

Duodenal Gist in an Elderly Female - A Rare Case Report

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Abstract: *The incidence of Gastrointestinal stromal tumors is most in stomach which accounts for 50% of all the cases. Most often they are asymptomatic and are recognised when either the mucosa overlying ulcerate leading to bleeding or are noticed incidentally on CT scan or EUS. When large it may present with nonspecific gastrointestinal symptoms or as an abdominal lump. Diagnosis can be confirmed by a CECT of abdomen with final diagnosis relying on the histopathology examination of specimen. Definitive treatment includes wide local excision with adjuvant tyrosine kinase inhibitors. Here we are reporting a case of GIST arising from duodenum in a 60year old female presenting with hypovolemic shock secondary to hematemesis and melena and treated by resection of mass. Histopathology repeat revealed the diagnosis as GIST.*

Keywords: GIST, Surgical resection, tyrosine kinase inhibitor

1. Introduction

Gastrointestinal stromal tumors are nonepithelial mesenchymal tumor that are slow growing arising from interstitial cells of Cajal (ICC) and are distinct from leiomyoma and leiomyosarcoma. Two third of all GISTs occur in the stomach. Epithelial cell stromal GIST is the most common type and cellular spindle type next common. Glomus tumor type is only seen in stomach. Almost all the GIST express c-KIT(CD117), CD34, PDGF receptors and actin, desmin. These are useful in differentiating between GIST and smooth muscle tumors histologically. Smaller lesions are incidental to find although occasionally may ulcerate, bleed while larger lesions cause weight loss, abdominal pain, fullness, bleeding. Mass may sometimes be palpable. Hematogenous metastasis and lymph node metastasis are seen rarely. Diagnosis is by endoscopy guided biopsy or EUS. Metastatic workup entails CT chest, abdomen and pelvis. Prognosis of patient depends mostly on tumor size, mitotic count and metastasis. Complete surgical resection with negative margins is recommended. Treatment with imatinibmesylate has shown improvement in management of unresectable or metastatic disease. In this case report we described a 60year old female with duodenal GIST treated with complete surgical resection.

2. Case Report

A 60year old female patient presented to the emergency department with chief complaints of multiple episodes of hematemesis and melena since 6-7 days. She had no history of similar complaint in past. No history of abdominal distension, bowel or bladder complaints. No past surgical or medical history. She was in her menopause and a mother of 3 children delivered by normal vaginal delivery.

She had a history that she got such episodes 6-7 days back for which she went to a nearby private hospital and was diagnosed to be in severe hypovolemia secondary to blood

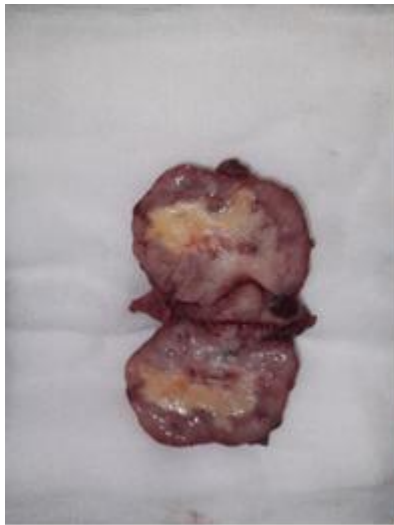
loss and was primarily investigated. 12 units blood transfusion were given to the patient meanwhile radiology investigation in the form of USG abdomen was done with suggested Large solid heterogeneous lymph node mass lesion over right suprarenal area possibly periportal lymph node with distorted hilum and changes of colitis in sigmoid colon. Following the USG findings a CECT abdomen and pelvis was done which gave inference of 44*55*51mm sized lobulated exophytic soft tissue density mass lesion with marked inhomogeneous enhancement arising from second part of duodenum with intraluminal extension with multiple prominent vessels seen around the surface of mass. Superiorly the mass was seen abutting the inferior surface of liver and the planes with right kidney and large bowel maintained.

With this report patient was referred for further intervention to our hospital. On arrival in emergency department she had a blood pressure of 60mmhg systolic in brachial artery and tachycardia of 110/min on inotropic support of noradrenaline and dopamine. Per abdomen was soft and revealed a diffuse lump on deep palpation in right lumbar region. She was immediately shifted towards and resuscitation started first by inserting a central venous access and measuring central pressure and accordingly continuing inotropic support and fluids. 6 blood and 16 frozen plasma were transfused in a period of 15 days with gradual improvement in vital status of patient and no episodes of blood loss.

Based on the CT findings an upper GI scopy was planned which revealed normal mucosa of duodenum. Following this a CT angiography was planned with the suspicion of GIST not being the cause of massive exsanguination which revealed Malignant GIST arising from duodenum with feeder vessel arising from gastroduodenal artery. Following all of this a laparotomy was planned and on table a 5*5cm sized highly vascular exophytic lesion was seen arising from first part of duodenum. Feeder were tied and mass excised

using linear cutter stapler and sent for histopathology examination. Report was suggestive of GIST with positive staining for CD117. Patient was stable postoperatively and

discharged on day 12 on Imatinibmesylate (400mg) for 3months.



3. Discussion

Gastrointestinal stromal tumor (GIST) is a nonepithelial, mesenchymal tumor first described by Mazur and Clark in 1983. GISTs are thought to arise from interstitial cells of Cajal (ICC) or their stem cell precursors which are normally part of the autonomic nervous system of the intestine [2, 3]. ICC serves as a pacemaker function in controlling motility. GISTs usually arise in the stomach in 40% to 70%, in the small intestine in 20% to 40%, and less than 10% in the esophagus, colon, and rectum [3–6]. Duodenal gastrointestinal stromal tumor (DGIST) accounted for 10–30% of all malignant tumors of the duodenum. GISTs can develop outside the intestinal tract, within the abdominopelvic cavity such as the omentum, mesentery, uterus, and the retroperitoneum; they are called extra gastrointestinal stromal tumors (eGIST), usually behaving aggressively [2, 4, 7, 8]. GIST has been shown to affect men (55%) more than women with median age of 55–60 [9].

The spectrum of clinical presentation in GIST is broad. It is largely dependent on tumor size and location. GIST-causing symptoms are usually larger in size, more than 6 cm in diameter [9]. The most common presentation of GIST is abdominal pain and/or GI bleeding [2]. This may be acute, as in melena, hematemesis, or chronic insidious bleeding leading to anemia [2]. GIST can also cause symptoms secondary to mass effect, including satiety, bloating, and abdominal pain [2, 9]. In our case review, patient has presented in a state of hypovolemic shock. Approximately 20% to 25% of gastric and 40% to 50% of small intestinal GISTs are clinically malignant. The most common metastatic sites include the abdominal cavity, liver, and rarely bones and soft tissues. GISTs very rarely, if not, metastasize to the lymph nodes and the skin. Less than 5% of GISTs can be associated with one of the four tumor syndromes: familial GISTs, neurofibromatosis type 1 (NF1), Carney's triad (CT), and, recently, the Carney-Stratakis triad (CSS) [4].

GISTs normally present a wide clinical-pathological spectrum, from a small incidental nodule to large pedunculated mass. They are usually described as a tan to white, well-circumscribed lesions within the walls of the stomach [4]. GISTs demonstrate either of the 3 main histologic cell types: spindle cell type (most common), epithelioid cell type, and the mixed spindle-epithelioid type [3–5, 9]. Spindle cell GISTs account for 70% of the tumors.

Diagnosis of GISTs relies heavily on KIT/CD117 immunohistochemical staining, which can detect most GISTs except for a few 3% to 5% that harbors PDGFRA mutation. Tumor size, location, and mitotic index remain the main variables used in risk stratification systems first developed by the National Institute of health, the so-called Fletcher's criteria [4, 5]. The first step in the management of a potentially resectable GIST is to determine its resectability with history/physical exam together with tests such as computed tomography (CT) and/or magnetic resonance imaging (MRI), chest imaging, endoscopic ultrasound (EUS), and endoscopy. PET scan is not routinely recommended [5].

Despite the impressive advances in targeted therapy, surgery resection remains the primary mode of therapy for localized GISTs [4, 5]. Surgery is used in three main approaches, most commonly as an initial treatment (primary surgery) after diagnosis, especially if the tumor is solitary and can be easily removed. It can be used after neoadjuvant treatment to shrink the size of the tumor; and, in some cases, surgery is used for advanced metastatic disease for symptomatic relief, termed debulking surgery. These tumors should be handled carefully to avoid tumor rupture and spread. GISTs respond poorly to conventional chemotherapy and radiation therapy [4]. Imatinibmesylate and sunitinib maleate are competitive inhibitors of KIT and PDGFRA [4, 10] and are used as adjuvant treatment post resection of GIST. For tumors of large size, imatinib has proved its utility in downgrading the size to allow surgical excision at a later date and also preservation of function of the organ.

4. Conclusion

GIST is a tumor with growing concern. The diagnosis of GIST is often delayed owing to its indolent symptoms that only present in advance and sometimes unresectable stage. Immunohistochemical staining is a useful aid in diagnosing GISTs. Surgery still remains as the primary mode of treatment despite a high incidence of recurrence, owing to the lack of alternative treatment options. Improvement in surgical techniques has decreased the incidence of tumor recurrence from tumor seeding. Postoperative imatinib treatment has also shown to improve relapse-free survival.

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