

Large Locally Advanced Duodenal Gastrointestinal Stromal Tumor (GIST) Successfully Managed by Combined Use of Neoadjuvant Imatinib and Surgical Resection

Yeap, J. H^{1*}, Tan, P. G¹, Leow, V. M², Ramasamy, U¹

¹Hospital Taiping, Department of General Surgery, Perak, Malaysia

²Hospital Sultanah Bahiyah, Hepatobiliary Department, Alor Setar, Kedah, Malaysia

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*Corresponding author: Yeap, J. H

Hospital Taiping, Department of General Surgery, Perak, Malaysia

Abstract

Case Report

Duodenal GISTs are rare mesenchymal neoplasms of the gastrointestinal tract. The usual presentation is gastrointestinal bleeding or abdominal pain. They can be asymptomatic until they reach a considerable size. Tyrosine kinase inhibitors (TKI) are important in high risk GISTs and in locally advanced disease. We report a 65-year-old man with abdominal discomfort, anemia and clinically palpable abdominal mass. A Computed Tomography (CT) scan of the abdomen and pelvis was done which showed a locally advanced duodenal GIST involving second and third part of duodenum. Neoadjuvant imatinib was given to this patient for 6 months. He underwent Whipple's procedure and right hemicolectomy. The histopathological examination report revealed a high-risk duodenal GIST. Adjuvant Imatinib was continued for 1 month after the operation. He continued to do well with no evidence of disease recurrence based on post-operative CT abdomen and pelvis.

Keywords: Duodenal Gastrointestinal Stromal Tumor (GIST), Neoadjuvant Imatinib, Surgical Resection, Whipple Procedure.

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INTRODUCTION

Gastrointestinal stroma tumor (GIST) is a rare mesenchymal neoplasm of the gastrointestinal tract. The most common site involved is the stomach (60%), small intestines (20%) and rarely the duodenum (5%) [1]. Patients with GISTs can present with abdominal pain and gastrointestinal bleed which cause them to become anemic [2]. When the tumor has reached a considerable size, patient can become symptomatic with palpable abdominal mass [2]. Tyrosine kinase inhibitor such as imatinib has been used as a neoadjuvant chemotherapy to improve the resectability of tumor and achieve clear margins during surgical resection [3]. The malignant potential of the tumor relies on histopathological findings. Non-gastric GISTs poses higher malignant potential compared to gastric GISTs [1]. Here we present a patient with large duodenal GIST including its initial presentation, diagnostic workup, combined chemotherapy and surgery performed, and the issues related to locally advanced duodenal GISTs.

CASE REPORT

A 65-year-old man with history of open cholecystectomy in 2017 presented with abdominal discomfort and recent loss of weight for 5 months. He denied other associated symptoms, including altered bowel habit, passing blackish stool, dysphagia or urinary symptoms. There was no family history of gastrointestinal malignancy. Abdominal examination revealed a 5x5cm right lumbar mass.

His blood investigations results were normal except that his hemoglobin level was 7.9g/dl. Ultrasound abdomen revealed a right lower intraabdominal mass. Subsequently patient had a computed tomography (CT) of the abdomen and pelvis which showed an 8.5x8.8x10.7cm mass arising from the second and third part of duodenum. Upper gastrointestinal endoscopy revealed an outpouching at third part of duodenum with irregular mucosa. Endoscopic biopsy showed a gastrointestinal stromal tumor.



Figure 1: CT image showing heterogenous enhancing mass from second and third part of the duodenum

Neoadjuvant imatinib was given to patient for 5 months. Repeated CT scan post neoadjuvant Imatinib showed a smaller size of the tumor. Subsequently, patient underwent pancreaticoduodenectomy (Whipple procedure) and right hemicolectomy. Histopathological examination revealed a large lobulated solid tumor measuring 10x9x8cm from third and fourth part of

duodenal wall. The tumor showed predominantly spindle cells arranged in interlacing fascicles with a mitotic count of 3/50 high power field (HPF). The tumor was seen just adjacent to the pancreatic tissue. Resected margins were free. The tumor was positive for C-Kit and DOG, but negative for CKAE1/AE3, CD 34, SMA, DESMIN and S100.

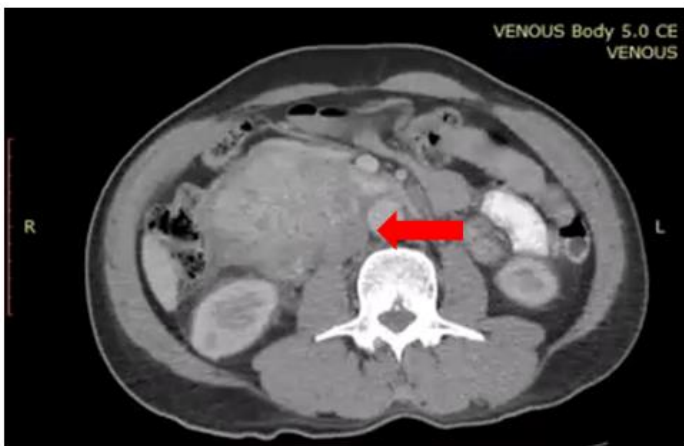


Figure 2: CT image showing smaller size of duodenal mass post neoadjuvant Imatinib

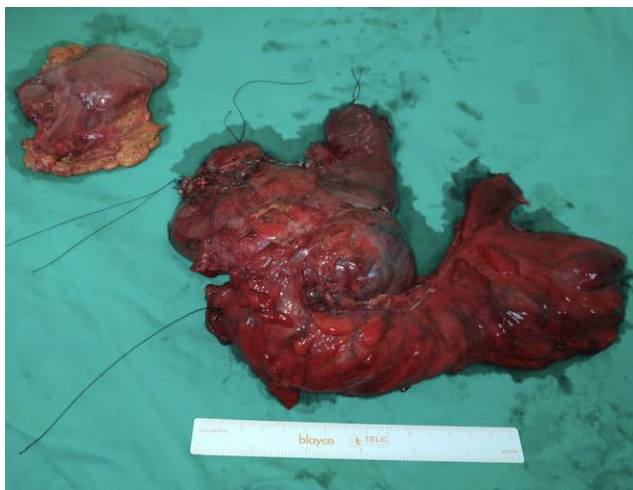


Figure 3: Resected Whipple specimen showing large solid tumor arising from third and fourth part of duodenal wall

Patient tolerated operation well and was discharged after 3 days of hospitalization. Another 1-month of adjuvant chemotherapy with Imatinib was given to patient. CT scan repeated post adjuvant chemotherapy showed no tumor recurrence and patient continued to do well.

DISCUSSION

GIST is the most common mesenchymal neoplasms of the gastrointestinal tract. However, duodenal involvement with this type of tumor is rare. The incidence of this tumor is estimated at 1 in 100,000 and it usually affected people in their sixties with male predominance [4]. The main clinical presentation is usually gastrointestinal bleeding. However, this was not found in this patient, who was diagnosed during clinical examination and radiological imaging which was CT scan. Distal lesions as the one described in this case may be missed as it may not be reached by conventional upper gastrointestinal endoscopy. Ultrasound abdomen may help in diagnosing abdominal mass but a CT scan is necessary for localizing lesion and staging of the disease [5]. Management of GISTs should involve multidisciplinary team including surgeon, oncologist, and radiologist [6]. Neoadjuvant treatment with imatinib has been showed to downsize tumor and hence ease the process of surgery and results in less morbidity [7]. Surgical treatment is the standard treatment for duodenal GISTS. Surgical strategy is determined by the tumor size and location [8]. In this case, patient underwent pancreaticoduodenectomy as second part of duodenum is involved. The recurrence of GISTs depends on biology of tumor [1, 4, 9, 10]. Higher malignant potential is seen in small intestine compared to gastric GIST [1, 4, 9, 10]. The contributing factors for malignant potential in GISTs are tumor size, tumor location and mitotic activity [1, 4, 9, 10]. Other factors which subject the patient to high risk of recurrence include rupture of tumor (before or during surgery), failure to achieve clear surgical margin and specific genotype [9, 10]. There are a few classification systems for risk stratification of GIST tumor. The latest classification system is called the Armed Forces Institute of Pathology (AFIP) classification system [9, 10]. Based on the AFIP system, this patient has high risk GIST tumor as the tumor size is 10cm, mitotic count is 3/50 HPF and tumor site is non gastric [10]. Considering that this patient has a high-risk GIST, it is justifiable for adjuvant chemotherapy with a tyrosine kinase inhibitor which is imatinib. The appropriate follow up time for patient with high-risk GISTS should be every 3-6 months with repeated CT scan [10].

CONCLUSION

This case report shows that duodenal GISTS can be asymptomatic until it reaches a considerable size and cause anemic symptoms. Neoadjuvant chemotherapy helps to decrease tumor size to facilitate ease of surgery.

Surgical resection remains as the gold standard treatment for GISTs. Adjuvant chemotherapy is mandatory in high risk GISTs to lower risk of recurrence and improve patient long term survival rate.

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