CASE REPORT

GASTROINTESTINAL STROMAL TUMUOR- CASE REPORT AND A REVIEW OF THE LITERATURE

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ABSTRACT

Gastrointestinal stromal tumours (GIST) are rare mesenchymal tumours, comprising 1% of all GI malignancies. This is a report of a middle-aged man diagnosed with GIST of the duodenum. He had gastrojejunostomy and para-aortic lymph node biopsy for histology. He was subsequently commenced on 5-fluorouracil but with little response.

Keywords: *Gastrointestinal tract, stromal tumour.*

INTRODUCTION

Gastrointestinal stromal tumours (GIST) are the most common mesenchymal tumours of the gastrointestinal tract. They are rare tumours, comprising 1% of all GI malignancies[1]. They represent approximately 5% of all soft tissue sarcomas[1]. Majority of these tumours arise from cells that are not clearly of smooth muscle or neurogenic origin. They are slow growing, indolent tumours. Over the past few years the term gastrointestinal stromal tumours (GISTs) has been introduced to classify mesenchymal tumours arising in the luminal tract[2]. They are usually extraluminal in origin, but may ulcerate through the overlying mucosa. Virtually all GISTs have gain – of – function c – kit mutation (a protooncogene growth factor receptor)[3]. The objective of this case report is to sensitize clinicians to the possibility of GIST in patients presenting with dyspepsia.

Case report

A 49 year-old man who presented with recurrent upper abdominal pain of 9 years, and 6 months history of right hypochondrial swelling and weight loss. He also had history of early satiety and vomiting. No history suggestive of upper gastrointestinal bleeding. Clinical examination revealed wasting, pallor and ankle oedema.

He had hepatomegaly which was firm, irregular and tender. Succussion splash was positive. An impression of gastric outlet obstruction secondary to antral carcinoma to rule out carcinoma head of pancreas with hepatic metastasis was made.

Abdominal ultrasound scan revealed hepatomegaly and a mass distal to the stomach.

Peripheral blood film examination revealed iron deficiency anaemia and thrombocytosis, while serum chemistry showed hypoalbuminaemia.

An upper gastrointestinal endoscopy revealed a mass in the second part of the duodenum which was ulcerating and vascular. Multiple biopsies were taken for histopathology. The tissue was sectioned and stained with haematoxylin and eosin. It showed spindle shaped cells containing large nuclei with blunt edges and were surrounded with mild to moderate cytoplasm. There is loss of overlying mucosa . Features are in keeping with gastrointestinal stromal tumour .

Exploratory laparotomy revealed a mobile mass in the 2nd and 3rd parts of the duodenum which was firm-hard, irregular, measuring about 12cm in diameter. There were metastases to the liver and paraaortic lymph nodes. He had gastrojejunostomy and paraaortic lymph node biopsy for histology which also revealed a metastatic gastrointestinal stromal tumour.

Postoperatively, patient was stable and he was discharged home 11 days after . He is presently stable and on follow up .He has been commenced on cyclical cytotoxic chemotherapy using 5- fluorouracil.

DISCUSSION

Gastrointestinal Stromal Tumours (GISTs) are slow growing and as such are usually asymptomatic until they become quite large. About 30% of GISTs are detected during investigations for unrelated disease. The typical age at presentation is over 40 years {median age is 50-60 years} and there is male predominance.[4] Symptoms at presentation include gastrointestinal

bleeding, dyspepsia, and with large tumours, obstructive symptoms. In this case report, our patient presented with dyspeptic and obstructive symptoms. His peripheral blood film also revealed iron deficiency anaemia, probably due to occult gastrointestinal bleeding. Our patient is within the usual age of presentation. GISTs can spread into regional lymph nodes, liver, lung and peritoneum. Our patient had metastases to the paraaortic lymph nodes and the liver. The initial diagnosis of GISTs is best made at the time of endoscopy. Probably, if our patient had presented earlier and had upper GI endoscopy, the diagnosis could have been made before he reached this advanced stage.

The most common affected site is the stomach in about 70% of cases followed by the small intestine {25-40%). The colon , rectum and oesophagus are less affected [4] . In our patient the tumour was found in the second part of the duodenum .It was well circumscribed with distinct margins and surrounded the papilla. In a study conducted in pakistan , 20% of 205 leiomyosarcomas were abdomimal[5].

GISTs are most common in the jejunum , least common in the duodenum. Until 1981, 123 cases of duodenal GISTs were reported in a review of literature.[6] The endoscopic appearance varies depending on the size of the tumour and whether it is intraluminar , intramural or exoenteric. The tumour may be submucosal and develop a central ulceration as it enlarges. The ulcerated form would presumably be the most common, since bleeding is the chief complaint in over half of the cases.[7] In this case presentation the tumour was intraluminar and ulcerated, although there was no evidence of upper GI bleeding. The chief complaint in our patient was upper abdominal pain.

Endoscopic ultrasound {EUS}is also helpful in determining the depth of invasion as well as regional lymph node involvement [2]. Although, our patient did not have EUS done, regional lymph node involvement was revealed at laparotomy.

Immunohistochemical analysis is used for confirmation of diagnosis C-kit {CD117}and CD 34 have significant importance in identifying GISTs as these tumours have a strong affinity for the respective antibodies. These tumours also show immunopostivity for smooth muscle actin {SMA} and S-100 protein [8]. In our patient the diagnosis was reached by a combination of clinical presentation, endoscopic appearance and histology.

The most useful clinical predictor of outcome is the mitotic index of the tumour. Those with a mitotic rate of greater than 2 per 10 high – power fields have a much higher risk for recurrence or metastases [9]. The size of the tumour is also a clinical predictor of outcome, with those larger than 5cm at higher risk of metastases [9]. GISTs of less than 2cm with negligible mitotic activity are considered very low risk, they may still metastasize [11]. Our patient had an extensive tumour, greater than 5cm and hence metastases to the liver and paraaortic lymph nodes.

Surgery is the treatment of choice. Some advocate use of adjunctive radiation and chemotherapy[2]. Recently, the tyrosine kinase inhibitor imatinib mesylate (ST 157, Gleevec) has been used successfully in a metastatic GIST.[10] Monitoring the patient long after resection is necessary as the tumour has a tendency to recur. Our patient was offered imatinib, but could not afford it because of cost, hence the choice of 5-fluorouracil.

CONCLUSION

GIST of the duodenum is an uncommon cause of gastric outlet obstruction. Our patient presented late with attending poor prognosis. The use of cytotoxic chemotherapy has not been found to be of proven efficacy. The cost of imatinib has placed it out of reach of use for patients in our environment.

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