Caecal Gastrointestinal Stromal Tumor with Perforation and Obstruction

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Introduction

The incidence of GIST is 11–15 patients/million/year and they constitute 0.1 % to 3 % of all gastrointestinal malignancy [2]. They commonly originate in the stomach, followed by small intestine [1–4]. They rarely arise in the caecum, with the entire colon and rectum contributing to less than 5 % of all GISTs [1–4]. Anecdotal reports of small intestinal GIST presenting with perforation exist in the literature [5]. Caecal GIST presenting with perforation has been reported only in canines but not in humans [6]. GIST arising in the small intestine usually bleeds as opposed to carcinoids, which present with obstruction [7].

Material and Method

A 60 years old female, presented with severe abdominal pain of 5 days duration. It was initially felt in the right iliac fossa, then became diffuse and severe. It was associated with frequent passage of small amounts of bright red blood per rectum, with no stool accompanying it. Immediately after onset of pain abdomen she had developed repeated episodes of large amounts of bilious vomiting lasting for 2 days. She also complained of obstipation for a day prior to admission. There was a past history of episodic mild bleeding per rectum, but without any bowel disturbances, 3 months ago.

On physical examination, the Patient was in acute distress with severe abdominal pain. She had a toxic look,

Febrile at 100-F, with Pulse of 104/min, respiratory rate of 30/min and was normotensive. Abdominal examination revealed a tender and huge, 15×18 cm mass, occupying the right iliac and lumbar areas with diffuse margins. The rest of the abdomen was soft but mildly tender. Bowel sounds were loud. On per rectal examination the finger stall was stained with bright red blood and mucus.

The Haemoglobin was 9 gms%. Total leucocyte count was 15,600/cumm with neutrophils being 90 %. x-ray abdomen revealed dilated small intestinal loops but no free gas under diaphragm (Fig. 1). CT scan of the abdomen revealed a huge tumor with areas of necrosis replacing the caecum and with moderate amounts of free fluid in the peritoneal cavity (Fig. 2).

Patient underwent an emergency laparotomy, after a rapid resuscitation. A huge localized tumor of 15×20 cms was found occupying the caecum, with a perforation of the anterior wall. The perforation had been sealed by greater omentum and by loops of distended discolored terminal ileum. Once these adhesions were released a foul smell emanated from the site of perforation. Moderate amounts of turbid fluid were found in the peritoneal cavity which was sent for culture and sensitivity. Multiple small and soft lymph nodes were found in the mesosolon. A right radical hemicolectomy, omentectomy and peritoneal lavage was done with a diagnosis of perforated carcinoma of caecum (Fig. 3). The patient recovered well postoperatively.

Histopathology of the resected specimen revealed a high-grade caecal GIST with more than 5 mitosis per 50 High power fields and c-kit being positive (Fig. 4). Culture of peritoneal fluid did not grow any micro organisms. Patient was administered imatinib (gleevec) postoperatively, as adjuvant therapy. Patient was followed up for 3 years, during which she was found to be disease free, before being lost to follow-up.

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Fig. 1 X-ray abdomen (erect posture) showing no air under domes of diaphragm

Discussion

Gastrointestinal stromal tumors are mesenchymal tumors of the Gastrointestinal tract with spindle cell or epithelioid histology and unequivocal immunohistochemical C-kit positivity. They arise from the interstitial cells of Cajal and they account for 0.1 % to 3 % of all G.I malignancies [2]. The site distribution of these tumours is as follows: Esophagus (3 %–5 %), stomach (60 %), small intestine (25 %–30 %), colon and rectum (1 %–5 %), others (2 %–6 %) [1–3]. Gastrointestinal stromal tumors of the small intestine presenting with perforation can be found as isolated reports [5, 8]. Perforated GIST is associated with reduced survival and adjuvant therapy with imatinib is recommended [5, 8]. In a study looking at specificity of clinical presentation of small intestinal tumors, it was reported that GIST mostly bleed, Carcinoids produce obstruction and Lymphomas cause perforation [7]. The commonest presentation of GIST in approximately 50 % of cases is bleeding [2]. It can also present as mass abdomen and in the small intestine, rarely with intestinal obstruction.

Fig. 2 CECT abdomen showing caecal tumor with heterogeneity and moderate amounts of free fluid in the abdomen

Fig. 3 Caecal tumor with perforation and mobilization of colon for right radical hemicolectomy

Till date no perforated caecal GIST in humans has been reported in the literature. Perforation of caecum secondary to GIST of splenic flexure has been reported [9]. A combination of perforation, obstruction and GI bleeding in a single patient is also not reported. According to the risk assessment proposed by Fletcher et al our patient displayed high risk features (Tumor size above 10 cms and >5 mitoses per 50 high power fields) [10]. Rutkowski et al indicated that tumors of size more than 10 cms and mitotic index of >5 mitosis per 50 high power fields had a metastatic rate as high as 84 %. Miettinen et al predicted a metastatic rate of 2 % to 3 % in those with tumors of size less than 10 cms and mitotic index of less than 5 mitosis per 50 HPF [11, 12]. In operable cases, the tumor should be removed en bloc with a clear margin. But how much should be the length of margin has not been clearly defined. The pseudo capsule should be removed and not penetrated. A segmental resection of colonic GIST can be done. If adjacent structures are involved they should be removed en bloc. Sleeve resection is advocated in small tumors of the stomach. The lymph node dissection is not recommended. Changchien CR et al found

Fig. 4 High grade caecal GIST with >5 mitosis per 50 high power fields

that the radical resection like abdomino-perineal resection or anterior resection of rectum in cases of malignant rectal GIST was superior to local excision with clear margins in the prevention of local recurrence but not for distant metastasis [13].

In a study involving 200 cases of GIST, the complete resection rate was approximately 85 %. The median survival was 66 months if complete resection had been done as compared to 22 months in those with incomplete resection[14]. The Survival with R-O resection of Gastrointestinal stromal tumors is 45 % in a localized tumor but 65.2 % with the use of adjuvant therapy after resection, at 36 months [15]. The indication for adjuvant therapy in our patient was the size of the tumor which was more than 10 cms, the presence of perforation and mitotic figure of more than 5 per 50 high power fields.

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