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Original Research Article

A study of variables in gastrointestinal stromal tumor: surgical experience at a single institute

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ABSTRACT

Background: Gastrointestinal stromal tumors (GIST) have been recognized as a biologically distinctive tumor type, different from the smooth muscle and neural tumors of the gastrointestinal tract. They constitute majority of the gastrointestinal mesenchymal tumors of the gastrointestinal tract and are considered to be refractory to conventional chemotherapy or radiation. Surgery remains the mainstay of primary tumor management of GIST.
Methods: This was a cross sectional study of the diagnosis, management and clinical outcome of cases diagnosed with GIST of the gastrointestinal tract from a teaching institute in a single year and a comparison of findings with previously published cases. Study included patients newly diagnosed with GIST and managed in department of general surgery from 10 April 2018 to 31 March 2019.
Results: Most patients were of age group 60-69 years with an almost equal incidence among males and females. Stomach and small bowel were found to be the commonest sites with an equal distribution of 45.45% with an average tumor size of 5 cms.
Conclusions: Surgical treatment remains the main stay of management for the GIST, depending on the location, staging and metastasis. Staging laparoscopy may be done to assess respectability as peritoneal deposits can be missed by CT scan. Immunohistochemistry is mandatory in defining GIST.

Keywords: GIST, Mesenchymal tumor, Cells of Cajal, Imatinib

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors of the gastrointestinal tract. They arise from the interstitial cells of Cajal, located in the wall of the gastrointestinal tract. They are pluripotent cells with neuronal characteristics such as smooth muscle cells being called pacemaker of peristalsis.[1] Gastrointestinal stromal tumours (GISTs) are rare malignancies, accounting for less than 1% of all gastrointestinal tumours, however, they are the most common mesenchymal tumors of the gastrointestinal tract.[2] Previously GIST were classified under smooth muscle neoplasms which included leiomyomas, leiomyosarcomas, leiomyoblastomas and other rare entities, but when the mutations in KIT and PDGFRA were discovered and Immunohistochemistry (IHC) showed lack of features of smooth muscle differentiation, GIST was established as a new diagnostic entity.[3,4] GIST are most common in middle aged and older patients with the mean age of approximately 65 years and peak incidence in the 70-79 year old.[4,5] They are rare in patients under 30 years pf age, although pediatric and young adult GIST do account for 0.5-2.7% of all GISTs.[6] The recognition of the KIT-gene, the role it has in the development of GIST and the ability to treat the disease with tyrosine kinase inhibitors have revolutionized the diagnosis, treatment and prognosis of GIST.

The histopathological appearance of GIST have three morphological appearances named as spindle cell type (70%), epitheloid type (20%) and, mixed (10%).[7] Spindle cell GISTs have relatively uniform cells arranged in fascicles. Nuclear palisading and juxtanuclear cytoplasmic vacuoles are often seen. Epitheloid GISTs have round to polygonal cells with variably basophilic cytoplasm and round nuclei. Mixed GISTs have abrupt transitions between the two appearances.[7,8] Although histological features can be suggestive of diagnosis of GIST but they are not specific and the diagnosis relies on the immunohistochemical (IHC) markers, the most important being CD117 and recently discovered DOG1. CD117 is expressed by KIT-gene and is positive in 90-95% of GISTs, it may also be positive in non GIST tumors including melanoma and kaposi sarcoma. Another IHC marker CD34 may be useful and is positive in 81% of cases, although it has much lower specificity. [8,9]

Stomach accounts around 40-60% of GISTs followed by jejunum and ileum which comprise 25-30% of all GISTs. Less frequently GISTs occur in colorectal area (5-15%), duodenum (5%) and esophageal (<1%). Very rarely mesentery, retroperitoneum comprise of extra gastrointestinal stromal tumors.[4,8] Metastasis most commonly occur in liver, peritoneum and omentum, very rarely pulmonary, bone and brain metastasis occur.

Patient present either symptomatically as having a vague abdominal pain or mass, early satiety, bloating, fullness of abdomen or fatigue. Some patients might present with occult bleeding per rectum. While the other group which are asymptomatic have an incidental finding of GIST while undergoing radiological investigation for some other suspected condition.[7]

The investigation of the patients begins with an ultrasound scanning which may demonstrate the mass, size and the tissue of origin. Once the ultrasound is confirmed, a CT scan or a PET scan may be useful imaging modality for diagnosis and staging. GIST appears as a solid mass with IV contrast enhancement either exophytic or endophytic sometimes with dual component so called dumbbell shaped lesion. Endoscopic ultrasound of lesion allows fine needle aspiration which is highly sensitive and specific for diagnosis of GIST.[10] Tissue diagnosis is important along with the CT scan or PET scan report in order to decide the further management of the patient..

METHODS

A cross sectional study was carried out from 01 April 2018 to 31 March 2019 at Department of General surgery, GCS Medical college Hospital and research Centre, Ahmedabad. The Institutional Ethics Committee approval was obtained before starting the study. Patients with a radiological investigation suggestive of a tumor arising from stomach or intestine with a query of gastrointestinal stromal tumor were selected. Some selected cases particularly query of GIST of stomach were investigated further for upper gastroscopy and biopsy. All these patients were explored with a diagnostic laparoscopy first followed by resection of the tumor according to location of tumor. Some cases with distant metastasis were excluded. Only the operated cases which had the final histopathology report of gastrointestinal stromal tumors were included. Statistical analysis of the data was done using microsoft excel software.

RESULTS

Out of the 11 patients operated and confirmed for GIST, 6 patients were male and 5 patients were female. The age group of the study ranged from 29 years to 70 years, median age being 60 years and the mean age of 56.1 years (Table 1).

Table 1: Age and gender wise distribution of cases.

Age in years	Male	Female	Total
20-29	0	1	1
30-39	0	1	1
40-49	0	1	1
50-59	1	0	1
60-69	5	1	6
>70	0	1	1
Total	6	5	11

Table 2: Distribution of cases according to location.

Location	No. of cases	%	Exophytic	Endophytic
Stomach	5	45.45	1	4
Duodenum	1	9.09	1	0
Jejunum	0	0	0	0
Ileum	5	45.45	5	0
Others	0	0	0	0
Total	11		8	3

2 patients out of the 11 had malignant GIST as histological report whereas rest all was benign tumors. Abdominal pain was found in 6 out of 11 patients, being the most common presenting feature in our study followed by malaise in 5 out of 11 patients.

The most common location in our study was small bowel accounting 54.54% particularly the ileum which was 5 out of 6 small bowel GIST (83.33%) and 1 was from the duodenum. Stomach GISTs comprised only 36.36% of the GISTs and one was from the abdominal wall (Table 2).

All patients were operated for Laparotomy and resection of the tumor. IHC reporting was done in all 11 patients. CD117 was positive in 11 cases and CD 34 were positive in 9 cases (Table 3).

Table 3: Frequency of IHC marker positivity.

IHC marker	Positivity in	Percentage (%)
CD117	11	100
CD34	9	81.81
S-100	0	0
CK	1	9.09
EMA	0	0

Table 4: Distribution according to location and size of tumor.

Site	Size		Total
	<5 cm	>5 cm	
Stomach	1	3	4
Duodenum	1	1	2
Jejunum	0	0	0
Ileum	1	4	5
Total	3	8	11

Most common size range of the tumor was >6 cm and number of patients with size >6 cm were 5 out of 11 patients. 8 were exophtyic and 3 were endophytic (Table 4).

DISCUSSION

GISTs are most commonly seen in 5th or 6th decade of the life. Experience from Italian group showed the mean age was 60 years with equal incidence in males and females.[13] Sorour et al reported the mean age of patients at diagnosis of GIST was nearly 50 years, whereas Miettinen et al revealed GISTs occurs rarely below 40 years and very unusual in children.[11,12] Our report shows that only 2 cases (20%) were diagnosed in patients 40 years old and 8 cases beyond 50 years of age. Wang et al reported high incidence of GIST between 50 and 59 years of age.[13] In a Middle East population, GIST occurred in males over 40 years of age in most of the cases.[12]

The most common site involved in GIST is the stomach which accounted 60%. GIST accounted nearly 41% in a Jordanian study population, and more than half of GIST cases in Saudi Arabian and Egyptian studies.[11,15] Moreover, one in five GIST patients in Saudi Arabian study had tumors in the small bowel.[15] Our study had stomach and small bowel as the most common site for gastrointestinal stromal tumor.

Although 30% of patients are asymptomatic, GIST associated symptoms varies with the site and size of the lesion, nearly half of the cases in the Egyptian study presented with gastrointestinal bleeding; followed by symptoms such as intestinal obstruction in nearly 30%, intraperitoneal hemorrhage in 15%, and rupture and peritonitis in 8%.[11] Abdominal pain was present in most of the patients (80%) in our study.

Tumor size is crucial in the progression of the disease. An Egyptian study on gastric stromal tumors which included 16 GIST patients reported tumor sizes between 8.4 and 20 cm.[16] In the present study, the median tumor size was 5cm ranging from 2.7to 10 cm. Our study also shows 72.72% of the cases are with tumor size greater than 5 cm. National Comprehensive Cancer Network (NCCN) suggested laparoscopic techniques should be used only in tumors less than 2 cm in size.[17] However, there are some reports indicating laparoscopy was effective and safe in removing larger GISTs but inadequate resection margins or tumor spillage leading to disease progression, recurrence and poor survival remain as main issues.[18]

CONCLUSION

Gastrointestinal stromal tumors are rare malignancies of the gastrointestinal tract. With the knowledge and understanding of the molecular biology of GIST, diagnosis and classification of the tumor is achieved. Surgical treatment remains the main stay management for the GIST, depending on the location, staging and metastasis. Laparoscopy should be the initial choice of surgery where metastasis is suspected. Immunohistochemistry is mandatory in defining GIST. After the surgery, patient should be referred to oncophysician and regular follow up of the patient should be taken. Six monthly CT scan should be done to assess the recurrence or metastasis and also to check the efficacy of the imatinib or sunatinib. Tumor size, location and mitotic index are the most important factors in predicting recurrence.

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REFERENCES

1. Kindblom LG, Remotti HE,Aldenborg Fand Meis-kindblom JM. Gastrointestinal pacemaker cell tumor (GIPACT). Gastrointestinal stromal tumors shows phenotypic characteristics of the interstitial cells of Cajal Am J Pathol. 1998;152:1259-69.
2. Judson I, Demetri G. Advances in the treatmentof gastrointestinal stromal tumours. Ann Oncol. 2007;18(Suppl 10):20–4.
3. Nishida T, Hirota S. Biological and clinicalreviewof stromal tumorsin the gastrointestinal tract. Histol Histopathol. 2000;15(4):1293-301.
4. Joensuu H. Gastrointestinal stromal tumor(GIST). Ann Oncol 2006;17(suppl 10):x280–6.
5. Ma GL, Murphy JD, Martinez ME, Sick lick JK. Epidemiology of Gastrointestinal stromal tumors in the era of histology codes: Results of a population-based study. Cancer Epidemiol Biomarkers Prev. 2015; 24(1):298–302.

6. Benesch M, Wardelmann E, Ferrari A, Brennan B, Verschuur A. Gastrointestinal stromal tumors (GIST) in children and adolescents: A comprehensive review of the current literature. Pediatr Blood Cancer. 2009;53(7):1171–9.
7. Fletcher CD, Berman JJ, Corless C. Diagnosis of gastrointestinal stromal tumors: a consensusapproach. Int J Surg Pathol. 2002;10(2):81–9.
8. Miettinen M, Majidi M, Lasota J. Pathology and diagnostic criteriaof gastrointestinal stromal tumors(GISTs): a review. Eur J Cancer. 2002;38(Suppl 5):S39–51.
9. Novelli M, Rossi S, Rodriguez-Justo M. DOG1 and CD117 are the antibodies of choice in the diagnosis of gastrointestinal stromal tumours. Histopathology. 2010;57(2):259–70.
10. Ando N, Goto H, Niwa Y. The diagnosis of GI stromal tumorswith EUS-guided fine needle aspirationwith immunohistochemical analysis. Gastrointest Endosc. 2002;55(1):37–43.
11. Sorour MA, Kassem MI, Ghazal AE. Gastrointestinal stromal tumors (GIST) related emergencies. Int J Surg. 2014;12(4):269-80.
12. Miettinen M, Lasota J, Review gastrointestinal stromal tumors: pathology and prognosis at different sites. Semin Diagn Pathol. 2006;23(2):70-83.
13. Wang ZH, Liang XB, Wang Y, Ma GL, Qu YQ, Tian XW. Epidemiology survey of gastrointestinal stromal tumor in Shanxi Province in 2011. Zhonghua yi xue za zhi. 2013;93(32):2541-4.
14. Barakat FH, Haddad HA, Matalka II, Al-Orjani MS, Al-Masri MM, Sughayer MA. Characteristics of gastrointestinal stromal tumors in a Middle Eastern population. Saudi Med J. 2010;31(7):797-802.
15. Bokhary RY, Al-Maghrabi JA. Gastrointestinal stromal tumors in western Saudi Arabia. Saudi Med. J. 2010;31:437-41.
16. Saied GM, Kensarah AM. Six months neoadjuvant imatinib improves resectability potential of gastric stromal tumors in Egyptian patients. Int J Surg. 2010;8(2):105-8.
17. Demetri GD, Benjamin RS, Blanke CD, Blay JY, Casali P, Choi H. management of patients with gastrointestinal stromal tumor (GIST)–update of the NCCN clinical practice guidelines. J Natl Compr Canc Netw. 2007;5(suppl 2):S1-29.
18. Desiderio J, Trastulli S, Cirocchi R, Boselli C, Noya G, Parisi A, Cavaliere D. Robotic gastric resection of large gastrointestinal stromal tumors. Int J Surg. 2013;11(2):191-6.

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