Atypical Presentation of Gastrointestinal Stromal Tumours—A Case Report

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Abstract Gastrointestinal stromal tumors (GISTs) are benign mesenchymal tumors of the gastrointestinal tract (GIT). Their clinical presentations are variable. We report a case of a 31-year-old man who presented with pain in the abdomen and vomiting. CT abdomen revealed a large exophytic mass in the epigastrium with enhancement pattern similar to hemangioma. No relationship of the mass could be made out with the adjacent structures on CT, histopathology proved it to be a GIST.

Keywords Hemangioma · Atypical GIST

Case Report

A 31-year-old man presented to the emergency department with pain in the abdomen and seven episodes of bilious vomiting for 1 day. The pain was sudden in onset with radiation to the back. On physical examination, the patient was afebrile (36.5 °C) and lethargic, with pulse rate 120/min, respiration rate 22/min, and blood pressure 130/90 mmHg. Pallor was present. Palpation of the abdomen revealed fullness in the upper abdomen. There was no rebound tenderness/muscle guarding. The rest of the

physical examination was unremarkable. A clinical diagnosis of acute cholecystitis was entertained.

Imaging

X-ray abdomen showed no obvious abnormality. Ultrasound abdomen showed free fluid in the peritoneal cavity with a large mass of mixed echogenicity predominantly hyperechoic in the epigastrium, superimposing the left lobe of the liver. CECT abdomen revealed the presence of a large heterogeneous exophytic soft tissue density lesion of size 11.4 (trans)×9.4 (AP) cms arising from the left lobe of the liver (Fig. 1). On contrast administration, the mass showed a peripheral enhancement in the arterial phase (Fig. 1), with centripetal filling in the venous and delayed (10 min) phase (Fig. 2). CT abdomen also showed ascites. No relationship of the mass could be made out with adjacent structures on CT. Compiling the above features—exophytic growth from the left lobe of the liver with a typical enhancement pattern defining hemangioma and free fluid—a diagnosis of ruptured hemangioma is considered. Rest of the abdominal organs were unremarkable. No further investigations were done due to the presence of free fluid in the abdomen which was suspected to be hemoperitoneum due to ruptured hemangioma. The patient was taken up for emergency surgery after blood transfusion for correction of anemia.

Peroperatively, a globular mass of size 12 cm×7 cm was found to be arising from the lesser curvature of the stomach. The mass was exophytic with no evidence of infiltration into the adjacent structures. A focal area of ulceration was noted in the posteromedial aspect of the mass. Wedge resection of the mass was done and the specimen was sent for histopathology. Cut surface of the specimen was tan fleshy with foci of cystic degeneration and

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Fig. 1 A large exophytic heterogeneous soft tissue density lesion arising from the left lobe of the liver

blackish areas. Histopathology confirmed GIST with a mitosis of 12/50 high-power field (HPF). Margins were free of tumor cells. Areas of hemorrhage (15 %) with dilated thin-walled vessels were noted focally. Tumor necrosis (10 %) was also noted. By immunohistochemistry the tumor cells were positive for CD 117, CD 34. Postoperative follow-up with CT abdomen and pelvis and screening the lung were unremarkable.

Discussion

GIST is a benign mesenchymal tumor arising from the intestinal cells of Cajal [1]. About 60–70 % of the

GIST arises from the stomach, 20–30 % from the small intestine, 10 % from the rest of the GIT [2]. The incidence of the GIST being 1.5/1,000,000/year [3], there is a slight male preponderance. Most of the GISTs are submucosal lesions and 15 % may be exogastric as in our case, and less than 5 % can be both extra- and intraluminal (dumbbell). GIST can also be multicentric [4] in less than 5 % of the cases.

Small GISTs are often asymptomatic and are detected incidentally. They become symptomatic because of their large size and their tendency to ulcerate and bleed [3]. Most common presenting signs and symptoms include abdominal pain, GI bleeding, and palpable mass. Very rarely a pedunculated GIST can cause obstruction to the pylorus and act as a lead point to intussusception. Histologically GIST can be benign (90 %) or malignant (10 %). GISTs with 1–5 mitosis/50 HPF are classified as a low grade, and GISTs with a mitosis of 10/10 HPF are classified as high-grade group [5]. The negative prognostic factors include size greater than 5 cm, high mitotic index, and a positive resection margins. Other poor prognostic factors include tumor rupture, more distal location, high cellularity, tumor necrosis, and presence of metastasis. The histological factors with a favorable prognosis include low mitotic index, size less than 5 cm, and lack of infiltration of the adjacent organs. Immunohistologically most GISTs have CD 34 antigen and CD 117 [6] antigen. CD 34 protein being a hematopoietic progenitor cell occurs in a variety of mesenchymal tumors.

Fig. 2 The arterial (a) and the delayed phases at 3 min (b) and 10 min; progressive centripetal filling in of contrast (c), and the ascitis (d)

CD 117 (also known as C-KIT protein) is a membrane receptor with a tyrosine component which is associated with malignant behavior of GIST. CT is the most common modality used to evaluate patients with GIST. Small GISTs are most commonly missed and appear as intramural masses. When they grow to large size (>5 cm) they become exophytic. Calcifications can also be seen. When tumors grow to large size, their blood supply tends to decrease and this leads to a central necrosis, eventually producing a ulceration. The most common metastasis is to the liver, followed by the peritoneum. Intracranial metastasis has also been reported. Lymphadenopathy is very rare in GIST. Very rarely GIST can arise from the pancreas [7].

Conclusion

Thus, GISTs being highly malignant tumors have a tendency to ulcerate and cause hemoperitoneum. GIST can be considered as a differential diagnosis in an untraumatic hemoperitoneum besides a ruptured hemangioma.

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