Malignant Gastrointestinal Stromal Tumor Of The Third Part Of The Duodenum Presenting As Gastric Outlet Obstruction: A Rare Presentation

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Citation

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Abstract

We hereby describe a case of gastrointestinal stromal tumor (GIST) in the third part of duodenum compressing the duodenum which was treated with wide local excision. The patient, a 60-year-old female, presented to us with features suggestive of gastric outlet obstruction and melena and on investigation was found to have a tumor in the paravertebral region anterior to the right kidney suggestive of GIST but no histological diagnosis was possible prior to laparotomy. Frozen section analysis showed it to be a GIST of intermediate-type malignancy, a local resection of the tumor arising from the third part of duodenum was performed and the margins were found to be negative for the tumor. Histological analysis revealed it to be GIST of intermediate-type malignancy.

Gastric outlet obstruction is an uncommon manifestation of GIST and whenever possible, a localized resection should be attempted given the low aggressive behavior of these tumors and the high morbidity of a more extensive procedure like pancreaticoduodenectomy.

DESCRIPTION

Duodenal GISTs are quite rare tumors and may present with vague symptoms. Since quite few cases have been reported in the literature till date, the management of these tumors has not been properly standardized. We hereby describe a case of malignant GIST in which the patient presented with features of gastric outlet obstruction secondary to extrinsic compression on the third part of the duodenum which is indeed an extremely rare presentation. A local resection of the tumor was done with anastomosis and the patient is doing extremely well after surgery with no signs of recurrence. Local resection should be favored over more extensive procedures like pancreaticoduodenectomy in these tumors of low aggressive potential with excellent results.

KEY POINTS

- Duodenal GISTs are quite uncommon tumors usually presenting with upper gastrointestinal bleeding.
- Gastric outlet obstruction is an extremely uncommon form of presentation of these tumors.
- These tumors have low aggressive potential.
- Extensive procedures like pancreaticoduodenectomy should be avoided and local resection favored as this produces good results.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common non-epithelial malignancies of the stomach and duodenum but they are quite rare tumors. These tumors usually present with features of upper gastrointestinal bleeding. Presentation as gastric outlet obstruction is quite rare. Surgery has been contemplated as the treatment modality of choice for these tumors but owing to the low incidence of duodenal GISTs (4.5% of all GISTs) the appropriate surgical procedure for these tumors is yet to be agreed upon. In our patient, local resection of the tumor achieving tumor-free margins was successful in treating a GIST in the third part of duodenum.

CASE REPORT

A 60-year-old female patient presented to our Out-Patient Department with features of early satiety, anorexia and episodes of vomiting occurring 2-3 hours after meals. She had associated fatigue and generalized weakness for the last

4 months. Over the last 3 months she had multiple episodes of melena treated conservatively but there was no history of hematemesis.

Physical examination revealed a poorly-defined rounded mass about 4 cm in diameter in the right upper quadrant of the abdomen which was freely mobile, hard in consistency and tender. On admission, blood pressure was 104/76 mm Hg, heart rate was 112/min and temperature was 36.8 degree Celsius. There was gross pallor. Hemoglobin on admission was 4.2 gm%. All other biochemical examinations were absolutely normal. Stool examination revealed trace quantities of blood.

Ultrasonography of the abdomen revealed a mass of 4.5 cm diameter in the paravertebral region adjacent to the inferior vena cava and a provisional diagnosis of enlarged lymph node was made. USG-guided FNAC from the mass revealed proteinaceous material and a definite diagnosis could not be made. A computed tomographic scan revealed an enhancing soft tissue mass in the right precaval region anterior to the right kidney suggestive of Gastrointestinal Stromal Tumor (GIST). An upper gastrointestinal endoscopy and a barium meal X-ray of the stomach both revealed features of gastric outlet obstruction with prolonged hold-up of barium inside the stomach. A CT-guided FNAC from the mass revealed proteinaceous material and no definite diagnosis could be made. The CEA and CA 19-9 levels were normal.

The patient was stabilized prior to the operation and anemia corrected. A formal laparotomy was done which revealed a solid tumor arising from the third part of the duodenum extending to the transverse mesocolon compressing the duodenum. No lymph nodes were found to be enlarged. Only a part of the duodenal wall was involved and since frozen section biopsy of the mass revealed it to be a malignant GIST of intermediate potential, a wide local excision was performed followed by anastomosis and the margins of resection were found to be free from the tumor. No other organ was involved in any form by the tumor and there was no evidence of any metastases. The total duration of the operation was 2 hours 50 min. and the estimated blood loss was 170 ml. The postoperative period was unremarkable and the patient was discharged after 2 weeks.

Histopathological examination revealed large epitheloid cells and spindle-shaped cells with increased cellularity and mitoses and a diagnosis of intermediate-type malignant GIST was made. The immunohistochemical study of the tumor revealed positivity for CD 117 (c-kit), CD 34, nestin and vimentin. The margins of resection were free from the tumor.

The patient is doing absolutely fine at 15 months follow-up with no signs of local recurrence or metastases.

Figure 1

Image 1: CT scan showing an enhancing soft-tissue mass anterior to the right kidney in the paravertebral region.

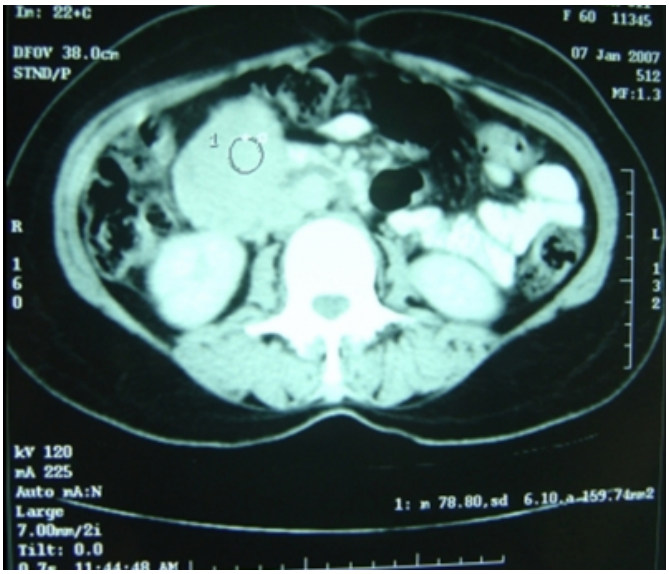


Figure 2

Image 2: Barium meal X-ray showing prolonged hold-up of barium within the stomach and duodenum.

Figure 3

Image 3: On laparotomy, a tumor is noted arising from the third part of the duodenum which is compressing it and projecting into the transverse mesocolon.

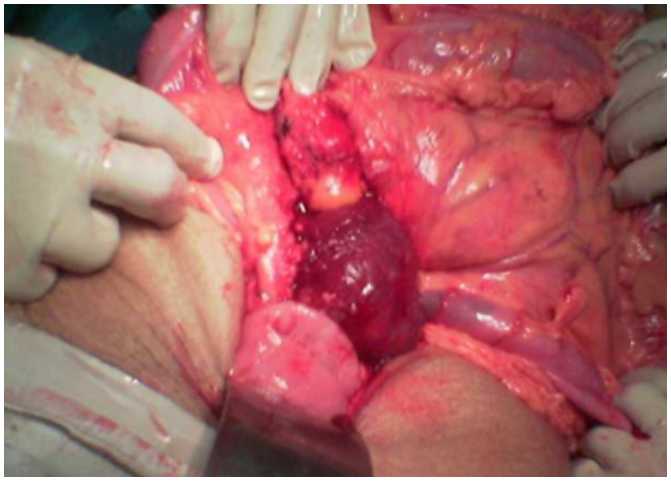
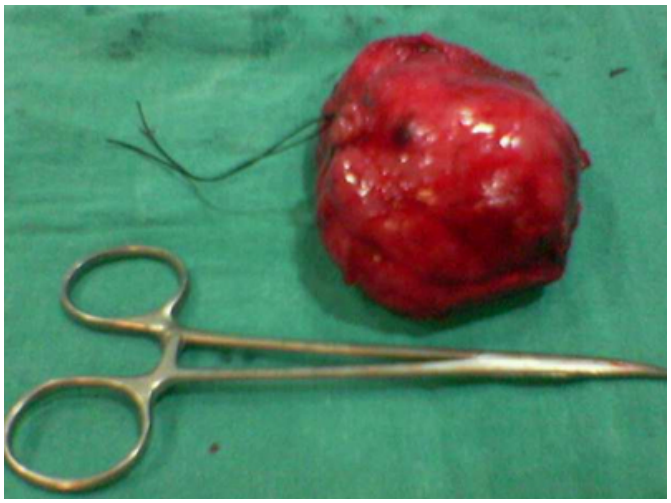


Figure 4

Image 4: Specimen of the tumor after local resection. The duodenal end has been marked with a thread.



DISCUSSION

Gastrointestinal stromal tumors (GIST) comprise of a rare group of tumors (0.3-1%) of the gastrointestinal tract (GIT) yet they are the most common mesenchymal neoplasms of the gastrointestinal tract. There is no gender predilection for these tumors which are seen most commonly in the 6 th and 7 th decades of life. Duodenal GIST is a rare entity comprising about 4.5% of all GISTs. [1]

Immuno-histochemical studies are essential for the classification of GIST into four prototypes: myoid, neural, dual differentiation and indeterminate types. The immunohistochemical expression of CD-117 (a growth factor receptor with tyrosine kinase activity) is the most important defining feature of GIST since a spindle shaped tumor in the GIT with CD-117 positivity is virtually diagnostic of GIST. [2] CD-117 is present in all GIST but not in true smooth muscle and neural tumors. Positivity for CD-34 is also common and occurs in 60-80% of GISTs. Other antigens that might be expressed by these tumors include vimentin, nestin (90-100% positivity), desmin, neuron specific enolase, neurofilament and smooth muscle actin. Preoperative diagnosis remains difficult and is not essential in all cases.

These tumors usually present with features of upper gastrointestinal bleeding, vomiting, anorexia, epigastric fullness and weight loss but features suggestive of gastric outlet obstruction as in our case are not usually seen with these tumors.

The size of the tumor has been shown to be an important prognostic factor with tumors larger than 5 cm in size being associated with an increased risk for malignancy. [3] There is, however, no clear cut-off point since the relationship between size and malignancy may be gradual.

Owing to the scarcity of these tumors, the treatment for duodenal GISTs has never been fully assessed. There has been no consensus since all available data are primarily from case series and individual case reports which attribute a grade E according to evidence-based medicine to the above. A wide variety of treatment options have been suggested including local resection to segmental duodenal resections and pancreatoduodenectomy for tumors with aggressive behaviors. [4] However, given the fact that these tumors grow expansively without being invasive with a low incidence of lymph node and distant metastases, local resection with tumor-free margins should be contemplated in small to medium sized tumors having low to intermediate degree of malignancy evident in frozen section given the morbidity of a more extensive procedure like Whipple's operation. [5,6,7]

CONCLUSION

Duodenal GISTs are quite rare tumors and even the malignant varieties have low aggressive potential. They usually present with features of upper gastrointestinal bleeding and anorexia. In our patient, the tumor presented with features of gastric outlet obstruction by extrinsic compression of the third part of the duodenum noted at laparotomy. An intraoperative frozen section characterized the tumor as malignant GIST and a localized excision was

performed. Biopsy revealed the margins to be free of the tumor and the patient is doing very well after surgery with no signs of recurrence. Therefore, radical procedures should be avoided in these tumors of low aggressiveness if they are found to be localized, considering the morbidity of more extensive procedures like pancreaticoduodenectomy.

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