# ATYPICAL CHARACTERISTICS AND BEHAVIOR OF GIST

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## Abstract

Gastrointestinal stromal tumors (GISTs), constituting digestive neoplasms, typically have a favorable prognosis, although occasional distant extension can occur. The main prognostic factors associated with GISTs are described as lesion size and the number of mitoses per field. These lesions primarily metastasize to the liver, omentum, and peritoneum, with lymphatic dissemination being exceptionally rare. Although the presence of lymph node involvement has been identified as an independent predictor of mortality, since it is an infrequent finding, further studies are needed to understand its prognostic and therapeutic implications. This clinical case describes the approach to a 75-year-old patient with no significant medical history presenting with gastric GIST, who notably had multiple tumor-associated lymph nodes on staging studies, requiring treatment with Imatinib.

**Keywords**: GIST, metastases, adenopathy.

## Introduction

GIST (gastrointestinal stromal tumours) are digestive neoplasms with a usually favourable prognosis, although they can present an aggressive clinical course with distant extension. The incidence in Spain is 1.24 cases each 100,000 inhabitants, with the main site being the stomach and causing epigastralgia, abdominal distension or gastrointestinal bleeding when symptomatic[1]. The endoscopic and radiological images of a GIST characterised by distant dissemination exclusively by the lymphatic route are presented below.

## Clinical Case

A 75-year-old patient with no personal history of interest was referred for oral intolerance and constitutional syndrome and underwent gastroscopy, where a subcardial lesion with a submucosal appearance and an ulcerated surface was found, which appeared to be malignant **(Figure 1)**.

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*Lecuona Muñoz M, Fernández García R, López González E, Tendero Peinado C, Redondo Cerezo E. Atypical Characteristics and Behavior of GIST RAPD 2024;47(3):133-135. DOI: 10.37352/2024473.7*


*Figure 1.* ***Gastroscopy . At the subcardial level, in the upper body towards the greater curvature, a submucosal lesion of about 25 mm, friable to friction and with an ulcerated centre covered by fibrin, can be identified.***

In the extension study performed by Computerised Axial Tomography **(Figures 2 and 3)** a stenosing lesion compatible with GIST was identified, with an approximate size of 16x11x14 cm (craniocaudal, transverse and anteroposterior diameters respectively). Adenopathies of tumour origin were observed in the left common and external iliac, celiac trunk, retroperitoneum and interaortocaval space, as well as contiguous infiltration of the upper pole of the spleen and left adrenal gland.

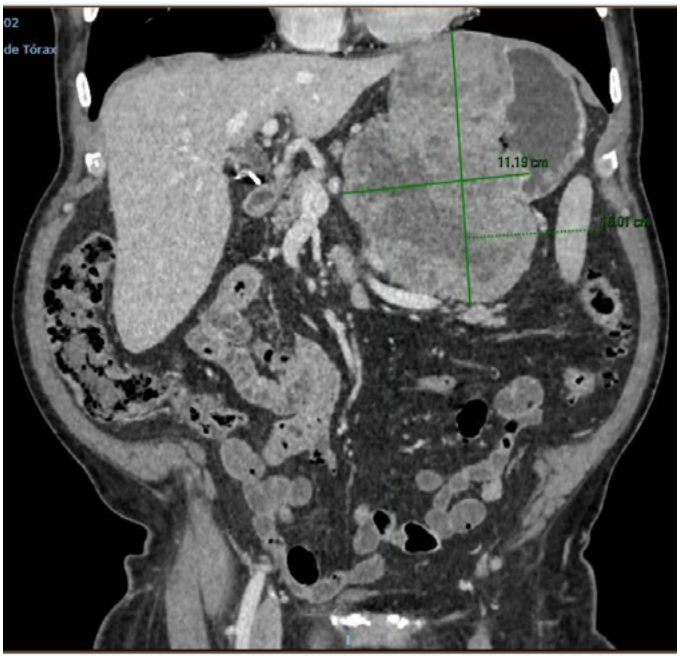


*Figure 2.* ***Abdominal CT scan. Bulky ulcerated submucosal gastric tumour with dependence on the cardia-fundus region compatible with gastrointestinal stromal tumour (GIST). Approximate maximum dimensions of 16x11x13.7 cm (cc x t x ap).***

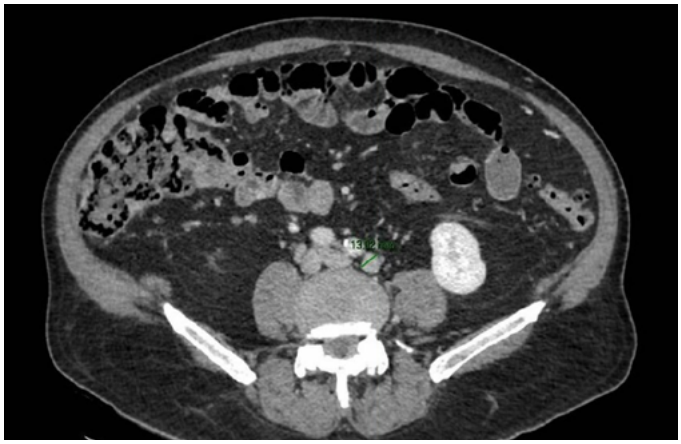

*Figure 3.* ***Abdominal CT scan. Left common and external iliac and left external iliac adenopathies of 13 and 10 mm, respectively.***

Biopsies obtained by endoscopy confirmed positivity for CD34, DOG1 and CD117, and six high magnification mitoses/field were counted. The study was completed by requesting PET-CT **(Figure 4)** and mutational analysis was also performed, identifying deletion 558 in exon 11 of the KIT gene. The patient was referred to Oncology and treatment with Imatinib was started and has continued for a year with adequate response

*Figure 4.* ***PET-CT scan. Large hypermetabolic mass in the hypochondrium and left flank, dependent on the gastric chamber, with invasion of the spleen and left adrenal gland. It has a SULmax of 15.19.***


DOI: 10.37352/2024473.7

and tolerance, with the only notable finding being the appearance of massive post-treatment necrosis two months after starting this therapy.

## Discussion

Gastrointestinal stromal tumours (GIST) are the most common digestive neoplasm of mesenchymal origin, and are characterised by the presence of a mutation in the C-Kit protein (CD117) in up to 95% of cases[2]. Our patient had some features that have been identified as risk factors for distant involvement, such as a size greater than 10 cm and a mitosis count greater than five mitoses per field[3]. In these neoplasms, mutational analysis prior to initiation of Imatinib treatment is recommended as it provides prognostic information and predicts the likelihood of response to tyrosine kinase inhibitor drugs. Mutations in exon 11 of the KIT gene, such as the one in our patient, are usually associated with a good response to Imatinib[4].

Regarding distant extension, GIST tumours mainly metastasise to the liver, omentum and peritoneum, with lymphatic spread being exceptional, unlike what was reported in the present case. A higher probability of adenopathy has been described in paediatric patients or when it occurs in association with Carney syndrome, but it is very rare in sporadic tumours that appear in adulthood. A study by Zachary E Stiles *et al*[5] included 5,018 patients with gastrointestinal GIST, of whom 301 (6%) had lymphatic involvement. In this study, the presence of lymph node metastases was associated with increased mortality, although given that this is an infrequent finding, further studies are needed to define its prognostic and therapeutic implications.

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*DOI: 10.37352/2024473.7*