ISSN: 2755-0117

Journal of Oncology Research Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Gastrointestinal Stromal Tumor of Esophageal Location: A Rare Case with Surgical Discussion. Case Report and Literature Review

Augusto Leite Canguçu[1]*, Virginia dos Santos Silva Neta[1], Sergio Tadeu Lima Fortunato[2], Igor Campos da Silva[3] and Rodrigo Antônio Vieira Guedes[1]

[1]Department of Clinical Oncology, Oncologia D'Or – Aliança Hospital/Rede D'Or São Luiz Hospital Group

[2]Department of Thoracic Radiology, Aliança/Rede D'Or São Luiz Hospital Group

[3]Department of Pathology, São Rafael Hospital/Rede D'Or São Luiz Hospital Group

ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal neoplasms. They arise from the intestinal wall and usually present as subepithelial neoplasms in the stomach and small intestine; however, they can appear anywhere in the gastrointestinal (GI) tract. Most of these tumors have mutations in KIT or platelet-derived growth receptor alpha (PDGFRA), while mutations in succinate dehydrogenase (SDH) or other genes are less frequent. New molecules have shown promising results in the therapy of these tumors.

***Corresponding author**

Augusto Leite Canguçu, Department of Clinical Oncology, Oncologia D'Or – Aliança Hospital/Rede D'Or São Luiz Hospital Group, Salvador-BA. Brazil. Tell: +55 71 99687-1064.

Received: July 26, 2023; **Accepted:** August 02, 2023; **Published:** August 10, 2023

Keywords: Gastrointestinal Stromal Tumor, Gist, Esophageal Gist

Introduction

Gastointestinal stromal tumor (GIST) is a rare tumor that accounts for about 3% of all tumors of the gastrointestinal (GI) tract [1]. Gastric location corresponds to 50 to 60% of cases, midgut to 30 to 35% and other sites of gastrointestinal tract are less common, such as colorectal and esophagus [2]. GISTs likely originate from the interstitial cells of Cajal (ICCs), sometimes referred to as the GI pacemaker cell and represent a distinct entity from other mesenchymal tumors of the GI tract [3]. The development of effective therapies is focused on the molecular profile of these tumors, where the KIT proto-oncogene mutation is the most common and on which therapies with better results have been sought [4].

Case Report

Female, 69 years old, with dyspeptic symptoms such as nausea and vomiting for 1 year. In upper digestive endoscopy, nodulation was visualized without alteration of the mucosa, not biopsied (Figure 1). During the period, she was diagnosed of pulmonary infection secondary to COVID and a chest X-ray identified an opacity in the power mediastinum (Figure 2). Chest CT scan showed heterogeneous expansive formation in the posterior mediastinum, located in the distal esophageal wall/esophageal-gastric transition, indeterminate, admitting among the differential diagnoses inflammatory pseudotumor and neoplasm, including benign lesions (leiomyoma) or malignant (GIST, for example). In endoscopic ultrasound (EUS), a hypoechoic formation was seen involving distal esophagus with a predominant exophytic component. Diagnostic puncture with pathological report showed spindle cell proliferation without significant atypia, suggestive of GIST versus leiomyoma; then underwent distal esophagectomy.

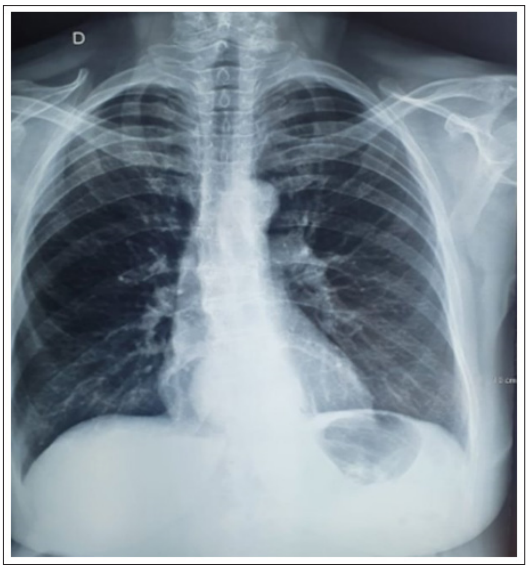


Figure 1: Chest X-ray showing enlargement in the superior mediastinum

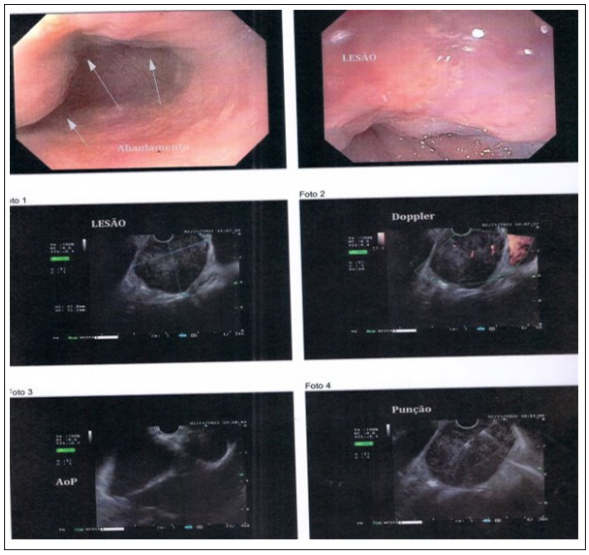
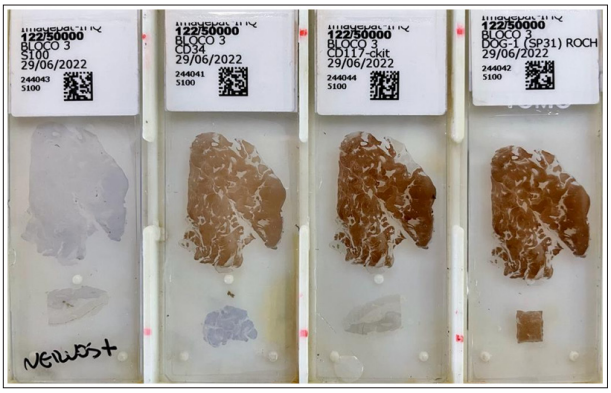


Figure 2: Upper digestive endoscopy image showing bulging mucous due to submucosal growth. At ultrasound (images below), evidence of submucosal expansive lesion

Pathological report confirmed spindle cell epithelioid neoplasia, measuring 6.0 x 4.0 x 3.5 centimeters (fragmented piece), mitotic count 9 mitoses per 5 mm, high cellularity, moderate cell pleomorphism, present necrosis at 15%, grade 2, positive angiolymphatic and perineural invasion, free margins, lymph node metastasis in 4/6 TEG lymph nodes (largest deposit measuring 10 mm), in addition to 2 nodules in the submucosa of the esophagus both measuring 7 mm. Immunohistochemistry was consistent with GIST due CD 117, DOG-1 and CD34 positive, S100 negative (Figure 3). Pathological staging pT3 pN1, stage IV.



Hospital discharge after 10 days of hospitalization with good evolution. The molecular profile of the GIST demonstrated positive cKIT mutation in Exon 11. PET-CT scan showed uptake in liver segments V/VI (SUV 3.3). Magnetic resonance imaging (MRI) of the abdomen confirmed multiple small liver lesions suggestive of secondary involvement [5-16]. Started Imatinib 400mg/day as a first line systemic treatment for metastatic GIST.

References

1. Fülöp E, Marcu S, Milutin D, Angela Borda (2009) Gastrointestinal stromal tumors: review on morphology, diagnosis and management. Rom J Morphol Embryol 50: 319-326.
2. Rossi CR, Mocellin S, Mencarelli R, Mirto Foletto, Pierluigi Pilati, et al. (2003) Gastrointestinal stromal tumors: from a surgical to a molecular approach. Int J Cancer 107: 171-176.
3. Fletcher CD, Berman JJ, Corless C, Fred Gorstein, Jerzy Lasota, et al. (2002) Diagnosis of gastrointestinal stromal tumors: a consensus approach. Int J Surg Pathol 33: 459-465.
4. Hirota S, Isozaki K, Moriyama Y, T Nishida, S Ishiguro (1998) Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. Science 279: 577-580.
5. Blesius A, Cassier PA, Bertucci F, Ray-Coquard I, Bui B, et al. (2011) Neoadjuvant imatinib in patients with locally advanced non metastatic GIST in the prospective BFR14 trial. BMC Cancer 15: 11-72.
6. Hoheberger P, Langer C, Wendtner CM, Werner Hohenberger, Annette Pustowka, et al. (2012) Neoadjuvant treatment of locally advanced GIST: Results of APOLLON, a prospective, open label phase II study in KIT- or PDGFRA-positive tumors. J Clin Oncol 30:15.
7. Kurokawa Y, Yang HK, Cho H, Ryu MH, Masuzawa T, et al. (2017) Phase II study of neoadjuvant imatinib in large gastrointestinal stromal tumours of the stomach. Br J Cancer 117: 25-32.
8. Rutkowski P, Gronchi A, Hohenberger P, Bonvalot S, Schöffski P, et al. (2013) Neoadjuvant imatinib in locally advanced gastrointestinal stromal tumors (GIST): the EORTC STBSG experience. Ann Surg Oncol 20: 2937-2943.
9. Miettinen M, Wang ZF, Lasota J (2009) DOG1 antibody in the differential diagnosis of gastrointestinal stromal tumors: a study of 1840 cases. Am J Surg Pathol 33: 1401-1408.
10. Debiec-Rychter M, Sciot R, Le Cesne A, Schlemmer M, Peter Hohenberger, et al. (2006) KIT mutations and dose selection for imatinib in patients with advanced gastrointestinal stromal tumours. Eur J Cancer 42: 1093-1103.
11. Heinrich MC, Owzar K, Corless CL, Hollis D, Borden EC, et al. (2008) Correlation of kinase genotype and clinical outcome in the North American Intergroup Phase III Trial of imatinib mesylate for treatment of advanced gastrointestinal stromal tumor: CALGB 150105 Study by Cancer and Leukemia Group B and Southwest Oncology Group. J Clin Oncol 26: 5360-5367.
12. Gastrointestinal Stromal Tumor Meta-Analysis Group (MetaGIST) (2010) Comparison of two doses of imatinib for the treatment of unresectable or metastatic gastrointestinal stromal tumors: a meta-analysis of 1,640 patients. J Clin Oncol 28: 1247-1253.
13. Boikos SA, Pappo AS, Killian JK, Michael P LaQuaglia, Chris B Weldon, et al. (2016) Molecular Subtypes of KIT/PDGFRA Wild-Type Gastrointestinal Stromal Tumors: A Report From the National Institutes of Health Gastrointestinal Stromal Tumor Clinic. JAMA Oncol 2: 922-928.
14. Heinrich Michael C, Robin L Jones, Margaret von Mehren , Patrick Schöffski , César Serrano et al. (2020) Avapritinib in advanced PDGFRA D842V-mutant gastrointestinal stromal tumour (NAVIGATOR): a multicentre, open-label, phase 1 trial. Lancet Oncol 21: 935-946.
15. Corless CL, Schroeder A, Griffith D, Ajia Town, Laura McGreevey, et al. (2005) PDGFRA mutations in gastrointestinal stromal tumors: frequency, spectrum and in vitro sensitivity to imatinib. J Clin Oncol 23: 5357-5364.
16. Boikos SA, Pappo AS, Killian JK, Michael P LaQuaglia, Chris B Weldon, et al. (2016) Molecular Subtypes of KIT/PDGFRA Wild-Type Gastrointestinal Stromal Tumors: A Report From the National Institutes of Health Gastrointestinal Stromal Tumor Clinic. JAMA Oncol 2: 922-928.