Introduction

Gastrointestinal stromal tumors (GISTs) are rare but considered the most common mesenchymal tumors of the gastrointestinal tract [1, 2]. Nevertheless, they account for 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 an 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 1 cm) 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 2017 and 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
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.

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