CASE REPORT DOI: 10.15713/ins.bhj.132

Large Duodenal GIST with Course Over 5 Years and it's Management: A Case Report

ABSTRACT

Gastrointestinal stromal tumor (GIST) is a rare type of neoplasm of the gastrointestinal tract. Due to the rarity of occurrence, there is not much literature available. Existing literature is in the form of either case report or small case series. This is a case report of a large duodenal GIST diagnosed in 2014. After evaluation and surgical opinion at outside hospital, the patient was started on chemotherapy. The patient was non-compliant with the chemotherapy, taking it on and off from 2014 to 2019. In 2019, he was operated with pancreatoduodenectomy at our center and has been on routine follow-up since. Duodenal GIST has anatomical complexities with close proximity to noble structures; there is a lack of consensus on appropriate surgical treatment, prognostic factors, and survival which constitute a subject of controversy.

Key words: Duodenum, Gastrointestinal stromal tumor, Imatinib, Pancreatoduodenectomy

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare gastrointestinal (GI) tumors, accounting for 1–2% of all GI neoplasms.[1] It is a type of mesenchymal tumor of GI tract. Worldwide incidence of GIST is 10–20 cases/million population/year.[2] GISTs most commonly arise in stomach (60–70%), followed by small intestine (25–30%) and less often in colon and rectum (5–10%).[3] Duodenal GISTs represent <5% of the cases.[4]

Several factors are involved in the diagnosis and management of duodenal GISTs. The non-specific clinical manifestations often mimic a wide range of clinical conditions. Anatomical complexities with close proximity to noble structures can lead to misdiagnosis and inappropriate management.[5] The aim of the paper is discussion of the course of the patient for 5 years with vague clinical manifestation at presentation, the problems encountered in the management, treatment given, role of neoadjuvant chemotherapy, and the clinical response to it. This case also shows the difficulty faced in deciding on the optimal treatment and operability of the GIST.

CASE REPORT

A 36-year-old male presented to us with chief complaints of post prandial abdominal fullness, nausea, vomiting, and pain in abdomen in September 2019. Per abdomen-large lump was palpable in the epigastrium and right hypochondrium.

On probing further, the patient revealed a long history of an abdominal tumor, that is, GIST, dating back to 2014, summary is as follows:

1. September 2014: The patient presented with headache to a physician. On evaluation, hemoglobin (Hb) – 7.1 g/dL. Sonography showed a 14.4*14*8.6 cm sized heterogenous echotexture mass lesion in the epigastric region. CT scan (A+P) revealed 7.9*16*14 cm lobulated heterogeneously enhancing and non-calcified mass lesion with central area of necrosis seen in retroperitoneum, involving 3rd part of duodenum. It is smoothly indenting and displacing but not involving the pancreas, gallbladder, and right lobe of liver, left laterally displacing the jejunal loops. The mass lesion is pushing the superior mesenteric artery (SMA) and SM vein anteriorly. The upper GI endoscopy was done which revealed ulcerative friable mucosa with nodularity involving half of circumference immediately after duodenal papilla. Microscopy revealed spindle cell tumor and consistent with duodenal GIST. IHC was C-Kit (CD 117), CD-34, DOG-1, and vimentin positive. The patient was, then, started on neoadjuvant Imatinib. No clinical or radiological assessment was done after December 2014.
2. Patient stopped taking tablet imatinib in December 2015.
3. January 2017: presented with Malena. Hb – 4 g/dL. He received four packed cells transfusion. CT scan (A+P)-mass of 9.8*11.8*8.2 cm [Figure 1]. Rest findings were same as above. On upper GI endoscopy – large submucosal mass with ulcerations and active oozing in second

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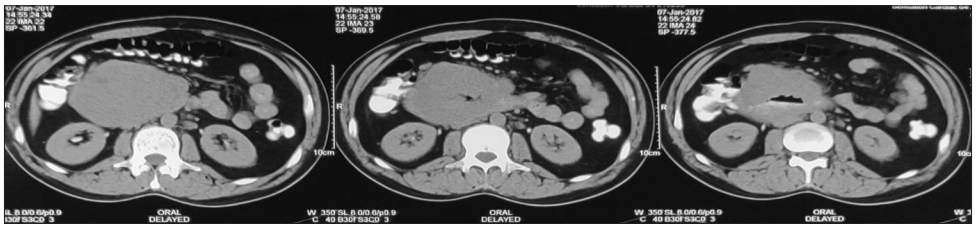

Figure 1: CT scan done in January 2017

part of duodenum. He consulted a surgical oncologist outside, who opined it as inoperable GIST and suggested angioembolization if possible. Interventional radiologist could not do angioembolization due to extensive nature of lesion and blood supply from major branches of SMA. He was restarted on tab imatinib. CT scan (A+P) done in April 2017 showed moderate resolution of heterogenous mass, which, now, measured 9.2*5.5 cm.

4. January–October 2018: Patient discontinued imatinib therapy.
5. September 2019: presented to us. CT scan (A+P) revealed heterogeneously enhancing soft-tissue mass lesion, measuring 9*16*10 cm, causing proximal partial small bowel obstruction. The mass is displacing the head and uncinate process of pancreas anteriorly with loss of fat planes. Posteriorly, the mass is seen to abut the liver and compress the IVC with loss of fat planes at places. There is perilesional fat stranding [Figure 2]. Compared to previous scan, there is significant increase in the size. PET CT scan revealed increase in size and avidity of the lesion. There was no evidence of any FDG avid lesion elsewhere in the body.

Management

The patient was already on imatinib. Since he was presenting with symptoms of obstruction, we decided to go ahead with surgical intervention. The tumor bulk was large and was abutting all the major structures. However, there was no evidence of distal metastasis or any upfront encasement of major vessel. Surgery was undertaken with a curative intent. Intraoperative findings were as follows: large mass seen to be arising from the second part of duodenum. Tumor was abutting the pancreatic head and lower CBD. Pancreatoduodenectomy (PD) (Whipple's procedure) was done and entire tumor removed en masse. Feeding jejunostomy was done. The patient withstood the surgery well. Post-operative period was uneventful and he was on full oral diet at the time of discharge.

Specimen [Figures 3 and 4] on gross examination was found to be adherent to CBD, but not infiltrating its wall.

HPE: s/o GIST, neoplastic spindle cells. Mitotic activity shows 7–8/50 hpf. Rest of the cut margins were free.

Postoperatively, the patient was again started on imatinib therapy. On routine follow-up since, he does not show any evidence of recurrence. He is tolerating adjuvant chemotherapy well and putting on weight.

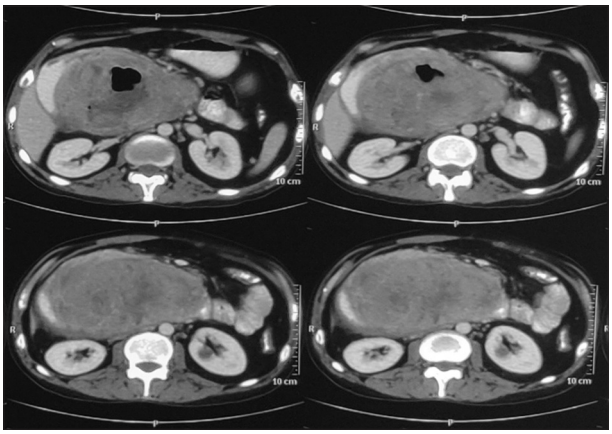

Figure 2: CT scan done in September 2019

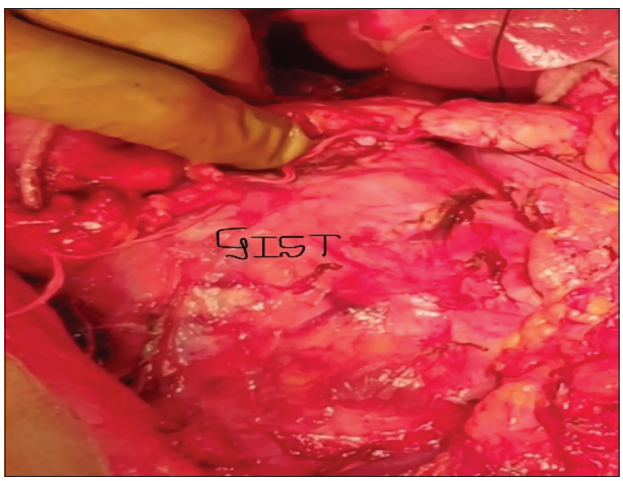

Figure 3: Intraoperative picture of duodenal gastrointestinal stromal tumor

DISCUSSION

This is a rarely encountered case of a large duodenal GIST. Most of the duodenal GISTs develop in 2nd and 3rd portion of duodenum.[6] This patient had it in 2nd part. The patient had an unusual presentation with headache. Hematological evidence of anemia led to further evaluation. Clinical manifestations are seen in 70% cases, 21% are found incidentally, and 10% on autopsy.[7] As per the literature, the most frequent manifestation of duodenal GIST is upper GI bleeding,[8] presenting as malena and chronic anemia.

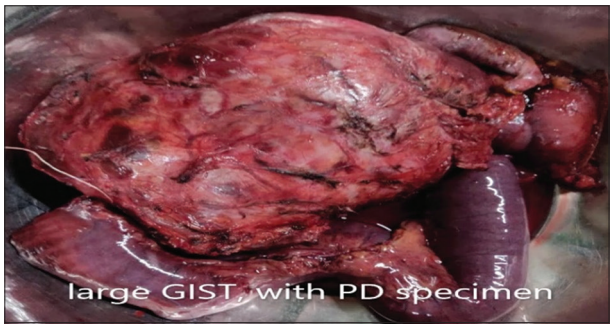

Figure 4: Specimen of pancreatoduodenectomy

Duodenal GIST may have various initial clinical manifestations leading to misdiagnosis. Therefore, diagnosis requires proper clinical history and examination supported by radiological and histopathological confirmation. Most present with acute bleeding or chronic anemia hence endoscopic evaluation of upper GI tract should be the first step.[4] It allows for biopsy and can also be therapeutic. Furthermore, endoscopy rules out other etiologies of upper GI bleed. In this case, upper GI scopy confirmed the mass to be arising from 2nd part of duodenum and also facilitated a biopsy for diagnosis. In cases of diagnostic uncertainty, endoscopic ultrasound (EUS) is an invaluable modality. EUS provides a precise evaluation of the size, border, layer of origin, echogenicity, and heterogeneity of the lesions and allows fine needle or trucut biopsy. Percutaneous biopsy should be avoided due to the risk of tumor spillage and dissemination.

A very important tool in the diagnosis of all abdominal pathologies is CT scan. CT scan is necessary to make an exact staging and decide management. This patient had 1st CT scan done in 2014, which showed the size, site, and extent of tumor and ruled out regional metastasis. In subsequent years from 2014 to 2019, CT scans revealed response to chemotherapy and increase in size of tumor on non-compliance. Fluorodeoxyglucose positron emission tomography is not routine tool but can be useful in prognosis and follow-up. Histopathology and molecular characteristics play an important role in diagnosis, treatment, and prognosis.

In contrast to the other locations, for duodenal GIST, there is no uniformly adopted surgical strategy due to the low incidence, lack of enough experience, and the complex anatomy of the duodenum. Imatinib mesylate has played a key role as a neoadjuvant therapy in the management of GISTs.[9] In locally advanced disease, neoadjuvant imatinib may downstage the tumor to allow R0 resection or even an organ preserving intervention.[9] Due to the bulk of the tumor, this patient was started on neoadjuvant chemotherapy in 2014. The patient was non-compliant with the imatinib therapy.

According to the latest guidelines, surgery is the primary treatment of choice for patients with localized or potentially resectable GIST lesions.[10] Therefore, individually tailored surgical approach is recommended. In fact, there are three main surgical options: PD, wedge resection, and segmental resection. Coming to this case, He was diagnosed incidentally and presented with most of the complications seen in a duodenal GIST, like upper GI bleeding, anemia, and GI obstruction. He was responsive to neoadjuvant imatinib therapy, but non-compliant in the duration of 5 years from 2014 to 2019, the patient twice took surgical oncology opinion at another hospital, but was labeled inoperable. In the duration of 5 years from 2014 to 2019, the patient twice took surgical oncology opinion at another hospital, but was labeled inoperable. This is one of the largest duodenal GISTs reported. This case shows an unusually long course of the disease with indecisiveness over operability and optimal surgical treatment. However, in experienced hands, the surgical outcome is good even with a supra-major surgery like pancreaticoduodenectomy. The rupture of GIST during the surgery should be avoided because is associated with nearly 100% risk for recurrence. Post-operative adjuvant imatinib therapy should follow.

CONCLUSION

Duodenal GIST, though rare, can have various manifestations. Important is adequate clinical evaluation, early diagnosis, and prompt treatment. This case showed that long duration of the illness and irregular neoadjuvant therapy led to complication of gastrointestinal obstruction. PD with clear margins gave the patient symptomatic relief as well as possible cure. Adjuvant imatinib therapy was still continued and the patient is under routine follow-up with no evidence of recurrence so far.

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How to cite this article: Lambe A, Yadav R. Large Duodenal GIST with Course Over 5 Years and it's Management: A Case Report. Bombay Hosp J 2022;64(4):46-49.

Source of support: Nil, **Conflicts of interest:** None