Intestinal Obstruction Secondary To Duodenal Gastrointestinal Stromal Tumour

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Abstract

Though gastrointestinal stromal tumours are common, a duodenal stromal tumour is a rare entity. We present such a rare case with duodenal obstruction which caused a dilemma in the management of this patient. Though a Whipple's operation was planned, intraoperatively, due to the pedunculated nature and free intraluminal mobility, resection of the polyp was done and histopathology was consistent with the diagnosis of a duodenal stromal tumour, which was reconfirmed with special stains/immunohistochemistry. Surgeons and pathologists should keep this rare primary tumour of the duodenum in mind as a differential diagnosis when examining/treating duodenal lesions, as a major surgical intervention can be avoided, cutting down on the cost, hospital stay, morbidity and mortality to the patient. A literature review and a discussion on the pertinent clinical, radiological, pathological and surgical aspects of stromal tumours have been discussed.

Introduction

Although stromal tumours are the most common gastrointestinal mesenchymal neoplasms, they are uncommon and duodenum is the least frequent site of occurrence (3-5%) [1]. Previously, they have been referred to as leiomyomas, leiomyoblastomas and leiomyosarcomas. The term ‘stromal tumour’ was first coined by Schaldenbrand and Appelman in 1984 [2]. Duodenal gastrointestinal stromal tumours (GIST) are extremely rare with a variable clinical presentation. A palpable abdominal mass is the most frequent sign. Most cases are asymptomatic until they reach large size and may cause acute haemorrhage in the GI tract. Obstructive jaundice may be seen when GIST is located near the ampulla of Vater [3]. Small asymptomatic lesions arising in the duodenum may be found incidentally during radiological investigations or laparotomy for other reasons [4]. Surgery remains the ‘gold standard’ for non-metastatic GISTs. Metastatic GISTs require chemotherapy and then might require surgery. Post-resection, the recurrence rate is around 35% [5]. A major diagnostic criterion of GISTs is expression of c-kit (CD117) and additional criteria include CD 34 and desmin positivity at IHC [6]. Due to nonspecific symptoms, delayed interval between the start of the lesion and clinical presentation, diagnosis and treatment becomes all the more difficult and challenging.

Case Report(s)

A 56-year-old male presented with history of pain abdomen, post-prandial vomiting with haematemesis (4 episodes) and maelena of one week duration. The pain was in the right upper abdomen and was of dull aching type and continuous. There was no history of jaundice. On general examination, pallor was present and per abdomen examination was unremarkable. A gastroduodenoscopy was done which showed a polypoidal lesion with a stalk at D1-D2 junction, which was biopsied (Illustration 1). The biopsy report was unremarkable. A clinical diagnosis of a periampullary carcinoma was made and a CT scan of abdomen was done to look for the size of the lesion, degree of luminal obstruction and for possible extraluminal spread and metastases, which revealed an intraluminal duodenal mass obstructing the lumen and no intra-abdominal metastases (Illustration 2). Patient underwent an exploratory laparotomy and was found to have a freely mobile, soft, fleshy tumour confined to the duodenal lumen. Hence, a duodenotomy and excision of the polypoidal mass with a 2 cm margin was done. Histopathology revealed ulcerated duodenal mucosa, with one focus showing a few crypts covered by fibrinous exudate with neutrophils, overlying a tumour composed of interlacing fascicles of spindle cells with elongated eosinophilic cytoplasm, wavy buckled nuclei few with intracytoplasmic eosinophilic globules, occasional mitoses (Illustration 3).

Discussion

GISTs are rare tumours with an annual incidence of approximately 3000-6000/year in the United States and are most commonly seen in the seventh decade of life, although they may arise at any age. Stomach is the most frequent site of occurrence (45-65%), followed by the small bowel (15-25%), large bowel including the rectum (5-10%), oesophagus (5-10%)
and duodenum (3-5%) [1]. GIST constitutes a distinct group of rare GI tumours that originate from the interstitial cells of Cajal which are regulators of gut peristalsis and normally express CD117, which is a product of the c-kit proto-oncogene that encodes a tyrosine kinase receptor, which regulates cellular proliferation in GISTs [2].

Most cases of GISTs are sporadic although few familial cases have been reported. In GIST cells, the c-kit gene is mutated in approximately 85% to 90% of cases. GIST cells that do not have a mutated c-kit (“wild-type”) do have a mutation in another gene, PDGFR-α (platelet derived growth factor receptor alpha), which is a related tyrosine kinase. Mutations in the exons of 11, 9 and rarely 13 and 17 of the c-kit gene are known to occur in GIST. D816V point mutations in c-kit exon 17 are responsible for resistance to targeted therapy drugs like Imatinib mesylate [7].

The common presenting symptoms of duodenal GISTs are GI bleeding, vomiting, abdominal discomfort and mass per abdomen. These symptoms result from the fact that GISTs are typically fast growing neoplasms that often outgrow their blood supply and as a result they frequently develop a necrotic centre. When this fistulises to the enteric lumen or erodes a vessel, GI bleeding ensues that at times can be quite profuse. Fatigue due to anaemia may be present. Duodenal GISTs may present with obstructive jaundice [3]. Duodenal GISTs are most frequently diagnosed in the workup of symptoms not specific to these masses. Investigations like gastroduodenoscopy, barium meal, CT, MRI, endoscopic ultrasound and PET scan has been mentioned in the workup of these patients, amongst which the most useful is a CT scan. Not only does this demonstrate the size and anatomic location of the mass, it may also demonstrate features suggestive of stromal tumours (well vascularised, necrotic centre, heterogenous appearance) and provide evidence of metastatic disease. There is no standard staging system for GIST; however, the two most important factors in determining the risk of a GIST being cancerous are the size of the tumour and the mitotic count [4].

Surgery is the mainstay of treatment and the goal is complete tumour removal. At times the tumour may be large or spread into nearby organs and such patients will require Imatinib before surgery [5]. Targeted therapy has found a place in the treatment on GIST in which, faulty genes or proteins that contribute to cancer growth and development are targeted [8]. The United States Food and Drug Administration (FDA) have approved Imatinib, a cytostatic drug and a tyrosine kinase inhibitor for the treatment of GIST, which is now the drug of choice. It is usually given as either the only treatment or in combination with surgery (before or after surgery). The usual dose of Imatinib is 400 mg daily that can be raised to 800 mg daily. The other drug is Sunitinib, a tyrosine kinase inhibitor with anti-angiogenic properties and was approved in 2006 by the FDA for treating GIST when the tumour continues to grow even after treatment with Imatinib or in cases where it cannot be given [9]. Radiation therapy is not effective for treating the tumour itself, but may be used as a palliative treatment in painful metastasis [10].

### Conclusion

GISTs, though rare, can present with varied symptoms from being silent to distant metastases. They are being increasingly picked up due to advances in IHC. Though surgery is the mainstay of treatment, chemotherapy plays a significant role in control and treatment of these tumours. Surgery is indicated in bowel obstruction, tumour bleed or to debulk the tumour. The survival in these patients is promising.

### References

6. Park SS, Ryu JS, Oh SY, Kim WB, Lee JH, Chae YS, Kim SJ, Kim CS, Mok YJ. Surgical outcomes and immunohistochemical features for gastrointestinal
Illustrations

Illustration 1

Gastroduodenoscopy showing a tumour at D1-D2 junction, completely occluding the lumen.

Illustration 1: Gastroduodenoscopy showing a tumour at D1-D2 junction, completely occluding the lumen.
Illustration 2

CT abdomen showing complete duodenal occlusion (arrow).
Illustration 3

GIST

Illustration 3: Interlacing fascicles of spindle cells, wavy buckled nuclei, intracytoplasmic eosinophilic globules and occasional mitoses (Haematoxylin & eosin, x40).
Illustration 4

GIST, high power

Illustration 4: IHC showing scattered cells with cytoplasmic and membrane positivity (c-kit staining x40).
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