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Rare Case of Appendiceal Gastrointestinal Stromal Tumor: Diagnosis, Surgical Management, and Long-term Outcome

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Abstract

Background/Aim: Appendiceal gastrointestinal stromal tumors (GISTs) are exceptionally rare neoplasms.

Case Report: We report a case of a 68-year-old man who presented with recurrent right lower quadrant abdominal pain. Ultrasonography revealed a 10×4 mm hypoechoic submucosal tumor adjacent to the appendiceal orifice. Contrast-enhanced computed tomography demonstrated a delayed-enhancing appendiceal lesion without regional lymphadenopathy. Laparoscopic appendectomy was performed. Histopathological examination revealed an 8-mm tumor composed of spindle-shaped cells arising from the muscularis propria of the appendix. Immunohistochemical analysis showed positivity for c-kit and CD34, leading to a diagnosis of very low-risk appendiceal GIST.

Conclusion: This case highlights the diagnostic challenges posed by this extremely rare entity and contributes additional long-term outcome data to the limited literature on appendiceal GISTs. Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, most frequently arising in the stomach (approximately 60%) and small intestine (approximately 30%) (1). In contrast, GISTs originating from the appendix are exceedingly rare, accounting for approximately 0.1% of all GISTs (2). Owing to their rarity and small size, appendiceal GISTs are often discovered incidentally and may present diagnostic challenges. Herein, we report a rare case of appendiceal GIST detected preoperatively and successfully treated by laparoscopic appendectomy, with long-term recurrence-free survival.

Keywords: Appendiceal GIST, gastrointestinal stromal tumor, CD34, laparoscopic appendectomy.

Case Report

A 68-year-old man presented with recurrent right lower quadrant abdominal pain persisting for six months. Physical examination revealed a soft, flat abdomen with mild tenderness in the right lower quadrant. Laboratory findings showed no leukocytosis or elevation of C-reactive protein. Tumor markers, including carcinoembryonic antigen (CEA, 2.0 ng/ml) and carbohydrate antigen 19-9 (CA19-9, 7.1 U/ml), were within normal ranges.

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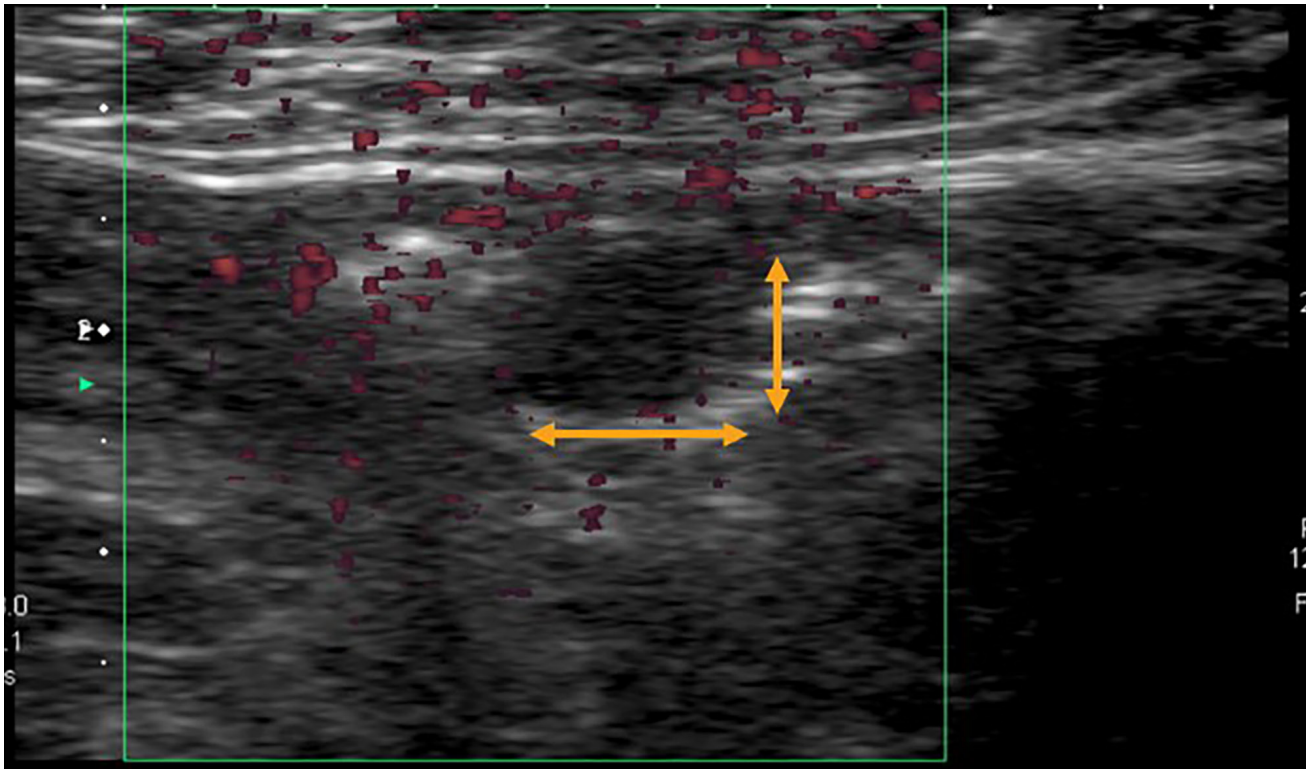

Figure 1. *Ultrasonography findings showing a hypoechoic appendiceal tumor measuring 10×4 mm.*

Abdominal ultrasonography revealed a 10×4 mm hypoechoic tumor connected to the muscular layer of the appendix, with no detectable intratumoral blood flow (Figure 1). Contrast-enhanced computed tomography demonstrated a delayed-enhancing nodular lesion in the appendix without evidence of regional lymph node enlargement (Figure 2). Colonoscopy showed no abnormal findings at the appendiceal orifice.

Based on these findings, because an appendiceal submucosal tumor was suspected, with GIST included in the differential diagnosis, biopsy for definitive histologic confirmation was not performed. The patient was referred to our department for definitive diagnosis and surgical treatment.

Laparoscopic exploration revealed a white tumor near the root of the appendix. Intraoperative frozen-section analysis suggested the possibility of GIST; therefore, laparoscopic appendectomy was performed without lymphadenectomy. Macroscopic examination of the resected specimen revealed an 8×8×6 mm white tumor located at the proximal appendix (Figure 3).

Microscopically, the tumor consisted of spindle-shaped cells originating from the muscularis propria (Figure 4a). Immunohistochemical staining was positive for c-kit, CD34, and α-smooth muscle actin, and negative for S-100 protein and desmin (Figure 4b). The MIB-1 labeling index was 0.6%. The tumor was diagnosed as an appendiceal GIST (pT1N0M0, G1, Stage I, R0) with a very low risk of recurrence according to the National Institutes of Health consensus criteria (3). No histopathological evidence of appendicitis was observed. The postoperative course was uneventful. The patient has remained alive and free of recurrence for 10 years after surgery.

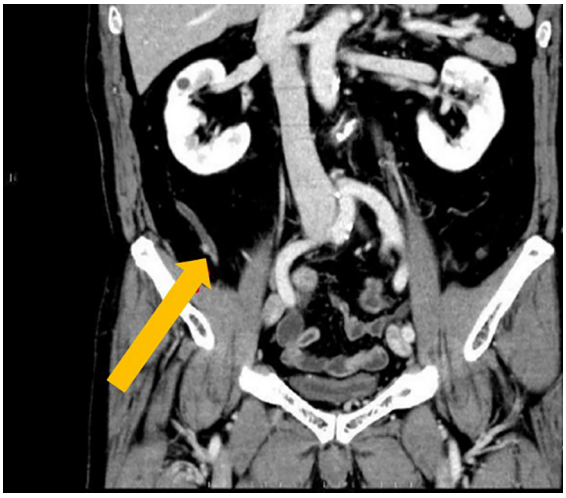

Figure 2. *Contrast-enhanced CT showing a delayed-enhancing appendiceal nodule (arrow). CT: Computed tomography.*

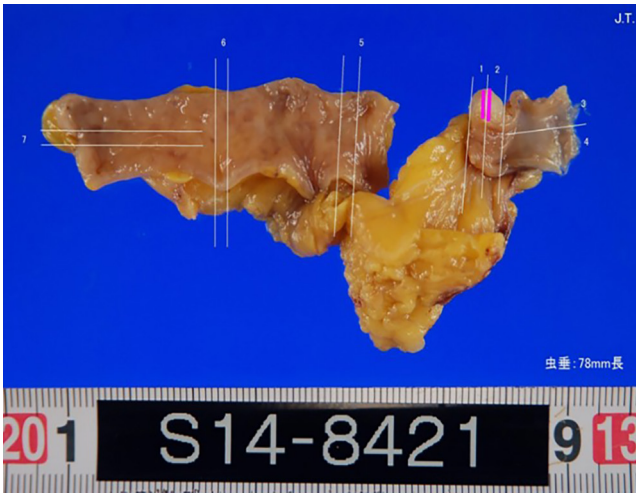

Figure 3. *Macroscopic appearance of the resected specimen showing a white tumor near the root of the appendix.*

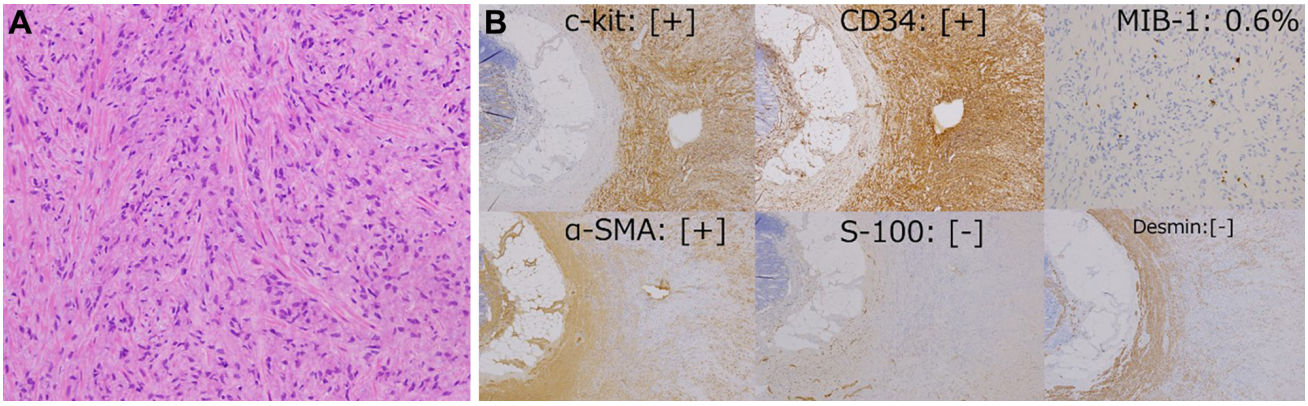

Figure 4. *Histopathological findings. (a) Hematoxylin and eosin staining showing spindle-shaped tumor cells. (b) Immunohistochemistry positive for c-kit and CD34. The MIB-1 labeling index is 0.6%.*

Ethics statement. Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Discussion

Appendiceal tumors are uncommon and are frequently detected incidentally following appendectomy performed for suspected acute appendicitis or during abdominal surgery for unrelated conditions (4). Appendiceal GISTs are particularly rare, accounting for approximately 0.1% of all GISTs (2), and are often small at the time of diagnosis.

Bouassida *et al*. reviewed eight reported cases of appendiceal GISTs, most of which were benign and measured less than 3 cm in diameter (5). Only one case demonstrated malignant features (6). Among these cases, several were discovered incidentally, while others presented with appendicitis-like symptoms. Histological evidence of acute appendicitis was absent in some cases, similar to the present case.

Regarding risk stratification, most reported appendiceal GISTs, including the present case, have been classified as very low risk according to the National Institutes of Health criteria. Long-term outcomes appear favorable, with no recurrence reported even in cases with tumor rupture managed without adjuvant therapy (5). Consistent with these findings, our patient has remained recurrence-free for a decade following complete surgical resection.

These observations suggest that appendiceal GISTs are generally indolent tumors with an excellent prognosis when completely resected. Nevertheless, accumulation of additional cases and long-term follow-up data is essential to better characterize their biological behavior and to establish optimal management strategies.

Conflicts of Interest

The Authors declare that they have no conflicts of interest.

Authors' Contributions

MU, AT, YO and SK were performed surgery and supervised the clinical management. MU, KY and SY, designed the study and collected the clinical data. MU and KY drafted the manuscript. MH, TO, SE, YF and TU supervised this study and critically revised the manuscript. All Authors read and approved the final manuscript.

Artificial Intelligence (AI) Disclosure

No artificial intelligence (AI) tools, including large language models or machine learning software, were used in the preparation, analysis, or presentation of this manuscript.

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