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# Gastrointestinal stromal tumor of the vermiform appendix mimicking Meckel's diverticulum: Case report with literature review



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## ABSTRACT

**INTRODUCTION:** Gastrointestinal stromal tumors (GISTs) of the appendix are extremely rare. To date, only 15 cases have been reported in the English literature. Here, we present a new case of an appendiceal GIST with appendicitis.

**PRESENTATION OF CASE:** A 68-year-old man who complained of right lower abdominal tenderness underwent surgery for a cystic mass mimicking Meckel's diverticulum. Laparoscopy revealed a mass protruding from the proximal appendix with distal appendicitis. Complete resection with adequate margins was performed. Histology showed a spindle cell GIST without mitotic activity as well as a strong expression of CD117 and CD34.

**CONCLUSION:** Primary appendiceal GIST occur at a very low rate and their symptoms are nonspecific. Accordingly, rare tumors of appendix including GISTs should be considered in the differential diagnosis of atypical symptoms or image findings.

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## 1. Introduction

Gastrointestinal stromal tumors (GISTs) are rare; they arise from mesenchymal tissues in the gastrointestinal tract. GISTs occur most commonly in the stomach (60%) and the small bowel (30%) [1]. Primary appendiceal GISTs are extremely rare, with only 15 reported cases in the English literature to date [2–12]. We present the clinicopathologic features of a new case of appendiceal GIST mimicking Meckel's diverticulum and discuss treatment proposals based on currently available literature.

## 2. Case presentation

### 2.1. Patient

A 68-year-old man presenting with right lower quadrant abdominal pain was referred to our hospital. His past medical and surgery history was negative. On physical examination, tenderness and rebound tenderness without a palpable mass were present in the right lower quadrant. Laboratory findings were: mild leukocytosis (11,600/ $\mu$ L) and neutrophilia (80%). Before five months ago, He presented to the internal medicine department of our hospital with a chief complaint of atypical abdominal pain. Computed

tomography revealed a cystic lesion with a solid component in the right lower quadrant of the abdomen. Surgical exploration was recommended; however, he refused. On his return visit, computed tomography showed a well-demarcated and enhanced tumor with a cystic component; it appeared to originate from the terminal ileum and measured approximately 2.5 × 6.0 × 2.5 cm (Fig. 1). Compared to the prior exam, the lesion slightly increased and inflammation was present. The clinical impression was complicated appendicitis or an inflammatory Meckel's diverticulum. Under laparoscopic exploration (Fig. 2), a solid mass with a cystic component protruded from the proximal appendix and the distal appendix was secondarily engorged. The lesion was located 1 cm from the base of the cecum with a safety margin. The mass was removed by a simple laparoscopic appendectomy. Postoperatively, he recovered and was discharged without any complications.

### 2.2. Pathologic findings

The gross specimen was a whitish-gray polypoid mass involving the muscle layer and bulging outward from the proximal appendix; it measured 3.0 × 2.5 × 2.5 cm. The distal portion the solid mass contain a cystic area measuring 2.3 × 2.3 cm. Appendiceal muscularis propria was present at the root of the tumor; it grossly formed a polypoid outward extension from the proximal appendix (Fig. 3). Histologically, the tumor was composed of spindle cells characteristic of GISTs, with irregular fascicles and variably sclerosed stroma (Fig. 4a). There was no nuclear atypia and mitotic activity was

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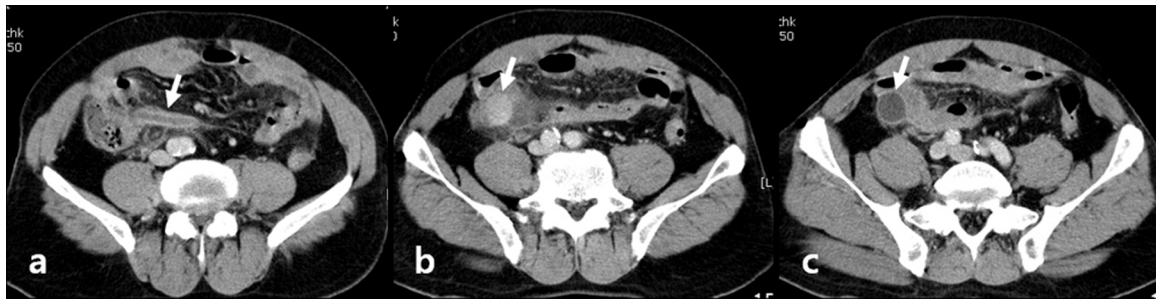


Fig. 1. Preoperative computed tomography. (a) Engorged inflammatory appendix (b) well-demarcated, enhanced mass (c) cystic lesion.

**Table 1**  
Clinical and histological characteristics of reported GISTs.

Case	Age/sex	Location	Presentation	/50 hpf	Reference
1	64/M	Tip	Autopsy	<1	[2]
2	56/M	Proximal	Appendicitis-like symptoms	<1	[2]
3	59/M	Middle	Gastric GIST resection	<1	[2]
4	72/M	Proximal	Acute appendicitis	<1	[2]
5	66/F	Middle	Appendicitis-like symptoms	<	[3]
6	56/M	Middle	Hematochezia	NR	[4]
7	65/F	NR	Recurrent lymphoma	<1	[5]
8	86/F	NR	Diverticulosis, incidentally found	<1	[5]
9	78/F	Proximal	Acute appendicitis	<1	[5]
10	72/M	Tip	Urocyctectomy, incidentally found	<1	[5]
11	57/M	Tip	Acute appendicitis	9	[7]
12	67/M	Middle	Appendicitis-like symptoms	<5	[9]
13	75/M	Middle	Acute appendicitis	NR	[10]
14	7/M	Proximal	Appendicitis-like symptoms	NR	[11]
15	88/F	Tip	Diverticulosis, incidentally found	<1	[12]
16	68/M	Proximal	Appendicitis-like symptoms	<1	Present case

NR: not reported; /50 hpf: number of mitoses per 50 high power fields.

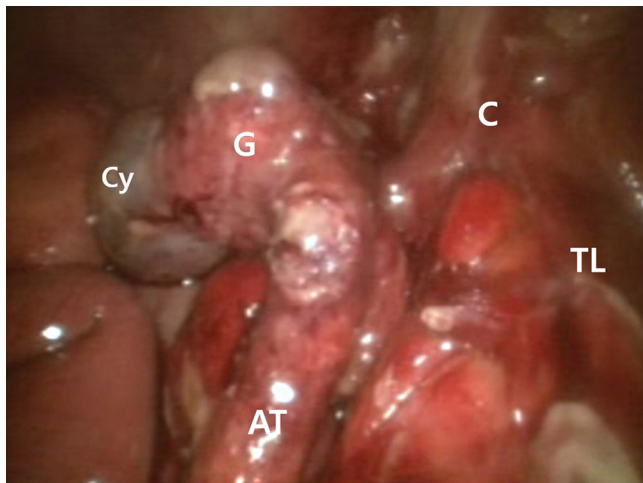


Fig. 2. Operative findings. C: cecum; TL: terminal ileum; G: solid component of GIST; Cy: cystic component of GIST; AT: appendiceal tip.

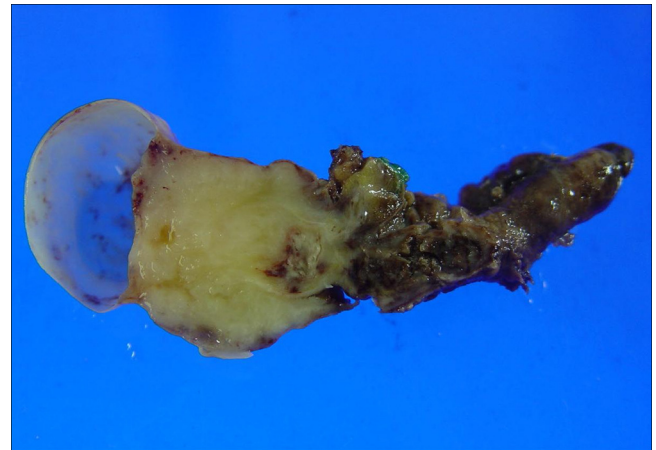


Fig. 3. Cross-sectioned gross findings: a pedunculated tumor attached to the proximal appendiceal muscle wall.

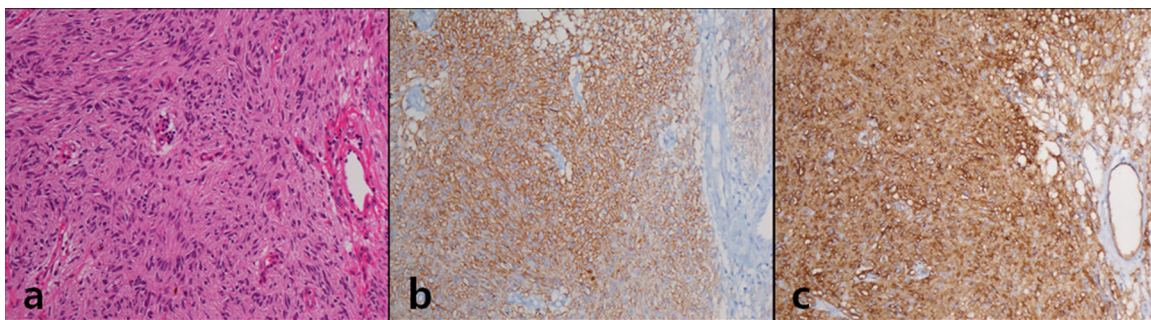
absent (<1/50/high power field). Immunohistochemistry showed strong expression of CD117 (Fig. 4b) and CD34 (Fig. 4c) in the tumor cells; however, no reactivity was seen for SMA, desmin, and protein S100. The risk classification according to Fletcher et al. [13] was low risk.

2.3. Postoperative course

Because of the low risk, the patient did not receive additional treatment. At the last follow-up, 35 months postoperatively, he was well without evidence of tumor recurrence.

3. Discussion

GISTs are relatively rare tumors which develop in the gastrointestinal tract, mesentery, or omentum; however, they are the most common mesenchymal neoplasms of the gastrointestinal tract. They differentiate similar to the interstitial cells of Cajal [1]. GISTs have a variable histologic spectrum including spindle, epithelioid, and pleomorphic cell morphology [1]. The differential diagnosis of GISTs from other stromal tumors is often difficult with a routine histologic examination. However, they can be distinguished from other stromal tumors such as leiomyomas, leiomyosarcomas, and neurofibromas by immunohistochemistry. Their diagnostic feature is expression of KIT (CD117), the stem cell/mast cell growth factor



**Fig. 4.** (a) Monotonous spindle morphology with irregular fascicles and variably sclerosed stroma (hematoxylin and eosin) (b, c) strong and diffuse expression, respectively, of CD117 and CD34.

receptor [8]. The most common locations of GISTs are the stomach (50–60%) and the small bowel (20–30%); however, they are occasionally found in the large bowel and the esophagus (<10%) [1]. Appendiceal GISTs are extremely rare; they represent only 0.1% of all GISTs [1]. Based on the previously reported 15 cases and our case, patients have a mean age of 66.3 years (range, 7–88 years) with a strong predilection for men (2.2:1); furthermore, nine reported cases were associated with appendicitis-like symptoms and other cases were found incidentally during surgery for other diseases or at autopsy [14]. Only the lack of elevated inflammatory serum markers and the presence of a mass or unusually enlarged appendix on imaging may indicate a GIST in this unusual location. Furthermore, it appears that primarily older men are affected; this age group is not typically affected by acute appendicitis [14].

The relationship of appendiceal GISTs to symptoms is difficult to determine because nine cases (including the present case) exhibited appendicitis-like pain, four cases were incidental findings during other surgery or autopsy, two cases were an intraluminal hemorrhage, and one case presented with diffuse abdominal pain. Appendicitis-like symptoms appear to originate from secondary inflammation due to luminal obstruction by GISTs.

Previously reported cases and the present case were adequately treated with a simple appendectomy and most cases were very low or low risk for malignancy, except only 1 case has behaved in a malignant fashion (9 mitoses per high power field); thus, adjuvant therapy (imatinib mesylate) is usually not recommended. However, the decision should be made on an individual basis because of their rarity and lack of a significant risk profile for malignancy (Table 1).

## Conclusion

Primary appendiceal GIST occur at a very low rate and their symptoms are nonspecific. Accordingly, rare tumors of appendix including GISTs should be considered in the differential diagnosis of atypical symptoms or image findings.

## Conflict of interest

There is no conflict of interest to declare.

## Funding

None.

## Ethical approval

Have obtained written consent from patient.

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## Consent

Have obtained written content from patient.

## Author's contribution

Jae Min Chun: primary author, composed paper.  
Kyoung Hoon Lim: reviewed and edited paper.

## Guarantor

Kyoung Hoon Lim and Jae Min Chun.

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