



Appendiceal Gastrointestinal Stromal Tumors in Adults Mini-Review of a Rare Clinical Entity

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Abstract

Gastrointestinal stromal tumors are extremely rarely located in the appendix and only a few cases have been described in the literature to date. Herein, we present a review of these case reports, focusing on epidemiology, clinical presentation of the condition, treatment, immunohistochemical characteristics, and other specific features.

Keywords: Appendix; Gastrointestinal stromal tumor; Appendiceal GIST

Abbreviations: GISTs: Gastrointestinal Stromal Tumors; NS: Not Specified; M: Male; F: Female; HPF: High Power Fields

Introduction

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors of the gastrointestinal track that express the phenotype of the interstitial cells of Cajal and are commonly associated with mutations of the tyrosine kinase receptors c-KIT [1]. They mainly occur in the stomach (60%) and the small intestine (30%) [2]; other rare locations include the esophagus, colon, rectum (<5% of cases [3]), and extra gastrointestinal areas [4].

However, presence of this tumor in the appendix is considered ever rarer; in fact, only a few cases of this entity have

been reported. Herein, a review of these cases is carried out, aiming to define the main characteristics of appendiceal GISTs.

Results and Discussion

Thorough bibliographic research was completed, with no restriction in date or language of publication, revealing thirteen articles, which included eighteen patients in total [5-16]. Unfortunately, characteristics of two cases [5] could not be retrieved; main features of the remaining sixteen cases are summarized in Table 1.

Table 1: Characteristics of reported cases of appendiceal GIST.

Author	Year	Sex	Age	Symptoms	/50 Hpf
Miettinen et al. [2]	2001	M	64	Autopsy	<1
Miettinen et al. [2]	2001	M	56	Symptoms of acute appendicitis	<1
Miettinen et al. [2]	2001	M	59	Incidental, during gastric GIST resection	<1
Miettinen et al. [2]	2001	M	72	Acute appendicitis	<1
Yap et al. [7]	2005	F	66	Symptoms of acute appendicitis	NS
Kim et al. [8]	2007	M	56	Hematochezia	NS
Agaimy et al. [9]	2008	F	78	Symptoms of acute appendicitis	<1
Agaimy et al. [9]	2008	M	72	Incidental, on bladder carcinoma follow-up	<1
Rahimi et al. [10]	2008	F	65	Coexisting with mantle cell lymphoma	<1

Elazary et al. [11]	2010	F	65	Periappendiceal abscess	9
Chung et al. [12]	2012	M	67	Symptoms of acute appendicitis	<5
Vassos et al. [13]	2013	M	48	Symptoms of acute appendicitis	NS
Bouassida et al. [14]	2013	M	75	Acute appendiceal peritonitis	NS
Ahmed et al. [15]	2014	F	52	Recurrent lower abdominal pain	NS
Back et al. [16]	2015	F	88	Incidental, history of diverticulosis	<1
Chun et al. [17]	2016	M	68	Symptoms of acute appendicitis	<1

NS: Not Specified; M: Male; F: Female; HPF: High Power Fields.

Appendiceal GIST was diagnosed in ten men and six women, with a mean age of 65.7 years and range 48-88 years (mean age for men: 63.7 years, mean age for women: 69 years). In many cases the initial presentation was a clinical condition mimicking acute appendicitis [6,7,9,12,13,17], or was indeed acute appendicitis [6,14], though it can also be an incidental finding [6,9] or show with an uncommon presentation, as hematochezia [8].

Appendiceal GISTs tend to be small-sized, benign tumors that present limited growth potential [9]. Diagnosis is mainly done postoperatively, during pathologic examination of the excised specimens, which reveals characteristic spindle cell tumors with low mitotic activity (<1/50 high power fields) in the majority of cases. Immunohistochemistry showed positivity on CD117 in nine cases [6,9,10,12,13]; CD34 was positive in eight cases [6,8,9,13,14,17]; negativity was shown on SMA, desmin and S-100 [6,8,14,17], with an exception of S-100 positivity in one case [10].

Preoperative differential diagnosis was difficult, as the tumor presented with vague and non-specific symptoms. In terms of differential diagnosis of appendiceal mesenchymal tumors, surgeons should be aware that the latest are rare entities mainly represented by leiomyoma, but they can also include leiomyosarcoma, Kaposi sarcoma, granular cell tumor, schwannoma, GIST and other rare tumors [18]. Additionally, although GISTs are the most frequent mesenchymal tumors of the gastrointestinal track, presence in the appendix is surprisingly uncommon.

In cases where the tumor is characterized as low risk, no additional treatment is considered necessary, other than appendectomy [17]. In fact, all cases were treated with appendectomy, except for one case, co-existing with mantle cell lymphoma, that was treated with right hemicolectomy [10]. As the majority of cases is considered as low risk for malignancy, with a limited number of mitoses, adjuvant therapy is not usually recommended; only one case was proved malignant [11], therefore adjuvant therapy with imatinib mesylate was started. Moreover, in another case, intraoperative tumor rupture demanded further treatment with adjuvant therapy with imatinib mesylate, as it was considered as a high-risk tumor [13].

In most cases, follow-up is relatively short (approximately after two years postoperatively), though with disease-free

results; in two cases, patients died after several years, but due to causes unrelated to the initial diagnosis of appendiceal GIST [6].

Conclusion

Gastrointestinal stromal tumors of the appendix are a rare, but remarkable entity, with unique characteristics, that often mimics the symptoms of acute appendicitis or presents non-specific symptoms. Treatment should be individualized according to the specific findings in every patient. Long-term follow-up examination is important in order to confirm the behavior of this uncommon tumor.

Conflict of Interest

No financial support was received for the preparation of this study. The authors declare that no conflict of interest exists.

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