Multicentric Jejunal and Omental GIST with an Unusual Clinical Presentation—A Case Report

Abid Iqbal · Fadl H. Veerankutty · M. S. Sulfekar ·
T. B. Culas

Received: 4 November 2013 / Accepted: 21 January 2014 / Published online: 9 February 2014
© Indian Association of Surgical Oncology 2014

Introduction

GISTs, are uncommon mesenchymal, malignant or potentially malignant tumors affecting the gastrointestinal tract derived from interstitial cells of Cajal (Intestinal pace maker cells). GISTs are the most common non epithelial tumors of the digestive tract, accounting for only 1 % of all gastrointestinal malignancies [1] and for 5.7 % of all sarcomas [2]. They are defined as specific Kit (CD117)-positive and Kit or platelet-derived growth factor receptor alpha (PDGFRA) mutation-driven tumors [3]. This paper reports a case of massive lower gastrointestinal bleeding due to a large GIST of the jejunum which presented as shock and cerebrovascular accident with a concurrent omental GIST.

Case Report

A 57 year old male with no past history of significant illnesses presented with history of massive bleeding per rectum of 2 hours duration. Patient collapsed in emergency room which was followed by one episode of generalized tonic clonic seizures. Patient was resuscitated and stabilized. He had pallor on clinical examination. Per rectal examination revealed

massive bleeding but no growth. Colonoscopy could not be done as view was obscured with blood. Nasogastric aspirate did not contain blood and an Upper GI endoscopy was done to further rule out upper GI bleed. A contrast enhanced CT scan of abdomen showed a heterogeneously enhancing tumour in relation to proximal small bowel.

An emergency exploratory laparotomy was performed. Intraoperatively there was an extra luminal hard polypoidal mass of 6 cm×5 cm in the antimesenteric border of proximal jejunum 15 cm away from Ligament of Treitz. It was excised with 5 cm proximal and distal margins. There was another hard 2 cm × 2 cm nodular lesion in greater omentum (Fig. 1). There was a 5 mm × 5 mm nodular lesion in mesentery of involved jejunum. Involved part of greater omentum also resected with an adequate gross margin. Patient developed aphasia in post operative period. CT scan of brain showed features suggestive of massive infarct in Left Middle Cerebral Artery territory. MRI was done to rule out a cerebral metastatic lesion.

A diagnosis of GIST was made on histopathological examination (Fig. 2a, c, d). Immunohistochemistry showed positivity for CD117 (Fig. 2b, e). Omental lesion showed numerous mitotic figures and was positive for Ki 67 also (Fig. 2f). Mesenteric node showed reactive hyperplasia only (Table 1). Patient was discharged on 12th post operative day and sent for neurological rehabilitation. Patient was started on Imatinib. There is no evidence of tumour recurrence after 6 months follow up.

Discussion

The term GIST was introduced by Mazur and Clark in order to indicate a distinct heterogeneous group of mesenchymal neoplasms of spindle or epithelioid cells of varying differentiation. The incidence of GIST is very low (2 in 1,00,000) while jejunal GISTs are rarest [4], accounting for 0.1–3 % of all

A. Iqbal (✉) · F. H. Veerankutty · M. S. Sulfekar · T. B. Culas
Department of General Surgery, Government Medical College
Thrissur, Mulamkunnathukavu, Thrissur Pincode 680596, Kerala,
India
e-mail: iqbal.abid@gmail.com

F. H. Veerankutty
e-mail: fadl_05@yahoo.com

M. S. Sulfekar
e-mail: drsulfekarms@hotmail.com

T. B. Culas
e-mail: tbculas@gmail.com

Fig. 1 Gross specimen. **a** and **c** Omental lesion. **b** and **d** Jejunal lesion

gastrointestinal (GI) tumors. The most common site of presentation is stomach (about 2/3). About one-fourth develop in the small intestine, usually in duodenum [5]. The most common clinical manifestation for symptomatic GISTs is occult gastrointestinal (GI) bleeding from mucosal ulceration.

GISTs often present with nausea, vomiting, abdominal pain, metastatic diseases, and bowel obstruction. Some are

found incidentally through medical imaging for other purposes or through surgery for other conditions. In our case, the patient presented with massive fulminant GI bleeding. Many studies emphasize the CD117 expression in GISTs [6]. A method of assessing prognosis of GIST and its malignant potential is by measuring the rate of cell turnover by either counting the mitotic figures in each

Fig. 2 Microscopy and IHC

Table 1 Immunohistochemistry status

Site	Ki67	CD117
Jejunum	–	+++
Omentum	++	+++

high power field or by indirect indicators of cell turn over like Ki-67 [7].

In this reported case, GIST was diagnosed as a malignancy with moderate level of pleomorphism and mitotic activity with IHC staining for CD117, and Ki-67. Omental lesion showed higher number of mitotic figures and was more intensely stained for Ki-67. The treatment of choice is the complete resection of the tumour with its pseudocapsule intact. Patients with unresectable tumours or with metastatic disease are treated with Kit/PDGFR tyrosine kinase inhibitors.

The abdominal CT correctly imaged the location and the size of the tumor in this case. It also excluded liver or peritoneal metastases and evaluated the extension of the primary tumor. As most GISTs occur as exophytic growths, CT imaging is more useful than endoscopy and barium studies. A contrast enhanced CT is the preferred investigation for omental lesions and metastatic lesions [8].

Conclusion

The purpose of this article is to report a case of multicentric GIST which presented as massive lower GI bleeding resulting in shock and cerebrovascular accident. Malignancies that present as ulcerated lesions that bleed persistently is most characteristic of GIST. We aim to highlight the need to consider the diagnosis of GIST in patients with fulminant GI bleeding, as well the need for proper intra operative assessment for extra GI sites for concurrent lesions in such patients.

Limitations

A molecular study of the resected specimen will be required to conclusively prove the multicentricity of this case of GIST.

References

1. Nowain A, Bhakta H, Pais S, Kanel G, Verma S (2005) Gastrointestinal stromal tumors: clinical profile, pathogenesis, treatment strategies and prognosis. *J Gastroenterol Hepatol* 20: 818–824
2. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF (2000) Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg* 231:51–58. doi: [10.1097/00000658-200001000-00008](https://doi.org/10.1097/00000658-200001000-00008)
3. Hirota S, Isozaki K, Moriyama Y, Hashimoto K, Nishida T, Ishiguro S, Kawano K, Hanada M, Kurata A, Takeda M, Muhammad Tunio G, Matsuzawa Y, Kanakura Y, Shinomura Y, Kitamura Y (1998) Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. *Science* 279:577–580
4. Miettinen M, Sobin L, Lasota J (2005) Gastrointestinal stromal tumors of the stomach: acclinicopathologic, immunohistochemical, and molecular genetic study of 1765 cases with long-term follow-up. *Am J Surg Pathol* 29(1):52–68
5. Kramer K, Siech M, Sträter J, Aschoff AJ, Henne-Bruns D (2005) GI hemorrhage with fulminant shock induced by jejunal gastrointestinal stromal tumor (GIST) coincident with duodenal neuroendocrine carcinoma (NET)+neurofibromatosis (NF)-Case report and review of the literature. *Z Gastroenterol* 43(3):281–288
6. Hasegawa T, Matsuno Y, Shimoda T, Hirohashi S (2003) Gastrointestinal stromal tumour: consistent CD117 immunostaining for diagnosis, and prognostic classification based on tumor size and MIB-1 grade. *Hum Pathol* 33:669–676
7. Romeo S, Debiec-Rychter M, Van Glabbeke M, Van Paassen H, Comite P, Van Eijk R, Oosting J, Verweij J, Terrier P, Schneider U et al (2009) Cell cycle/apoptosis molecule expression correlates with imatinib response in patients with advanced gastrointestinal stromal tumors. *Clin Cancer Res* 15:4191–4198
8. Sandrasegaran K, Rajesh A, Rushing DA, Rydberg J, Akisik FM, Henley JD (2005) Gastrointestinal stromal tumors: CT and MRI findings. *Eur Radiol* 15:1407–1414