Gastrointestinal stromal tumour of the duodenum

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Introduction

Gastrointestinal tumours (GIST) are described as gastrointestinal mesenchymal masses expressing a proto-oncogene called CD 117, detected by immunohistochemistry. GISTS account for about 1-3% of all gastrointestinal malignancies. The clinical features of GISTS depend on the size and site of the tumour. Complete surgical excision is the treatment of choice for a localized GIST. The role of radiotherapy in the treatment of GISTs is limited. The anti-tumour effects of the molecular inhibitor imatinib mesylate in metastatic and inoperable GISTS have been a remarkable breakthrough in the treatment of GISTS.

Case Report

A 41 year old house wife from Jaffna was admitted with right sided upper abdominal pain for the last 6 months. She had episodic vomiting during the last 3 months and the vomitus contained bile-stained food particles. She had no haematemesis or melaena. Her past surgical history only included surgery for a perforated duodenal ulcer 6 years ago. Clinical examination revealed an intra-abdominal mass in the right lumbar region which was not pulsatile or ballotable. Other system examinations were normal.

Routine blood investigations were within normal limits. An ultrasound scan of the abdomen revealed a mass in the right para-aortic region measuring about 8.5 cm x 9.9 cm, with both solid and cystic areas. The pancreatic head was obscured by the mass, but other organs were found to be normal.

A contrast CT abdomen, pelvis and chest revealed a contrast enhancing mass with a solid and cystic nature in the 3rd part of the duodenum, measuring 7.5 cm x 7.4 cm. It seemed to obstruct the 3rd part of the duodenum and obscure the head of the pancreas, but had no pancreatic infiltration. The radiological appearance was suggestive of a tumour arising from the 3rd part of the duodenum without evidence of metastases.

The case was discussed at the multidisciplinary meeting and it was decided that a surgical excision of the tumour would be done. An upper midline laparotomy was carried out and the tumour was found in the third part of the duodenum without any macroscopic evidence of metastases to the peritoneum or liver. The tumour was completely resected with the 3rd and 4th part of the duodenum without any damage to the head of the pancreas of the 2nd part of the duodenum. A proximal jejunal loop was mobilized and anastomosed with the 2nd part of the duodenum and stomach. A gastro-jejunostomy was performed considering the possibility of avoiding long term strictures at the duodeno-jejunal anastomotic site (Figure 1).

Figure 1. Location of tumour in the duodenum.

Figure 2. Surgical anastomoses.

The histologic examination of the specimen revealed

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complete excision of the encapsulated tumour of the duodenum without any infiltration into the surrounding tissue. Histological features raised the possibilities of a Rhomboid tumour, Para-ganglioneuroma or a stromal tumour. However, the CD 117 staining was diffusely positive in tumour cells in keeping with a diagnosis of a GIST. The patient was referred to the oncologist and she is followed up in at the surgical & oncology clinic regularly without any further medication or surgery.

Discussion

GISTs are relatively uncommon tumours of the gastrointestinal tract. They arise from mesenchymal cells of the gastrointestinal tract. GISTs can be benign or malignant. Stromal tumours in the abdominal cavity are divided into extra gastrointestinal stromal tumours (omentum, peritoneum and mesentery) and gastrointestinal stromal tumours. These tumours are mostly sited in the stomach (45.0%), followed by the small intestine (32.0%), omentum & peritoneum (12.6%), large intestine (9.3%) and oesophagus (1.1%). GISTs usually appears as a single mass, but multiple masses have been reported in several papers [2,5].

It usually affects the middle age and elderly patients (median age group is 50 – 60 years). The clinical picture depends on the size and site of the tumour. These tumours can present with acute and chronic bleeding, abdominal discomfort, intestinal obstruction, projectile vomiting, dysphagia, altered bowel habit and as an externally palpable intra-abdominal mass.

A GIST is usually an encapsulated well-circumscribed mass, commonly a larger lesion and ulceration of the overlying mucosa is also present. The predominant histologic pattern is spindle cell type (70-80%). Expression of CD 117 is found in 95% of GISTs regardless of the site of the tumour. It also expresses CD 34 (50%), neuron specific endolase (43%),

smooth muscle actin (26%), desmin (4%) and S. protein (4%) [2,3].

GISTs can metastasize to the liver, bone, lungs and rarely to the lymph nodes [3]. The small intestine GISTs are more aggressive than others and are found to have metastases at the time of surgery [2,3]. It is difficult to predict the malignant potential of GISTs based on its' clinical and pathological features.

Confirmation of GISTs requires tissue histology with demonstration of CD 117 positivity. The computing tomography (CT) and magnetic resonance imaging (MRI) will help to determine the site, extent and metastatic sites of the disease. Complete surgical excision of the tumour is the treatment choice for localized GISTs and the role of radiotherapy is limited by potential toxicity to surrounding structures. GISTs are usually resistant to chemotherapy, but Imatinib mesylate (Glivec®) is a new drug which has an established role in the management of inoperable and metastatic stromal tumours.

References

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Key Point:

- The stromal tumours of the gastrointestinal tract should be diagnosed accurately.
- Surgical excision is the best option for localized tumours.
- Imatinib mesylate (Glivec) is a major break through which has an established role in the management of inoperable and metastatic GISTs.