

Presentation and Management of Duodenal Gastrointestinal Stromal Tumours: A Case Series



Giovanna Mennitti Shimoda¹, Alexia Paganotti¹, Tiago Caserta Asvolinsque Peixoto¹, Mariana Langanke Gonçalves¹, Pedro de Souza Lucarelli Antunes¹, Luca Giovanni Antonio Pivetta¹, José Gustavo Parreira¹, José Cesar Assef¹



1 – Faculdade de Ciências Médicas da Santa Casa de São Paulo, Disciplina de Cirurgia – São Paulo – Brasil

INTRODUCTION

Gastrointestinal Stromal Tumours (GISTs) represent 0,1-3% of all gastrointestinal neoplasms and frequently result from activating mutations of the gene KIT¹. Duodenal GISTs are even rarer, since duodenum is considered a less common primary site of the tumour and are also described as symptomatic in over 80% of the cases. The symptoms include early satiety, fatigue, intraperitoneal hemorrhage, intraluminal gastrointestinal bleeding and abdominal pain². This series of two cases describes the range of symptoms, diagnosis and standard management of GISTs located on an extremely rare site - the fourth part of the duodenum - and which were also presented as an uncommon source of important episodes of gastrointestinal bleeding.

CASE PRESENTATION

Case 1: a 54-year-old male, was admitted referring darkened feces, which started a week before admission, as well as general malaise, palpitation, pallor and dyspnea. It was administered three packed red blood cells, since laboratory exams pointed to decreased levels of hemoglobin (5,7g/dL). An angiotomography demonstrated the existence of a nodular intramural formation, without promoting stenosis, in the fourth portion of duodenum. The patient was submitted to a wedge resection duodenectomy followed by duodenorrhaphy. The biopsy showed a 4.9 cm tumour with low histological grades and the absence of necrosis, composed of fusiform cells. Immunostaining of the tumor cells were strongly positive for CD34 and c-KIT, strongly suggesting the diagnosis of GIST.

Case 2: a 59-year-old woman was admitted with a one-day history of four episodes of haematemesis and two of melena, besides paleness and tachycardia.

Due to decreased levels of hemoglobin (6.2 g/dL), the conduct was transfusion of two packed red blood cells. An esophagogastroduodenoscopy revealed an enantematic pangastritis with mild duodenal scars, without active bleeding and, due to a suspected tumour, the patient was submitted to a duodenectomy. During surgery, the presence of a 5 cm tumour lesion in the fourth duodenal portion was confirmed. The biopsy showed a tumour with low histological grades, necrosis below 5% and composed of fusiform cells. Immunostaining was positive for C-KIT and CD34, corroborating the diagnosis of GIST.

DISCUSSION

Despite being rare, GISTs represent the most common mesenchymal tumour that can arise anywhere from the esophagus to the anus³. The clinical presentation is based on location, size and presence of intraluminal bleeding, which stands out

particularly in duodenal tumours², as observed in this case series. Early surgical intervention is the treatment of choice and, in general, complete surgical resection is accomplished in 40-60% of all GISTs patients⁴. The case series demonstrated a rarer presentation of completely excised duodenal GISTs and highlighted the importance of surgical treatment and a long-term follow-up to prevent poor prognosis, recurrences or metastasis.

REFERENCES

- [1] Joensuu, H. Gastrointestinal stromal tumor (GIST). *Ann Oncol.* 2006 Sep;17 Suppl 10:x280-6.
- [2] Demetri GD, von Mehren M, Antonescu CR, et al. NCCN Task Force report: update on the management of patients with gastrointestinal stromal tumors. *J Natl Compr Canc Netw.* 2010 Apr;8 Suppl 2(02):S1-41; quiz S42-4.
- [3] Boselli C, Cirocchi R, Gemini A, et al. Urgency surgical treatment for duodenal GISTs: analysis of aged patients and review of the literature. *Aging Clin Exp Res.* 2017 Feb;29(Suppl 1):1-6.
- [4] Nishida T, Blay JY, Hirota S, et al. The standard diagnosis, treatment, and follow-up of gastrointestinal stromal tumors based on guidelines. *Gastric Cancer.* 2016 Jan;19(1):3-14.