Multicentric Gastric Gastrointestinal Tumours – One Benign and the Other Malignant

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Gastrointestinal stromal tumor (GIST) is a rare tumor of the gastrointestinal tract. It most commonly involves the stomach and presents incidentally on imaging or with upper gastrointestinal bleeding. Most of the time, it is solitary, and multicentric gastric GIST is an extremely rare condition. Multicentric gastric GIST is usually associated with certain familial conditions like neurofibromatosis, Carney-Stratakis syndrome, and Carney triad; therefore, it should be investigated whenever encountered. A GIST of 2 cm or larger in size should be excised due to its malignant potential. Herein, we describe a case of a 45-year-old female who presented with hematemesis and melena. Initial evaluation revealed a large proximal gastric mass suggestive of GIST. Intraoperatively, a second small lesion was palpated in the antrum. Total gastrectomy with peri-gastric nodal excision was performed. Histopathology confirmed a malignant GIST at the cardia and a benign GIST in the antrum. No nodal involvement was observed. The patient received adjuvant imatinib therapy and remains recurrence-free at 10-year follow-up. Concomitant benign and malignant multicentric gastric GISTs represent an extremely rare condition. Anatomical resection remains the preferred management approach, with careful long-term follow-up.

Keywords: benign, gastrointestinal tumor, malignant, stomach, total gastrectomy.

Gastrointestinal stromal tumors (GISTs) account for <2% of all gastrointestinal (GI) tumors in adults.[1] The stomach is the most commonly involved organ, representing approximately 56% of all GIST cases.[2] These tumors originate from the interstitial cells and exhibit variable malignant potential, ranging from benign to aggressive behavior.[3,4] Multicentric primary GISTs of the stomach are extremely rare and are usually associated with familial syndromes, while sporadic multicentric cases are even less common.[5] Most GISTs present as a single lesion, and the occurrence of both benign and malignant tumors in the same patient is seldom reported. Sporadic multicentric GISTs of the stomach are extremely uncommon, and the literature is limited to a few case reports and series. Clinical presentation may vary from incidental detection to symptoms of abdominal pain, GI bleeding, or mass effect.[6,7] In this report, we present a rare case of multicentric gastric GISTs, with one tumor being malignant and the other benign, and review the literature on similar cases.

Case Details

This was a 45-year-old lady who presented to us with hematemesis and melena. She had a couple of black tarry stools in the past as well. Otherwise, there was no significant medical and surgical history. She was anemic with a hemoglobin level of 6.5 g/dl, not jaundiced, and there was no supraclavicular lymph node or palpable abdominal lump.

She was transfused with 2 units of whole blood, and an emergency esophagogastroduodenoscopy (EGD) was performed. It revealed a large mass, around the cardia, ulcerated, with evidence of old clot on it, as shown in **Figure 1**; however, there was no active bleeding.

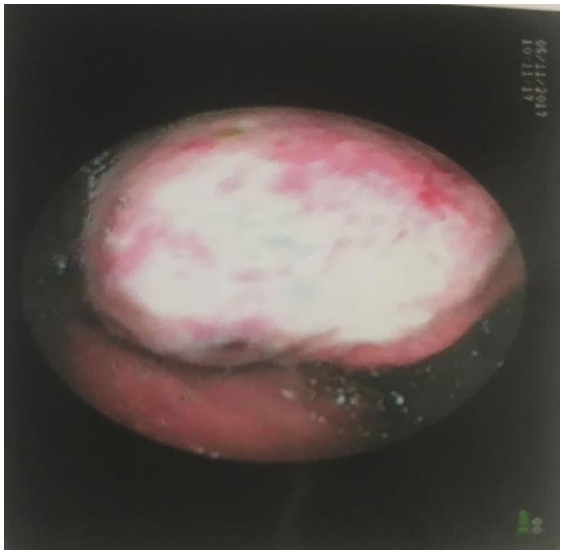

Figure 1: Endoscopy showing intraluminal growth in retroflexion

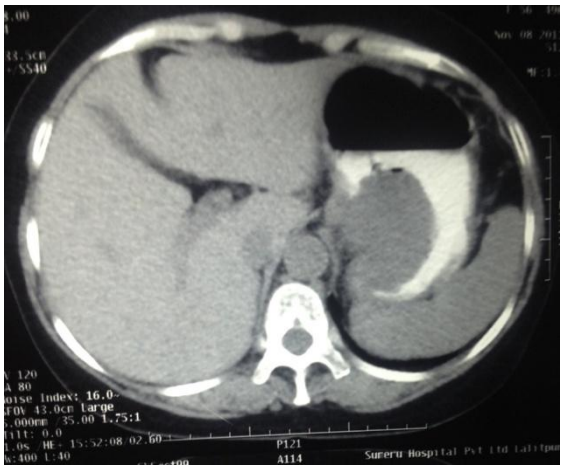

Figure 2: CT scan showing a large endophytic proximal gastric tumor

The biopsy was taken. Contrast-enhanced

CT scan revealed a large intraluminal proximal gastric mass in the body suggestive of gastrointestinal stromal tumor (GIST); non-significant perigastric nodes were visualized as presented in **Figure 2**. The biopsy report confirmed GIST.

The patient was prepared for surgery, and a proximal gastrectomy was planned. Intraoperatively, an intraluminal tumor was confirmed at the cardia on its anterior surface. Interestingly, another lump, approximately 1.5 cm, was also palpated over the anterior surface of the gastric antrum. As a frozen section facility was not available, the second lump was also thought to be GIST or a tumor of another nature. Hence, total gastrectomy with peri-gastric nodal excision was performed as shown in **Figure 3**.

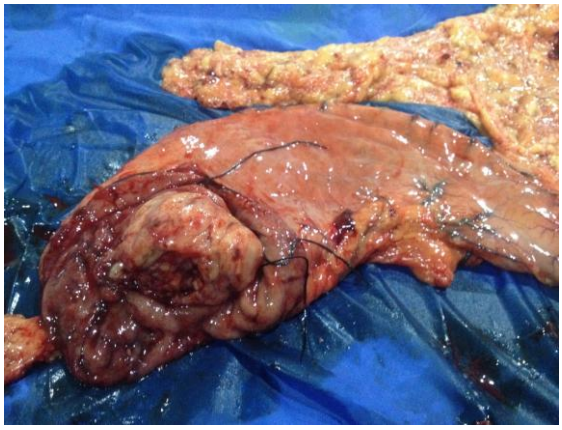

Figure 3: Total gastrectomy specimen showing a large ulcerated proximal gastric tumor

As there were no obviously palpable nodes, D2 dissection was deliberately not attempted. The spleen was preserved. Roux-en-Y esophagojejunostomy was reconstructed.

Postoperatively, she did well and was discharged on the 10th postoperative day. Subsequent histopathology confirmed the malignant GIST of the proximal stomach and the benign GIST of the antrum. Peri-gastric nodes were negative for tumors. Adjuvant imatinib therapy was started. She is on regular follow-up, and at 10 years, without any recurrence.

Discussion

The stomach is the most frequent site of GISTs; it may arise anywhere, but is more common at the fundus. GIST is usually a solitary tumor. Multiple GISTs are thought to be metastatic in origin from the main primary lesion, especially when there is a size discrepancy, and minor lesions are seen in the omentum or mesentery.[8,9] Primary GIST may be multicentric, usually in familial and occasionally in sporadic form.[5] Familial GIST, usually of the stomach, may be associated with neurofibromatosis type 1, Carney-Stratakis syndrome (GIST plus paraganglioma), and Carney triad (GIST, paraganglioma, and pulmonary chondroma), especially in young men and women.[10,11] Our patient had a small benign GIST at the antrum, whereas the larger GIST in the cardiac region was found to be malignant. She had no obvious associated features of familial GIST as mentioned above.

Most gastric GISTs are asymptomatic and incidentally detected. The most frequent symptom is bleeding in the form of hematemesis, melena, or iron deficiency anemia; our patients presented with similar symptoms. The bleeding is considered an independent risk factor for recurrence.[10,12] Other symptoms are abdominal pain, abdominal fullness, abdominal lump, and early satiety.[13,14]

GISTs with a size of 2 cm or larger should be resected due to the high potential for malignancy.[15] Tumor is resected free of margin, without any inadvertent rupture, and unlike adenocarcinoma, without any lymphadenectomy unless there are obviously enlarged nodes. Based on the tumor size and its location, a subtotal or total gastrectomy may be performed for gastric GIST.[16] GIST at and around the gastroesophageal junction or at the cardiac region is treated with proximal gastrectomy or total gastrectomy. We had planned to perform a proximal gastrectomy on the patient. However, the intraoperative finding of a separate lesion in the antrum prompted us to perform the total gastrectomy, as there was uncertainty about the nature of the lesion and the unavailability of the frozen section. Interestingly, the lesion at the antrum was not visible during endoscopy or on CT scan.

To our surprise, histopathology revealed benign GIST of the antral lesion and malignant GIST of the cardiac lesion. Publication related to concomitant benign and malignant multicentric primary gastric GISTs couldn't be searched for and identified in the literature. That's why this case seems to be a unique case.

Conclusion

Multicentric gastric GIST is a rare tumor. Whenever it is encountered, associated neurofibromatosis, Carney-Stratakis syndrome (GIST plus paraganglioma), and Carney triad (GIST, paraganglioma, and pulmonary chondroma) should be investigated. The most common presentation is upper gastrointestinal bleeding. Multicentric gastric GIST should be treated with anatomical resection whenever possible.

Concomitant benign and malignant primary gastric GISTs appear in an extremely rare condition.

Conflict of Interest: None.

References

1. Nilsson B, Bümming P, Meis-Kindblom JM, Odén A, Dortok A, Gustavsson B, et al. Gastrointestinal stromal tumors: the incidence, prevalence, clinical course, and prognostication in the preimatinib mesylate era--a population-based study in western Sweden. Cancer.

2005 Feb;103(4):821–9. DOI: 10.1002/cncr.20862

2. Sanchez-Hidalgo JM, Duran-Martinez M, Molero-Payan R, Rufian-Peña S, Arjona-Sanchez A, Casado-Adam A, et al. Gastrointestinal stromal tumors: A multidisciplinary challenge. World J Gastroenterol. 2018 May;24(18):1925–41. DOI: 10.3748/wjg.v24.i18.1925
3. Gheorghe G, Bacalbasa N, Ceobanu G, Ilie M, Enache V, Constantinescu G, et al. Gastrointestinal Stromal Tumors-A Mini Review. J Pers Med. 2021 Jul;11(8).DOI: 10.3390/jpm11080694
4. Corless CL. Gastrointestinal stromal tumors: what do we know now? Mod Pathol [Internet]. 2014;27(1):S1–16. Available from: https://doi.org/10.1038/modpathol.2013.173
5. Yin TC, Yang SF, Wang JY. Multicentric gastrointestinal stromal tumor of the stomach with wild-type KIT and PDGFRA. Genomic Med Biomarkers, Heal Sci [Internet]. 2011;3(2):81–5. Available from: https://www.sciencedirect.com/science/article/pii/S2211425411000070
6. Tarallo M, Carruezzo C, Dentice Di Accadia FM, Del Gaudio A, Caruso D, Polici M, et al. A Case Report of Multiple Gastrointestinal Stromal Tumors: Imaging Findings, Surgical Approach, and Review of the Literature. Front Surg. 2022;9:886135. DOI: 10.3389/fsurg.2022.886135
7. Shen YY, Ma XL, Yang LX, Zhao WY, Tu L, Zhuang C, et al. Clinicopathologic characteristics, diagnostic clues, and prognoses of patients with multiple sporadic gastrointestinal stromal tumors: a case series and review of the literature. Diagn Pathol [Internet]. 2020;15(1):56. Available from: https://doi.org/10.1186/s13000-020-00939-7
8. Arabi NA, Musaad AM, Ahmed EE, Abdo AA, Elhassan AM, Hassan H, et al. Primary extragastrointestinal stromal tumour of the whole abdominal cavity, omentum, peritoneum, and mesentery: a case report and review of the literature. J Med Case Rep [Internet]. 2014;8(1):337. Available from: https://doi.org/10.1186/1752-1947-8-337
9. Feng F, Tian Y, Liu Z, Liu S, Xu G, Guo M, et al. Clinicopathological features and prognosis of omental gastrointestinal stromal tumor: evaluation of a pooled case series. Sci Rep [Internet]. 2016;6(1):30748.

Available from: https://doi.org/10.1038/srep30748

10. Liu Q, Kong F, Zhou J, Dong M, Dong Q. Management of hemorrhage in gastrointestinal stromal tumors: a review. Cancer Manag Res. 2018;10:735–43.
11. Recht HS, Fishman EK. Carney-Stratakis syndrome: A dyad of familial paraganglioma and gastrointestinal stromal tumor. Radiol case reports. 2020 Nov;15(11):2071–5. DOI: 10.1016/j.radcr.2020.08.002
12. Pih GY, Jeon SJ, Ahn JY, Na HK, Lee JH, Jung KW, et al. Clinical outcomes of upper gastrointestinal bleeding in patients with gastric gastrointestinal stromal tumor. Surg Endosc. 2020 Feb;34(2):696–706.DOI: 10.1007/s00464-019-06816-9
13. Parab TM, Derogatis MJ, Boaz AM, Grasso SA, Issack PS, Duarte DA, et al. Gastrointestinal stromal tumors : a comprehensive review. J Gastrointest Oncol. 2019;10(1):144–54. DOI: 10.21037/jgo.2018.08.20
14. Caterino S, Lorenzon L, Petrucciani N, Iannicelli E, Pilozzi E, Romiti A, et al. Gastrointestinal stromal tumors: correlation between symptoms at presentation, tumor location, and prognostic factors in 47 consecutive patients. World J Surg Oncol. 2011 Feb;9:13. DOI: 10.1186/1477-7819-9-13
15. Blay JY, Bonvalot S, Casali P, Choi H, Debiec-Richter M, Dei Tos AP, et al. Consensus meeting for the management of gastrointestinal stromal tumors. Report of the GIST Consensus Conference of 20-21 March 2004, under the auspices of ESMO. Ann Oncol Off J Eur Soc Med Oncol. 2005 Apr;16(4):566–78. DOI: 10.1093/annonc/mdi127
16. Kang YK, Kang HJ, Kim KM, Sohn T, Choi D, Ryu MH, et al. Clinical practice guideline for accurate diagnosis and effective treatment of gastrointestinal stromal tumor in Korea. Cancer Res Treat. 2012 Jun;44(2):85–96. doi: 10.4143/crt.2012.44.2.85