Unusual presentation of jejunal gastrointestinal stromal tumour with large metastatic deposits in the liver and peritoneum: Advantageous surgical exploration

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Summary

Gastrointestinal stromal tumour (GIST) is a relatively rare neoplasm accounting for less than 3% of all malignant gastrointestinal tumours. The diagnosis of GIST is often delayed due to its indolent symptoms. Patients present in advanced stages with metastatic deposits and at times unresectable stage.

This report presents a case of a jejunal GIST with subsequent metastasis to the liver and multiple peritoneal deposits in a 59-year-old female patient managed surgically.

Keywords - gastrointestinal stromal tumours (GIST), metastasis, surgery, treatment outcome

Introduction

GISTs are the most common mesenchymal tumours of the gastro-intestinal system, although they represent less than 3% of all gastrointestinal tract malignancies[9]. Most GISTs arise from the stomach (50-70%) and small intestine (20-30%)1.

The clinical symptoms associated with small bowel GISTs are usually nonspecific and varied[3]. This results in late presentation with advanced disease. Around 10-25% of patients present with metastatic disease, the most common sites being the liver (65%) and peritoneum (21%)[2]. GISTs express a KIT protein (CD117) as its characteristic feature, which is confirmed by immunohistochemistry. Early intervention and surgical exploration of metastatic GISTs are associated with prolonged survival[1].

This report presents a case of operating a GIST with liver metastases and multiple peritoneal deposits in a 59-year-old female patient who presented with symptomatic anaemia.

Case Report

A 59-year-old female patient presented with anaemic features for two weeks having a haemoglobin level of 7.3 g/dl. Her stool was positive for occult blood. She had a history of recurrent vomiting and reflux symptoms for the last one and half years.

Her abdominal examination was significant with a non-tender nodular right hypochondrial mass.

Non-contrast Computed Tomography (CT) abdomen revealed a 63 x 8 x 75 mm well-defined lesion in segment VII of the liver. There was a 20 x 145 mm calcific focus and an area of central non-enhancing hypo-attenuated area suggestive of necrosis (Figure 1).

The patient underwent a core biopsy and histopathology revealed a malignant spindle cell tumour which was confirmed as GIST with strong CD117 immunoreactivity.

The patient was pre-operatively optimized and offered right hepatectomy and resection of multiple peritoneal metastases with small bowel resection.

Peritoneal cavity was accessed through a long midline laparotomy with right lateral extension. There was a large liver deposit involving segment VII and VIII adherent to the diaphragm (Figure 2) with omental and peritoneal nodules (Figure 3). The patient underwent right hepatectomy with resection of metastatic nodules and resection of small bowel. Other deposits were removed by diathermy.

Recovery was uneventful following surgery. Patient was discharged after nine days from admission. Subsequent histopathology report confirmed the diagnosis of small bowel GIST with multiple peritoneal deposits and liver metastasis.

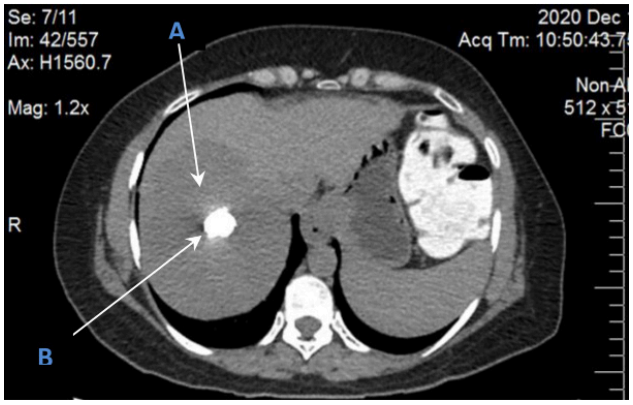

Figure 1: Axial slice of the CT abdomen demonstrates 63 x 8 x 75 mm well-defined lesion in segment VII of liver (A) with 20 x 14 mm calcific focus and area of central non-enhancing necrotic area (B).

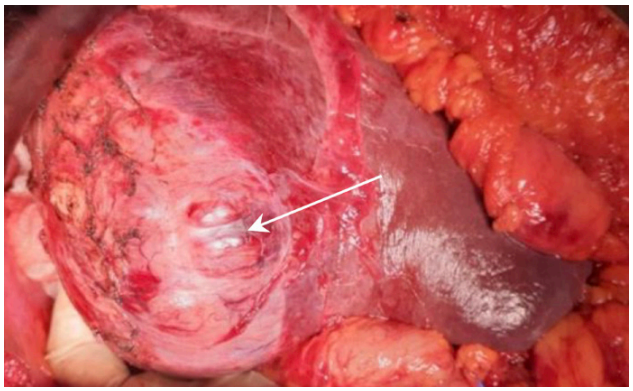

Figure 2: Well-defined metastatic lesion in right lobe of liver involving segment VII & VIII adherent to the diaphragm. The mass has no attachment to nearby structures.

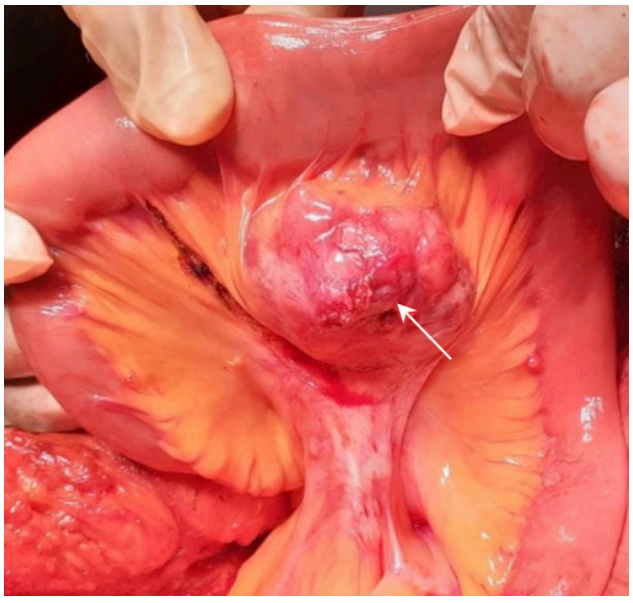

Figure 3: A metastatic deposit in the mesenteric border of the small bowel.

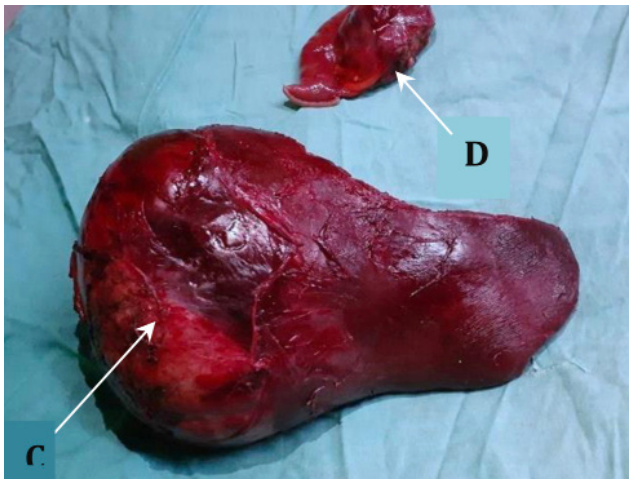

Figure 4: The resected specimen. Metastatic deposit in the segment VII of the liver, measuring 63 x 8 x 75 mm in diameter (Arrow C). Resected mesenteric nodule (Arrow D).

Discussion

GISTs with liver and peritoneal metastases are extremely rare in literature. Its' incidence and prevalence has been under-recognized and its' devastating disease course is underestimated[7]. Sreeramulu et al. in 2017 reported a series of 11 cases concluding the uncommon and rare presentation of GISTs showing different presentations owing to its anatomical location. Nisson B et al. performed a population-based study and reported that forty-four percent of symptomatic, clinically detectable GISTs were categorized as high risk (29%) or overtly malignant (15%).

Surgical intervention for GIST is the preferred choice of treatment for localized, resectable GIST. In some instances 'debulking surgery' is used for advanced metastatic disease for symptomatic treatment[3]. The 5-year survival rate after surgical resection has been reported as 30%–65% among patients with GISTs[4,10].

DeMatteo et al., who evaluated a series of 200 GISTs, have shown median survival as 66 months following complete resection. There is some evidence that metastasectomy can improve survival in selected patients. Chen H. et al. reviewed 11 consecutive patients who underwent resection of isolated metastases concluding that median survival of all patients after liver resection was 39 months.

Based on this evidence, the patient was offered small intestinal tumour resection and metastasectomy with right side hepatectomy for a favourable outcome.

Following successful surgical resection, further improvement of 5-year survival is targeted with adjuvant chemotherapy.

Before imatinib mesylate was introduced, resected GISTs had a higher rate of recurrence and a failure with 5 year survival of 28-35%[6]. Survival of advanced GIST has improved following increased access to tyrosine kinase inhibitors[8].

Zhu J. et al. proposed a two year survival rate of 95.2% in the patients who had only a liver-metastatic GIST after the prior radical resection combined with the treatment of imatinib mesylate. Based on this evidence, the patient was started on imatinib mesylate and is on long term follow-up.

Conclusion

GISTs with peritoneal and liver metastasis are extremely rare but can be operated safely with a favourable surgical outcome.

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