No other significant abnormalities or lymphadenopathy were noted. The only unrelated finding was the presence of an aneurysm of the distal aorta measuring 2.7 cm.

At the follow-up visit, it was decided to remove the fundic lesion by endoscopic excision. During the procedure, mucinous/clear fluid exuded from the cystic lesion.

Macroscopic examination showed a mixed cystic and solid submucosal lesion, which consisted of a piece of polypoid tan-brown tissue measuring 2.0 × 1.4 × 1.5 cm. The lesion was

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**Figure 1.** A: The cystic consists of gastric foveolar-type lined mucosa with an intraluminal polypoid structure attached to the cyst wall by a short stalk. B: The wall of the cyst consists of three layers: the most external facing the gastric lumen is comprised of gastric oxyntic mucosa; the intermediate is shared muscularis mucosa and the proper lining of the cyst which includes gastric foveolar epithelium, antral and oxyntic type glands. C: The polypoid intraluminal structure is comprised of interdigitating bundles of bland smooth muscle. Neither cytological atypia nor mitotic figures are seen. D: The surface of the intraluminal polypoid structure appears partially ulcerated. E: Immunohistochemistry for caldesmon highlights the muscular component of the lesion.

entirely submitted for histological evaluation. Microscopically, the surface was lined by unremarkable gastric oxyntic mucosa which was embedded within the gastric wall forming an intramural submucosal cyst (**Figure 1A**). The cyst wall was also lined by gastric foveolar, antral-type epithelium with oxyntic glands, resting on a loose connective tissue, resembling lamina propria, rich in plasma cells, lymphocytes, lymphoid aggregates surrounded by smooth muscle, which was in continuity with

the muscularis mucosae of the overlying surface oxyntic mucosa (**Figure 1B**). The epithelium lining the cyst showed focal reactive changes and was negative for dysplasia. The lumen of the cyst was occupied by a protuberant, polypoid structure connected with the cyst wall by a short stalk (**Figure 1A**). The polypoid intraluminal structure consisted of broad interdigitating bundles of bland smooth spindle cells in continuity with the smooth muscle of the muscularis mucosae (**Figure 1C**). No mitotic figures

or cytological atypia were seen. The surface of the polypoid structure was focally lined by antral-type mucosa with areas of granulation tissue and fibrinopurulent exudate, consisting with ulceration (**Figure 1D**). Neither ectopic pancreatic tissue, nor Brunner's glands were identified within the bundles of smooth muscle or in the wall of the cyst. Immunohistochemistry showed the spindle cells to be immunoreactive for desmin, SMA and caldesmon (**Figure 1E**). Immunohistochemistry showed the spindle cells to be negative for CD117, DOG-1 and S-100. CD117 highlighted numerous interstitial cells of Cajal cells intermixed with the smooth muscles fibres. The histopathological findings were in keeping with a benign, adenomyoma of the gastric fundus.

### Discussion

GAs are rare benign hamartomatous lesions, which consist of a variable mixture of bundles of smooth muscle and epithelial elements in the submucosa. The epithelial element may include gastric foveolar glands with/without goblet cells, Brunner's glands and pancreatic elements. Adenomyomas can be found in the stomach, duodenum, jejunum, ampulla and gallbladder where they present as solid mural nodules. Presentation as a cystic polypoid lesion is rare. The origin of the adenomyoma is controversial. Gastrointestinal epithelium originates from endoderm, whereas muscle, connective tissue and peritoneal elements derive from the mesoderm. Abnormal interactions between endodermal and mesodermal elements may result in the formation of adenomyomas. Primordial epithelial buds may develop and variably differentiate, resulting in the spectrum of adenomyoma, including aberrant pancreas and Brunner's glands [5, 6]. The smooth muscle bundles may arise from embryonic muscle or may represent a proliferation of normal muscle induced by the misplaced epithelium [7]. These lesions are classified according to which cell type predominate; aberrant pancreas is the most common variant [5].

GAs of the stomach are most frequently found in the distal stomach, antrum and pylorus. They may occur at any age and there is a slight predilection for women [7]. The presentation of these lesions is quite variable and depends on the site and size of the lesions. They may be asymptomatic or cause obstruction, bleeding

with melena [8], and intussusception [9] or cause non-specific symptoms such as epigastric pain and vomiting. Asymptomatic lesions can be found incidentally during endoscopy or imaging for other reasons.

Given the submucosal localization of these lesions, endoscopic biopsies have a very low yield [10] and surgical resection is usually necessary to confirm the diagnosis. Histologically, the diagnosis generally is straightforward and based on the presence of bundles of smooth muscle intimately associated with epithelial elements. Invasive gastric adenocarcinoma associated with GAs has been reported [3, 11]. The differential diagnosis depends on the age of the patient and includes Brunner's gland hamartomas, which generally occurs in the first part of the duodenum, myofibromas of the pylorus composed by myoid cells with thin-walled blood vessels. When epithelial elements are lacking or scarce, gastrointestinal stromal tumors (GISTs) and leiomyomas should be considered [9]. Another condition which may enter in the differential diagnosis is gastritis cystica profunda. This condition is characterized by misplacement of hyperplastic foveolar glands, cystically dilated into the gastric submucosal layer. The lack of smooth muscle bundles around the cysts helps to distinguish gastritis cystica profunda from GAs.

To the best of our knowledge this is the first case in the literature of a GA of the gastric fundus (in close proximity of the gastroesophageal junction). Our lesion was cystic and contained an unusual combination of epithelial elements and smooth muscle: antral mucosa with oxyntic glands embedded in a loose connective tissue resembling the lamina propria and associated smooth muscle from the muscularis mucosae. These morphological features are unusual for GAs which typically consists of glands embedded within muscle in the wall. They are rarely cystic, and this "cyst within a cyst" appearance is even more unique.

Yoon et al reported a case of GA of the gastric body [7]; and their lesion shows similar features to the GA described herein. Both were submucosal with mixed cystic and solid components, filled with mucinous/clear liquid. However, histologically, Yoon et al describe broad interdigitating bundles of smooth muscle present between the ducts, which were occasional-

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ly dilated and lined by tall columnar cells [7]. In our case, the cyst was lined by gastric mucosa with antral and oxyntic type glands while the bundles of smooth muscle were mainly protruding into the lumen of the cyst and was in continuity with the smooth muscle of the muscularis mucosae of the overlying oxyntic gastric mucosa.

In conclusion, although GAs are commonly located in the distal stomach, we describe a rare, incidental case of GA of the gastric fundus, which appeared as a polyp on endoscopy, and was a polypoid cystic lesion pathologically. Histologically, it was typified by gastric type mucosa with oxyntic glands, devoid of both Brunner's glands and pancreatic elements, both of which are rare for a GA.

### Disclosure of conflict of interest

None.

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