Review Article

Gastrointestinal Stromal Tumor – A Systemic Review Literature

Debajani Deka, Md Faizzal[1], Bipul Kumar Das[2]

Demonstrator, Gauhati Medical College and Hospital, Gauhati, Assam, [1]Senior Resident, All India Institute of Medical Sciences, Jodhpur, Rajasthan, [2]Assistant Professor, Tezpur Medical College and Hospital, Tezpur, Assam, India

Abstract

Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors of the gastrointestinal tract and originate from the interstitial cells of Cajal. They arise most commonly from the stomach or small intestine, with a median age of 60 years. We did not find any relationship between GIST and ABO blood group and Rh factor. Mutations in KIT exon 11 are found to be more common in larger tumors, and the presence of this mutation has been shown to have an adverse prognostic influence. Deletions compared with point mutations in exon 11 have also been found to be a significant unfavorable factor in patients with gastric GISTs. The study group includes all cases of GIST and extra-GIST. Review literatures were taken from Internet Google search and some websites of high index journals. The data extruded mainly were sites of the gastrointestinal tract, histological types, different immunohistochemical markers, etc., Stomach is the most common site of GIST around 70%. Spindle cell is the most common histological variety; CD117 is frequently found immunohistochemical finding. From the abovementioned results, we can easily come to the diagnosis of specific type of GIST and its respective therapeutic measures.

Keywords: Blood group, deletions, gastrointestinal, mutations, spindle

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Introduction

Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors of the gastrointestinal tract and originate from the interstitial cells of Cajal. They arise most commonly from the stomach or small intestine, with a median age of 60 years.[1] Majority of GISTs are sporadic, but rare familial cases may be found. GISTs are generally transmural tumors with frequent intraluminal and outward bulg.[2] The average size of extra-GIST (EGIST) (15.5 cm) was more than double of GIST (7.55 cm).[3] EGIST occurs most commonly in the retroperitoneum, liver, hepatobiliary system, spleen, pancreas, urinary bladder, prostate, and rectouterine septum.[4] The Cajal cells from which GISTs arise both produce ghrelin and express the ghrelin receptor.

Ghrelin is currently considered to be the main endogenous ligand of growth receptors. The ghrelin coding gene is located on chromosome 3 (3p25–26). Some of the many functions of ghrelin include regulation of growth hormone secretion, energy balance, gastrointestinal motility, gastric acid secretion, cardiovascular activity, pancreatic hormone secretion, glucose metabolism, prolactin and adrenocorticotropic hormone secretion, sleep, and gonadal hormone secretion. Several studies have shown that ghrelin can promote the development of malignant tumors through a variety of signaling pathways that increase cell proliferation and metastasis, including the phosphatidylinositol 3-kinase/AKT/mammalian target of rapamycin, Ras/RAF/extracellular signal-regulated kinases, Janus kinase/signal transducers and activators of transcription, and Src kinase pathways.[5] We did not find any relationship between GIST and ABO blood group and Rh factor.[6] Mutations in KIT exon 11 are found to be more common in larger tumors, and the presence of this mutation has been shown to have an adverse prognostic influence. Deletions compared with point mutations in exon 11 have also been found to be a significant unfavorable factor in patients with gastric GISTs.[7] Interstitial cells of Cajal, also known as pacemaker cells for peristaltic contraction, are a group of cells found in the muscularis propria and around the myenteric plexus along the GI tract and have the

Address for correspondence: Dr. Debajani Deka, C/o. P. C. Prafulla Chandra Deka, Bishnu Rabha Road, Tezpur, Assam, India.
E-mail: debajani.deka31@gmail.com

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Table 1: Most common site of gastrointestinal stromal tumors

Authors	Stomach (%)	Small intestine (%)	Large intestine (%)	Esophagus (%)	Appendix (%)
Huda T, Pratap, Singh M. (2019)[9]	40-60	5-30	15	-	-
Fangxing Peng, Yaolin (2020)	50-60	20-30	5-10	5	-
Binh LT, Mao NV, Huy TV, Tri NH, Khoan LT. (2020)[10]	60-70	20-30	2-5	1	-
E.N. Valdes Peregrinaa, M. Hernández Gonzáleza, b, O. de Leónb Pachecoc, S. Mendoza Ramírezа[11]	50	25-30	5	1	-
N Iqbal, A Sharma, Nk Shukla, BK Mohanti, SVS Deo, P Sahni, S Pal, S Pathy, V Raina, L Kumar[12]	19.6	42.8	19.6	-	-
R Zappacosta, B Zappacosta, S Capanna, C D'Angelo, D Gatta and S Rosini	60	30	5	5	5
Mohamad A, Quresi T Al, Rakha S M[13]	60-70	20-25	5	5	-
Dan Xu, MM, Xuyong Lin, Xueshan Qiu[16]	50-70	25-36	5-7	1-3	-
R Abuduwayite, A Muhemaiti, H Biekemituofu	60-70	25-35	5	2-3	5

Table 2: Most common age group effected

Authors	Age group
Klaus Kramer, Uwe Knippschild, Benjamin Mayer, Kira Bögelspacher, Hanno Spatz, Doris Henne-Bruns, Abbas Agaimy, Matthias Schwab and Michael Schmieder[19]	70 years, <40 years. (<10%) pediatric GIST
Nikki S. IJzerman, Cas Drabbe, Dide den Hollander, Mahmoud Mohammadi, Hester van Boven, Ingrid M. E. Desar, Hans Gelderblom, Dirk J. Grünhagen, An K. L., Reyners, Max M. van Noesel, Ron H. J., Mathijssen, Neeltje Steeghs and Winette T., A. van der Graaf[20]	65 years, <40 years (<10%)

GIST: Gastrointestinal stromal tumor

Table 3: Most common variety of gastrointestinal stromal tumors

Authors (%)	Spindle cell variety (%)	Epithelioid variety (%)	Mixed variety (%)
Wai Chin Foo, Bernadette Liegl, Atzwanger and Alexander J. Lazar[14]	70	20	10
Xiaohui Zhao, Changjun Yue[15]	70	20-25	5-10
Cicilia Marcella, RuiHua Shi and Shakeel Sarwar	70	20	10
Xu, Dan; Lin, Xuyong; Qiu, Xueshan[16]	70	20	10
Loiy Alkhatiba, Omar, Albtoushc, Nesreen, Batainehb, Kamal, Gharaibeha, Ismail, Matalka b, Yasuharu, Tokuda	70	20	10

immunophenotypic and ultrastructural characteristics of both the neural and smooth muscle elements.[8]

Materials and Methods

The study group includes all cases of GIST and EGIST. Review literatures were taken from Internet Google search and some websites of high index journals. The data extruded mainly were sites of the gastrointestinal tract, histological types, different immunohistochemical markers, etc.[9]

Aims and objectives

The aim of the study is analysis and identification of different varieties of GIST through histology and immunohistochemical markers.

Objectives

1. Most frequent site of GIST
2. Different immunohistochemical markers of GIST
3. Different histological types of GIST
4. Most common age group effected.

Results

From the Table 1 it has been seen that stomach is the most commonly effected site of GIST (40%-70%) while esophagus and appendix(2%-5%) are least commonly effected site . From Table 2 it has been observed that age <40yrs and >60 yrs are effected most and regarding pediatric age group it is <10yrs yrs. From Table 3 we found spindle variety as most common variety(70%) of GIST [Table 4]. Immunomarker CD117, DOG1 and S100 frequently found in GIST.In majority of cases they are positive.

Discussion

From the above literatures, it has been found that GIST is mainly found in the stomach followed by small intestine and this is according to the authors R. J. Hartley, j. H. R. Becker, and h. Van der walt. The age group commonly effected in GIST is <40 years and >70 years which is coinciding with the authors Kjetil Sreidea, b, Oddvar M. Sandvika, Jon Arne Sreidea, b, Vanja Giljacac, Andrea Jureckovad, and V. Ramesh Bulusu.

From the above literatures, it has been found that spindle cell variety is the most common variety of GIST. This is also according to the authors Albtoushc, Nesreen Batainehb, Kamal Gharaibeha, Ismail Matalka b, and Yasuharu Tokuda.

Table 4: Most common immunomarker

Authors	Immunomarkers	Remarks
Sonja E Steigen, Bodil Bjerkehagen, Hans K Haugland, Ivar S Nordrum, Else Marit Løberg, Vidar Isaksen, Tor J Eide and Torsten O Nielsen[7]	CD117	+
	Smooth muscle actin	+
	S100	+
	Desmin	+
		+
		+
Michael Stamatakos, Emmanouel Douzinas, Charikleia Stefanaki, Panagiotis Safioleas, Electra Polyzou, Georgia Levidou and Michael Safioleas[8]	CD117	+
	DOG1	+
		+
Vandana L. Gaopande, Avinash R. Joshi, Pallavi D. Bhayekar, Siddhi G. S., Khandeparkar[17]	CD117	+
	S100	+
	DOG1	+
		+
lei Zhou, Yusheng liao, Jie Wu Jing Yang heng Zhang Xiangyang Wang shengbin sun[18]	CD117	+
	DOG1	+
	S100	+
	Smooth muscle actin	+
	Desmin	+
		+

CD117 is the most common immunohistochemical marker associated with GIST which is according to the authors Yu Tung Lo, David Siu Kei Mak, and Colum Patrick Nolan.

Conclusion

From the abovementioned results, we can easily come to the diagnosis of specific type of GIST and its respective therapeutic measures.

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Conflicts of interest

There are no conflicts of interest.

References

1. Kale SS, Sachdev MS, Ismail MK, Davila R, Tombazzi CR. A case and literature review of complicated gastrointestinal stromal tumors. Gastroenterol Hepatol (N Y) 2008;4:650-7.
2. Niazi AK, Kaley K, Saif MW. Gastrointestinal stromal tumor of colon: A case report and review of literature. Anticancer Res 2014;34:2547-50.
3. Gaopande VL, Joshi AR, Bhayekar PD, Khandeparkar SG. Clinicopathologic and immunohistochemical study of gastrointestinal stromal tumor (ten cases) and extragastrointestinal stromal tumor (six cases) with review of literature. J Curr Res Sci Med 2016;2:84-91.
4. Katsoulis IE, Tzortzopoulou A, Tziakou P, Arnogiannaki N, Kostoglou-Athanassiou I, Lypas G, *et al.* Extragastrointestinal stromal tumour of the lesser omentum: A case report and literature review. Int J Surg Case Rep 2017;37:17-21.
5. Brodsky SV, Gimenez C, Ghosh C, Melamed M, Ramaswam G. Estrogen and progesterone receptors expression in gastrointestinal stromal tumour intramural gastrointestinal Leimyoma. J Gastrointest Cancer 2006;37:129-32.
6. Ürün Y, Utkan G, Yalcin Ş, Coşkun HŞ, Koçer M, Özdemir NY, *et al.* Lack of any relationship between ABO and Rh blood groups and clinicopathological features in patients with gastrointestinal stromal tumors: Turkish oncology group. Asian Pac J Cancer Prev 2012;13:4129-31.
7. Steigen SE, Bjerkehagen B, Haugland HK, Nordrum IS, Løberg EM, Isaksen V, *et al.* Diagnostic and prognostic markers for gastrointestinal stromal tumors in Norway. Mod Pathol 2008;21:46-53.
8. Stamatakos M, Douzinas E, Stefanaki C, Safioleas P, Polyzou E, Levidou G, *et al*. Gastrointestinal stromal tumor. World J Surg Oncol 2009;7:61.
9. Huda T, Singh MP. Gastrointestinal stromal tumors of small intestine. Surg J (N Y) 2019;5:e92-5.
10. Binh LT, Mao NV, Huy TV, Tri NH, Khoan LT. Early diagnosis and treatment of a small gastric stromal Tumor – A case report and literature review. Asp Biomed Clin Case Rep 2020;3:135-40.
11. Valdes-Peregrinaa EN, Hernández-González M, De León-Pachecoc O, Ramírez SM. Extra-gastrointestinal stromal tumour. Report of primary tumour in the omentum. Rev Med Hospgenmex 2018;81:221-5.
12. Iqbal N, Sharma A, Shukla N, Mohanti BK, Deo SV, Sahni P, *et al*. Advanced gastrointestinal stromal tumors: 10-years experience from a tertiary care centre. Trop Gastroenterol 2015;36:168-73.
13. Mohamed A, Al Qureshi T, Rakha SM. Giant gastrointestinal stromal tumors of the stomach successfully treated with laparoscopic resection: Case report and literature review. Cureus 2021;13:e13584.
14. Foo WC, Liegl-Atzwanger B, Lazar AJ. Pathology of gastrointestinal stromal tumors. Clin Med Insights Pathol 2012;5:23-33.
15. Zhao X, Yue C. Gastrointestinal stromal tumor. J Gastrointest Oncol 2012;3:189-208.
16. Xu D, Lin X, Qiu X. The epithelioid gastrointestinal stromal tumor with pulmonary metastasis: A rare case report and literature review. Medicine (Baltimore) 2020;99:e19346.
17. Gaopande VL, Joshi AR, Bhayekar PD, Khandeparkar SG. Clinicopathologic and immunohistochemical study of gastrointestinal stromal tumor (ten cases) and extragastrointestinal stromal tumor (six cases) with review of literature. J Curr Res Sci Med 2016;2.
18. Zhou L, Liao Y, Wu J, Yang J, Zhang H, Wang X, *et al*. Small bowel gastrointestinal stromal tumor: A retrospective study of 32 cases at a single center and review of the literature. Ther Clin Risk Manag 2018;14:1467-81.
19. Kramer K, Knippschild U, Mayer B, Bögelspacher K, Spatz H, Henne-Bruns D, *et al*. Impact of age and gender on tumor related prognosis in gastrointestinal stromal tumors (GIST). BMC Cancer 2015;15:57.
20. IJzerman NS, Drabbe C, den Hollander D, Mohammadi M, van Boven H, Desar IM, *et al.* Gastrointestinal stromal tumours (GIST) in young adult (18-40 Years) patients: A report from the Dutch GIST registry. Cancers (Basel) 2020;12:730.