Gastrointestinal Stromal Sarcoma – A Case Report of Palliative Enteral Stenting

H B Yim, J R Saltzman, D L Carr-Locke

ABSTRACT

We report a case of metastatic gastrointestinal stromal sarcoma (GISS) in a 33-year-old man who subsequently underwent successful palliative endoscopically-placed enteral stenting for duodenal stenosis secondary to extrinsic compression. Enteral stenting for palliative relief of malignant gastrointestinal obstruction is recommended for its safety, efficacy and cost-effectiveness.

Keywords: Enteral stenting, endoscopic, gastrointestinal stromal sarcoma, gastrointestinal obstruction, palliative

Singapore Med J 2001 Vol 42(11):534-536

INTRODUCTION

Gastrointestinal stromal sarcomas (GISS) are very rare tumours that are usually very aggressive in nature with a poor prognosis. The origin of these tumours was once a source of much speculation and controversy. However, they have recently been shown to originate from connective tissue elements of the bowel wall[1]. Aggressive surgical resection in early cases, achieving complete resection, might lead to prolongation of life[2]. However, most cases are diagnosed at the advanced stage where surgery is not feasible or contraindicated. Endoscopic enteral stenting is now widely accepted as an efficacious and cost-effective method to palliate such malignant enteral obstruction.

CASE REPORT

A 33-year-old man from Venezuela first noticed a hard mass in his anterior abdominal wall in August 1998, initially thought to be a lipoma. However, the mass progressively enlarged and abdominal CT scan performed in March 1999 revealed a 7 cm by 6.5 cm by 4.2 cm mass of the anterior abdominal wall without definite involvement of any intra-abdominal organ. Subsequently, the mass was resected and the pathology revealed a high grade gastrointestinal stromal sarcoma grade 3/3 with epithelioid features. A repeat post-operative CT scan one month later revealed a new right-sided colonic mass of 4 cm diameter. This was resected via a right hemicolectomy and pathology confirmed the origin to be similar to that of the previous abdominal wall mass.

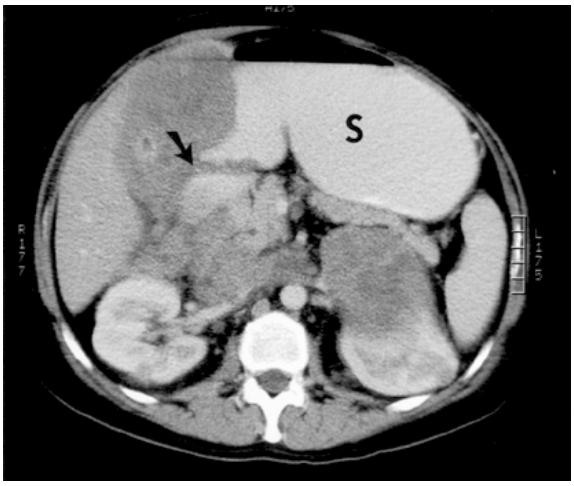

Fig. 1 CT scan of the abdomen showing a distended stomach with fluid level (S) and extrinsic compression of the first and second parts of the duodenum by the tumour (arrow) with no flow of contrast into the small bowel.

A CT scan in July 1999 showed bilateral adrenal gland masses and a new mass in the right kidney. Chemotherapy was initiated with a protocol using the experimental drug CT2584 (xanthine analogue). However, a post-chemotherapy CT scan in late August 1999 showed further progression of the abdominal tumour and the chemotherapy was discontinued. He then presented with lower back pain and MRI revealed a metastatic mass to his lumbar spine compressing the L3 nerve root. Radiation therapy was then initiated.

Most recently, he presented in September 1999 with a one-week history of vomiting and regurgitation with inability to retain food or liquids. This was associated with abdominal pain but no significant change in bowel habits. He was started on anti-emetics but without improvement. Subsequently, he developed coffee-ground vomitus and an upper endoscopy revealed oesophagitis and gastritis with a stenosed first part of the duodenum which did not allow passage of the gastroscope.

CT scan of the abdomen at this time (Fig. 1) revealed a markedly distended stomach with no contrast flowing

Gastroenterology Unit
Department of General Medicine
Tan Tock Seng Hospital
11 Jalan Tan Tock Seng
Singapore 308433

H B Yim, MBBS, MRCP, FAMS
Consultant

Division of Gastroenterology
Department of Medicine
Brigham and Women's Hospital
Harvard Medical School
Boston
Massachusetts, USA

J R Saltzman, MD
Associate Director of Endoscopy

D L Carr-Locke, MD, FACG, FRCP
Director of Endoscopy

Correspondence to:
Dr Yim Heng Boon
Tel: (65) 256 6011
Email: heng_boon_yim@ttsh.com.sg

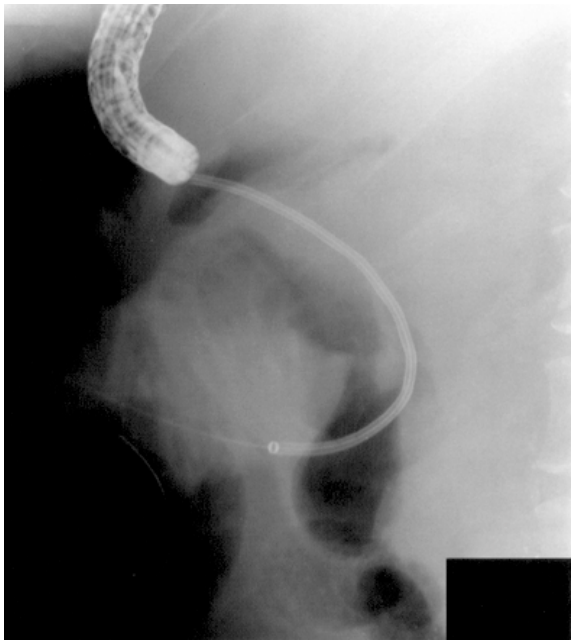

Fig. 2 Radiograph of the endoscopic enteral stenting procedure showing the placement of the Enteral Wallstent over a guidewire just prior to deployment.

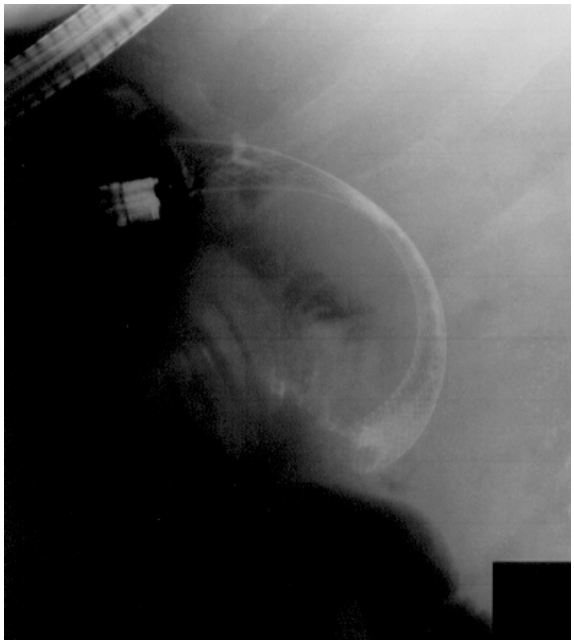

Fig. 3 Radiograph of the fully deployed Enteral Wallstent across the duodenal narrowing.

past the pylorus. The 2nd and 3rd parts of the duodenum appeared encased by the tumour which was also seen to involve the liver, abdominal wall, adrenal glands, right kidney, and retroperitoneal lymph nodes. Patient was referred for endoscopy to relieve the obstruction by stent placement.

An endoscope (Fujinon gastroscope EG 410D) was passed easily into the duodenal bulb. Distal to the bulb, a moderate stenosis of the junction of the first and second parts of the duodenum, secondary to extrinsic compression of the tumour, was present. With application of pressure, the gastroscope (diameter 9.5 mm) could pass through the narrowing to the junction of the second and third parts of the duodenum which appeared patent. An ERCP catheter was passed through the accessory channel of the gastroscope into the duodenum. Contrast was injected and patency beyond the second part was confirmed fluoroscopically. A Jagwire guidewire (Boston Scientific Microvasive, Natick, MA, USA) was passed through the catheter and the endoscope withdrawn to the pylorus, taking note of the distance from the distal to the proximal end of the narrowing which measured 6 cm. Finally, an Enteral Wallstent (Boston Scientific Microvasive, Natick, MA, USA) of diameter 22 mm and length 90 mm was deployed successfully across the stenosis (Figs. 2 and 3). At completion of the procedure, the proximal end of the stent was at the pylorus and distal end at the junction of the second and third parts of the duodenum. Contrast was then injected into the proximal end of the stent and was seen to flow to the duodenum distal to the stent, confirming technical success of the procedure.

The patient was discharged a few days later tolerating oral nutrition very well. He was last seen at the outpatient clinic one week later with resolution of his obstructive symptoms and was eating a regular diet. He returned to his country of origin and has not had further follow-up.

DISCUSSION

Gastrointestinal stromal sarcomas (GISS) are rare malignant tumours which represent about 2.5% of all soft tissue sarcomas[3]. They are part of a larger group of tumours called gastrointestinal stromal tumours (GIST)[4] which are thought to arise from connective tissue elements located along the entire length of the gut. The nomenclature of GIST has undergone numerous changes over the past years, reflecting the controversies surrounding its histogenesis. Such tumours have been labelled as bizarre leiomyomas, leiomyoblastomas, and epithelioid leiomyomas, but these have largely been replaced by GIST. GIST itself has been classified into three subgroups: (a) tumours with well-developed features of differentiation; (b) gastrointestinal stromal tumours; and (c) gastrointestinal stromal sarcomas (GISS)[1].

The exact incidence of GISS is not well established but it is demonstrated to be most commonly found in the stomach (47%), followed by small bowel (35%), colorectum (12%), and oesophagus (5%)[5]. The typical presenting symptoms include upper gastrointestinal tract bleeding (generally as a result of mucosal ulceration), abdominal pain, a palpable mass, intestinal obstruction, nausea, and dyspepsia[3]. Factors responsible for clinical

outcome in such patients include age, histologic grade and clinical stage. There appears to be an increased survival in patients with localised tumours as compared to those with infiltrative or metastatic disease at the time of diagnosis[5]. Invasion of adjacent organs at the time of surgery is associated with highly aggressive behaviour and early death[6].

Surgical resection is the treatment of choice for early stage, non-metastatic GISS[7,8]. Reported survival data after surgery for GISS varies widely[7-12]. The overall five-year survival rate of operable gastric sarcoma ranges between 19% and 56% whereas those in the small or large intestine ranges have survivals between 40% to 50% following complete excision. Generally, the prognosis of GISS is poor due to its aggressive nature and many patients have metastases at the time of diagnosis which renders the tumour inoperable. The effectiveness of adjuvant chemotherapy and radiotherapy has not been established.

For patients with advanced metastatic disease due to GISS, palliative measures are indicated for relief of symptoms. Our patient had gastric outlet obstruction due to advanced metastatic disease. Surgery carries the risk of significant morbidity and mortality as gastrojejunostomy may be associated with up to 10% mortality rate as in patients with gastric outlet obstruction due to pancreatic carcinoma[13]. Several studies have shown encouraging outcomes in treatment of malignant gastric-outlet, duodenal and small-intestinal obstruction by endoscopically-placed stents[14-20]. Large-diameter (22 mm) stents provide significant lasting relief from obstructive symptoms and allow the majority of patients to eat a regular diet. Oesophageal stents have been used for gastroduodenal obstruction but these are limited by their short delivery systems and need for non-endoscopic placement.

The enteral Wallstent, previously approved for use in malignant colonic obstruction, has recently been FDA-approved for use in malignant gastroduodenal obstruction and is placed trans-endoscopically[19]. To our knowledge, there has been no previous case report on enteral stenting in a patient with obstructive, metastatic GISS. In this patient, an Enteral Wallstent was deployed successfully across the obstruction and within a day, the patient tolerated oral nutrition. He was on regular diet upon discharge a few days later. The use of endoscopic stent placement appears to extend and improve the quality of such patients' poor life expectancy. In a number of studies involving patients with malignant gastric-outlet, duodenal and small-intestinal obstruction who underwent enteral stenting, the mean follow-up period was reported to be about 13 weeks[17-20]. The majority of deaths in these patients were due to progression of the malignancy rather than recurrent gastrointestinal obstruction. We conclude that patients with extrinsic, inoperable adjacent compression of the gastrointestinal tract should be considered for endoscopically-placed enteral stents for palliation.

REFERENCES

1. Suster S. Gastrointestinal stromal tumours. Semin Diagn Pathol 1996 Nov; 13(4):297-313.
2. Lev D, Kariv Y, Issakov J, et al. Gastrointestinal stromal sarcomas. Br J Surg 1999 Apr; 86 (4):545-9.
3. Licht JD, Weissmann LB, Antman K. Gastrointestinal sarcomas. Semin Oncol 1988 Apr; 15(2):181-8.
4. Ueyama T, Guo KJ, Hashimoto H, et al. A clinicopathologic and immunohistochemical study of gastrointestinal stromal tumours. Cancer 1992 Feb 15; 69(4):947-55.
5. McGrath PC, Neifeld JP, Lawrence W Jr, et al. Gastrointestinal sarcomas. Analysis of prognostic factors. Ann Surg 1987 Dec; 206(6):706-10.
6. Shiu MH, Farr GH, Papachristou DN, et al. Myosarcomas of the stomach: Natural history, prognostic factors and management. Cancer 1982 Jan 1; 49(1):177-87.
7. Grant CS, Kim CH, Farrugia G, et al. Gastric leiomyosarcoma. Prognostic factors and surgical management. Arch Surg 1991 Aug; 126(8):985-90.
8. Fishbein T, Carroll M, Beazley RN. Duodenal leiomyosarcoma. J Surg Oncol 1995 Jan; 58(1):70-3.
9. Lindsay PC, Ordonez N, Raaf JH. Gastric leiomyosarcoma: clinical and pathological review of fifty patients. J Surg Oncol 1981; 18(4):399-421.
10. Bedikian AY, Khankhanian N, Valdivieso M, et al. Sarcoma of the stomach: clinico-pathologic study of 43 cases. J Surg Oncol 1980; 13(2):121-7.
11. Lee YT. Leiomyosarcoma of the gastrointestinal tract: general pattern of metastasis and recurrence. Cancer Treat Rev 1983 Jun; 10(2):91-101.
12. Conlon KC, Casper ES, Brennan MF. Primary gastrointestinal sarcomas: analysis of prognostic variables. Ann Surg Oncol 1995 Jan; 2(1):26-31.
13. Weaver D, Winczek R, Bowman D, et al. Gastrojejunostomy: is it helpful for patients with pancreatic cancer? Surgery 1987 Oct; 102(4):608-13.
14. Feretis C, Benakis P, Dimopoulos C, et al. Palliation of malignant gastric outlet obstruction with self-expanding metal stents. Endoscopy 1996 Feb; 28(2):225-8.
15. Kozarek RA, Brandabur JJ, Raltz SL. Expandable stents: unusual locations. Am J Gastroenterol 1997 May; 92(5):812-5.
16. Baron TH, Morgan DE, Yates MR. Use of expandable metal stents for malignant gastrointestinal luminal obstruction. Clin Perspectives in Gastroenterol Jan/Feb 1999:16-24.
17. Bethge N, Breitkreutz C, Vakil N. Metal stents for the palliation of inoperable upper gastrointestinal stenoses. Am J Gastroenterol 1998 Apr; 93(4):643-5.
18. De Baere T, Harry G, Ducreux M, et al. Self-expanding metallic stents as palliative treatment of malignant gastroduodenal stenosis. Am J Roentgenol 1997 Oct; 169(4):1079-83.
19. Soetikno RM, Lichtenstein DR, Vandervoort J, et al. Palliation of malignant gastric outlet obstruction using an endoscopically placed Wallstent. Gastrointest Endosc 1998 Mar; 47(3):267-70.
20. Yates MR 3rd, Morgan DE, Baron TH. Palliation of malignant gastric and small intestinal strictures with self-expandable metal stents. Endoscopy 1998 Mar; 30(3):266-72.