International Journal of Surgery and Research (IJSR)
ISSN: 2379-156X

Gastrointestinal Stromal Tumors in Al-Noor Specialist Hospital, Makah, Saudi Arabia: Report of Five Cases

Research Article

Suleiman Jastaniah*

Department of General Surgery, Faculty of Medicine, Umm Al-Qura University, Makah, Saudi Arabia.
Department of General Surgery, Al Noor Specialist Hospital, Saudi Arabia.

Abstract

Gastrointestinal stromal tumors (GISTs) are rare, but they are the most common mesenchymal neoplasm of the GI tract. GISTs are usually found in the stomach or small intestine but can occur anywhere along the GI tract and rarely have extra-GI involvement. Five cases of (GIST) are recorded between Jan 2017 and December 2019 in the surgical department in Al Noor Specialist Hospital. The records of the five cases of GI stromal tumor were reviewed. Three males and two females, with mean age of 55.8 ± 7years. The stomach was involved in all patients. The most common symptom at presentation was abdominal pain, followed by gastrointestinal bleeding. The size of the tumors varies between 3.5X4 cm to 21.7X10.8cm. Four cases were treated with the complete surgical resection and the large tumor was treated medically. Follow up of the patients for 2-years. there was no recurrence in four cases. The fifth case with large tumor treated medically died after 18 months.

Complete surgical resection with tumor free margin is the standard treatment for GISTs [1].

Keywords: Gastrointestinal (GIST) - Complete Surgical Excision - Spindle Cells - CD117.

Introduction

Gastrointestinal stromal tumors (GISTs) are rare but considered the most common mesenchymal tumors of the gastrointestinal tract [1, 2]. Nevertheless they account only 0.1%-3% of all malignancies [2-4]. In the United states, an annual incidence of 3300-6000 new cases has been reported while in Saudi Arabia the annual incidence has increased from 2.1 per million in 1995 to 12.7 per million in 2003 [5, 6]. The cause for this is to increased understanding of GIST pathology and the availability of the diagnostic immuno histochemical antibody directed against the CD 117 antigen [6]. The exact incidence rate is unknown and the true incidence may be much higher ,as micro-GIST lesions (less than 1cm) may be present without clinical evidence [7, 8]. GISTs can occur anywhere in the gastrointestinal tract but is the most common in the stomach (60%) and small intestine (30%), but it can occur anywhere in the gastrointestinal tract [9]. Complete surgical resection followed by imatinib treatment is the treatment of choice for these tumors [10].

A Case series of five patients diagnosed with GISTs is presented, four of them complete surgical resection was achieved and the fifth case was treated with medical treatment (imatinib) followed by surgical resection.

Case Report

Report of 5 cases

Five cases diagnosed with GISTs were admitted to the surgical department between Jan 2017and December 2019. Four of them treated with complete surgical resection and the fifth one treated with medical treatment followed by complete resection at Al-Noor Specialist hospital (Makah, Kingdom of Saudi Arabia). Three patients were males (60%) and two patients were females (40%). The mean age of 55.8 ± 7 years (range 48-70 years). The

***Corresponding Author**:
Suleiman Jastaniah,
Department of General Surgery, Faculty of Medicine, Umm Al-Qura University, Makah, Saudi Arabia.
Department of General Surgery, Al Noor Specialist Hospital, Saudi Arabia.
Tel: 966505539003
Fax: 966125544900
Email: sa.jastaniah@hotmail.com

Received: February 11, 2020
Accepted: February 26, 2020
Published: February 29, 2020

Citation: Suleiman Jastaniah. Gastrointestinal Stromal Tumors in Al-Noor Specialist Hospital, Makah, Saudi Arabia: Report of Five Cases. *Int J Surg Res.* 2020;7(2):146-148.
doi: http://dx.doi.org/10.19070/2379-156X-2000031

stomach is the site of the lesion in all cases (100%) in addition that the fifth case there was liver metastasis. All five patients underwent CT-scan abdomen with oral and IV contrast and upper endoscopy with biopsy. The reports of CT-scan and the biopsies are highly suggestive of GIST. Blood investigations for all 5 patients were normal. Pain, upper gastrointestinal bleeding and weight loss were the most common presenting symptoms. Complete surgical resection of the tumor was done in four patients and the fifth patient after medical treatment with imatinib. All patients have histologically negative margins. Histological examination of all specimens revealed mixed spindle and epithelioid cells in four patients (80%) and spindle cells in one patient. Four patients have CD117 and CD34 positive while one patient has CD34 positive (20%).The resected tumors in three patients were subtyped as gastrointestinal stromal tumors with low risk of progressive disease less than 5/50 mitosis per HPF, one patient not mentioned and the fifth patient was classified as GIST with high risk more than 25/50 mitosis per HPF. The patient with high risk received Imatinib 400 mg daily for eighteen months. The follow up was for two years. Four patients remained free of the disease.

Discussion

In the early literatures the Gastrointestinal Stromal Tumors (GISTs) as mesenchymal tumors involving the wall of the bowel [6]. In 1983 Mazur and Clark identified GIST as mesenchymal tumors of the gut, arising from the myenteric nervous system [11, 12]. In 1998 Hirota reported that GISTs contained activating mutations in the cKit proto-oncogene, associated with expression of Kit protein known as CD117 [13]. GISTs tumors have no sex preference and common between the age 50 and 80 years [14]. The clinical presentations are different according to the site and the size as well as its aggressiveness [15]. The most common clinical presentation is gastrointestinal bleeding, other symptoms may exist as a cute abdominal pain, intestinal obstruction and small tumors may remain a symptomatic [6-8]. In this series all five patients with GISTs tumors were clinically evident and presented with gastrointestinal bleeding.

GISTs are positive for CD117 in 95%of cases, and 70-80% of the tumors are positive for CD34 expression [16, 17]. All patients in this series the lesions were positive for the expression of these markers.

The stomach is the most common site involved (60%), then the small intestine (30%), the rectum (3%), colon (1-2%) and esophagus (<1%) [11]. In present study the stomach was the affected organ.

CT scan is the best modality of the diagnosis and follow up. The most common CT feature is a heterogeneously enhancing mass arising from the intestinal wall and protruding into the abdominal cavity, with central hypodense non enhancing areas of necrosis and central degeneration [18, 19]. In this study CT scan was the primary diagnostic and follow up tool. Upper endoscopy can suggest the diagnosis due to the presence of intact mucosa over the mass, most of the biopsies are not adequate or not conclusive, as seen in the present study where it was performed in all five cases only one was diagnostic.

The important part of the management of GISTs is to assess the risk factors. It is also important for its prognostic and therapeutic implications. According to the Armed Forces Institute of Pathology criteria, the main factors involved are the anatomic site of the tumor, its size, and mitotic index [23-25].

Complete surgical resection is the standard treatment for localized GIST, with the use of Tyrosine Kinase Inhibitors (TKIs) as adjuvant treatment in high risk patients [25-27]. TKIs are also the standard treatment for unresectable, recurrent, and metastatic GISTs, with surgery for selected cases [20-23]. Laparoscopic surgical resection can be done in selected cases but tumor rupture will increase the recurrence risk [24]. In the present series complete surgical resection was achieved in four patients with no recurrence was detected with follow up to 2 years, and TKIs was used for the fifth case for 18 months but the patient died because of sepsis.

Conclusion

In conclusion complete surgical resection with negative margins is the standard treatment for GISTs, and TKIs either alone or in sequence are as adjuvant therapy if the risk of progressive disease is high. The role of laparoscopic surgery in GISTs needs more research.

References

[1]. Al-Jiffry BO, Allam HM, Hatem M. Single-center experience of surgically resected gastrointestinal stromal tumors: A report of six cases, including a rare case involving the loweresophagus.Oncol Lett. 2015 Feb;9(2):745-748. Pubmed PMID: 25624901.

[2]. Zhang L, Smyrk TC, Young WF Jr., Stratakis CA, Carney JA. Gastric stromal tumors in carney triad are different clinically, pathologically, and behaviorally from sporadic gastric gastrointestinal stromal tumors: Findings in 104 cases. Am J Surg Pathol. 2010 Jan;34(1):53-64. Pubmed PMID: 19935059.

[3]. Joensuu H, Hohenberger P, Corless CL. Gastrointestinal stromal tumour Lancet. 2013 Sep 14;382(9896):973-83. Pubmed PMID: 23623056.

[4]. Corless CL ,Heinrich MC. Molecular pathobiology of gastrointestinal stromal sarcomas Annu Rev Pathol. 2008;3:557-86. Pubmed PMID: 18039140.

[5]. Corless CL. Gastrointestinal stromal tumors: What do we know now?. Mod Pathol. 2014 Jan;27 Suppl 1:S1-16. Pubmed PMID: 24384849.

[6]. Al Hussaini HF. GIST in Saudi Arabia: multicentric histopathological genetic study of 75 surgically excised cases. Gulf J Oncolog. 2012 Jan;(11):31-7. Pubmed PMID: 22227543.

[7]. Agaimy A, Wünsch PH, Hofstaedter F, Blaszyk H, Rümmele P, Gaumann A, et al. Minute gastric sclerosing stromal tumors (GIST tumorlets) are common in adults and frequently show c-KIT mutations. Am J Surg Pathol. 2007 Jan;31(1):113-20. Pubmed PMID: 17197927.

[8]. Kawanowa K, Sakuma Y, Sakurai S, Hishima T, Iwasaki Y, Saito K, et al. High incidence of microscopic gastrointestinal stromal tumors in the stomach. Hum Pathol. 2006 Dec;37(12):1527-35. Pubmed PMID: 16996566.

[9]. Miettinen M, Lasota J. Histopathology of gastrointestinal stromal tumor. J Surg Oncol. 2011 Dec 15;104(8):865-73.

[10]. Blackstein ME, Blay JY, Corless C, Driman DK, Riddell R, Soulières D, et al. Canadian Advisory Committee on GIST. Gastrointestinal stromal tumours: consensus statement on diagnosis and treatment. Can J Gastroenterol. 2006 Mar;20(3):157-63. Pubmed PMID: 16550259.

[11]. Mazur MT, Clark HB. Gastric stromal tumors. Reappraisal of histogenesis. Am J Surg Pathol. 1983 Sep;7(6):507-19. Pubmed PMID: 6625048.

[12]. Rabie ME, Hummadi A, Bazeed M, El Hakeem I, Al Qahtani AS, Haroon H, et al. Gastrointestinal stromal tumors: Do we follow the current guidelines? A self-critique. Saudi Surgical Journal. 2019 Jul 1;7(3):100-107.

[13]. Hirota S, Isozaki K, Moriyama Y, Hashimoto K, Nishida T, Ishiguro S. Gain of function mutations of c-kit in human gastrointestinal stromal tumors. Science. 1998 Jan 23; 279(5350): 577-80. PMID: 9438854.

[14]. Nowain A, Bhakta H, Pais S, Kanel G, Verma S. Gastrointestinal stromal tumors: Clinical profile, pathogenesis, treatment strategies and prognosis. J Gastroenterol Hepatol. 2005 Jun; 20(6): 818-24. PMID: 15946127.

[15]. Demetri GD. Gastrointestinal stromal tumor. 9th ed. DeVita VT Jr., Lawrence TS, Rosenberg SA, editors. Cancer: Principles and Practice of Oncology. Philadelphia, Pa: Lippincott Williams & Wilkins; 2011
[16]. Miettinen M, Lasota J. Gastrointestinal stromal tumors - definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. Virchows Arch. 2001 Jan;438(1):1-12. PMID: 11213830.
[17]. Kindblom LG, Remotti HE, Aldenborg F, Meis-Kindblom JM. Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. Am J Pathol. 1998 May; 152(5): 1259-69. PMID: 9588894.
[18]. Catalano O, De Lutio di Castelguidone E, Nunziata A, De Rosa V, Siani A. Gastrointestinal stromal tumors: A pictorial review. Radiol Med. 2005 Nov-Dec;110(5-6):484-91.. Pubmed PMID: 16437034.
[19]. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: Radiologic features with pathologic correlation. Radiographics. 2003 Mar-Apr;23(2):283-304, Pubmed PMID: 12640147.
[20]. Joensuu H, Hohenberger P, Corless CL. Gastrointestinal stromal tumour. Lancet. 2013 Sep 14;382(9896):973-83. Pubmed PMID: 23623056.
[21]. Miettinen M, Sobin LH, Lasota J. Gastrointestinal stromal tumors of the stomach: A clinicopathologic, immunohistochemical, and molecular genetic study of 1765 cases with long-term follow-up. Am J Surg Pathol. 2005 Jan;29(1):52-68. Pubmed PMID: 15613856.
[22]. Miettinen M, Furlong M, Sarlomo-Rikala M, Burke A, Sobin LH, Lasota J. Gastrointestinal stromal tumors, intramural leiomyomas, and leiomyosarcomas in the rectum and anus: A clinicopathologic, immunohistochemical, and molecular genetic study of 144 cases. Am J Surg Pathol. 2001 Sep;25(9):1121-33. Pubmed PMID: 11688571.
[23]. Nishida T, Doi T, Naito Y. Tyrosine kinase inhibitors in the treatment of unresectable or metastatic gastrointestinal stromal tumors. Expert Opin Pharmacother. 2014 Oct;15(14):1979-89. Pubmed PMID: 24990162.
[24]. Ohtani H, Maeda K, Noda E, Nagahara H, Shibutani M, Ohira M, et al. Meta-analysis of laparoscopic and open surgery for gastrointestinal stromal tumor. Anticancer Res. 2013 Nov;33(11):5031-41.Pubmed PMID: 24222147.
[25]. National Comprehensive Cancer Network. NCCN clinical practice guidelines in oncology. Soft tissue sarcoma. 2; 2018. Available from: https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf.
[26]. Judson I, Bulusu R, Seddon B, Dangoor A, Wong N, Mudan S. UK clinical practice guidelines for the management of gastrointestinal stromal tumours (GIST). Clin Sarcoma Res. 2017 Apr 21;7:6. Pubmed PMID: 28465823.
[27]. Casali PG, Abecassis N, Bauer S, Biagini R, Bielack S, Bonvalot S, et al. Gastrointestinal stromal tumours: ESMO-EURACAN clinical practice guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2018 Oct 1;29(Suppl 4):iv68-iv78. Pubmed PMID: 29846513.