CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 37 (2017) 17–21

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Extragastrointestinal stromal tumour of the lesser omentum: A case report and literature review

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ARTICLE INFO

Article history:
Received 29 January 2017
Received in revised form 28 May 2017
Accepted 29 May 2017
Available online 8 June 2017

Keywords:
Extragastrointestinal stromal tumour
Lesser omentum
Abdominal mass
Case report

ABSTRACT

INTRODUCTION: Extragastrointestinal stromal tumours (EGISTs) are very uncommon compared to their gastrointestinal counterparts. Most of them originate from the intestinal mesentery and the omentum.
CASE REPORT: A 70 year-old Caucasian woman presented with a bulky abdominal mass which on laparotomy was found to originate from the lesser omentum and was completely resected. Histological examination revealed spindle cells with severe pleomorphism and high mitotic activity. Immunohistochemically, the tumour cells showed strong positivity for c-kit (CD117), DOG-1 and human haematopoietic progenitor cell antigen (CD34). An exon 11 deleterious mutation was identified and thus regular dosing of 400 mg imatinib mesylate was initiated.
DISCUSSION: There have been only a few previous reports of EGISTs arising in the lesser omentum. Although EGISTs seem to have morphological and immunohistochemical similarities with GISTs, their pathogenesis, incidence, genetic background and prognosis are not completely known because they are extremely rare. It is strongly believed that such tumours originate from cells, which have similar pathological characteristics and biological behaviour as the intestinal cells of Cajal. In most series of EGISTs, a female predominance, a greater size and a higher mitotic index than GISTs were observed.
CONCLUSION: EGISTs are very rare mesenchymal tumours which originate from cells outside the gastrointestinal tract and tend to have a more aggressive biological behaviour than their GI counterparts. Complete surgical resection is the most effective treatment associated with the use of imatinib in the presence of adverse prognostic factors. In any case a strict follow-up is necessary due to high recurrence rates.

1. Introduction

Gastrointestinal stromal tumours (GISTs) represent a distinct pathological entity that has been recognised since the discovery of c-kit (CD117) in 1998 [1]. GISTs are nonepithelial, mesenchymal tumours, which may occur in all sites of the gastrointestinal (GI) tract, but most commonly affect the stomach and the small intestine. They constitute 1–2% of all GI neoplasms and they arise from the intestinal pace-maker cells of Cajal or their stem cell precursors as a result of oncogenic mutation in the KIT tyrosine kinase [2].

GISTs are rarely found as primary tumours in extragastrointestinal tissues. Extragastrointestinal stromal tumours (EGISTs) are very uncommon compared to their gastrointestinal counterparts and typically are not connected to the walls or serosal surfaces of gastrointestinal tubular organs. Most of them originate from the intestinal mesentery and the omentum but there have been sporadic reports of EGISTs in other sites such as retroperitoneum, liver, hepatobiliary tree, pancreas, spleen, uterus, vagina, inguinal hernia sac, rectovaginal septum, ovary, pleura, pericardium, prostate, urinary bladder, scrotum, seminal vesicles and abdominal wall [3–35]. Although EGISTs seem to have morphological and immunohistochemical similarities with GISTs, their pathogenesis, incidence, genetic background and prognosis are not completely known because they are extremely rare [6].

We report the interesting case of an EGIST located in the lesser sac and review the relevant literature.

2. Case presentation

A 70 year-old Caucasian woman presented to our department with symptoms of early satiety and epigastric fullness. Physical examination revealed a mass in the left upper abdominal quadrant. The results of laboratory tests including complete blood count, amylase, liver function tests, and all tumour markers were within normal range.

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http://dx.doi.org/10.1016/j.ijscr.2017.05.040

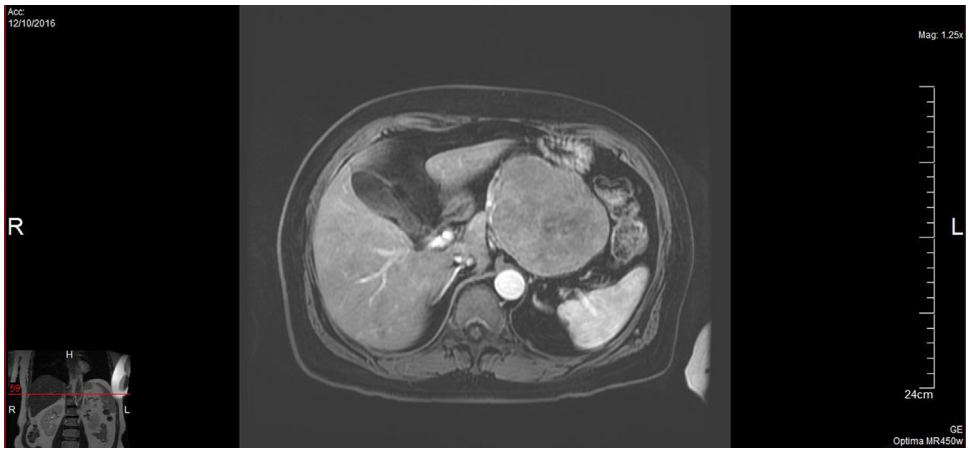

Fig. 1. MRI abdominal scan showing a large solid mass between the left hepatic lobe, the stomach and the retroperitoneum.

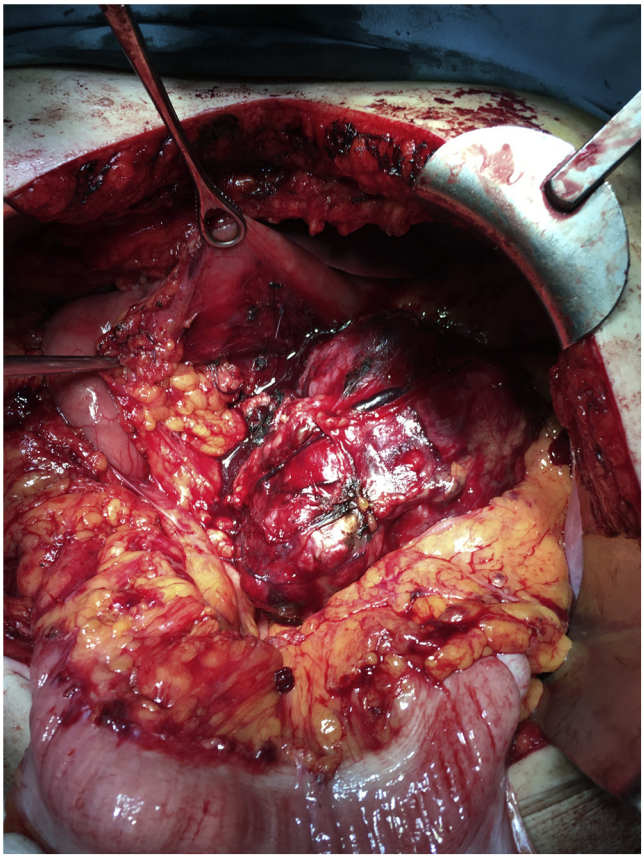

Fig. 2. Intraoperative exposure of lesser sac containing the tumour.

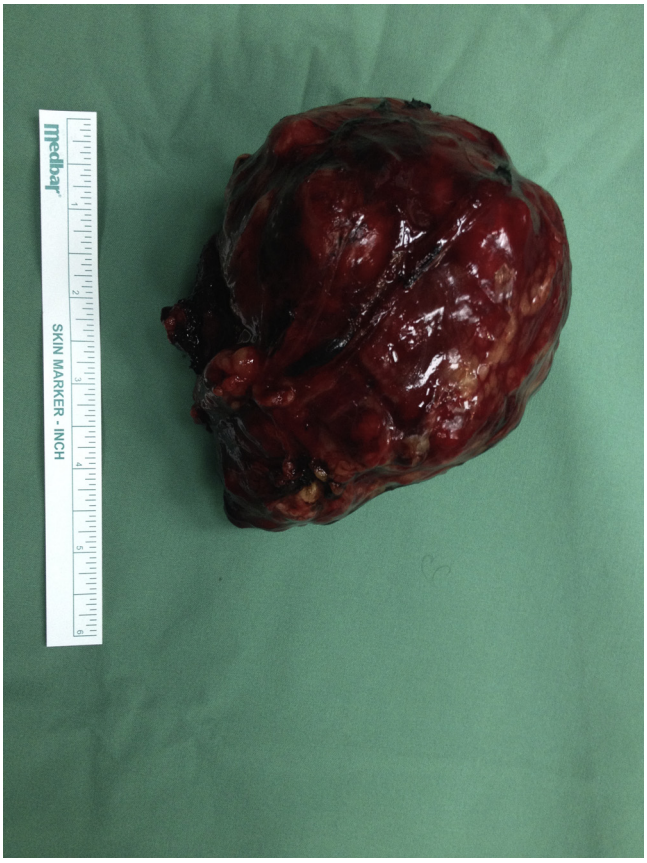

Fig. 3. The specimen measured approximately 5 inches (equal to 12.7 centimetres).

A computed tomography scan (CT scan) and a magnetic resonance imaging (MRI) of her abdomen both showed a large mass (maximum diameter 12 cms) that was confined between the left hepatic lobe, the stomach and the retroperitoneum (Fig. 1). An endoscopic ultrasound guided fine-needle aspiration was performed and cytology was consistent with a stromal tumour.

On exploratory laparotomy, after entering the lesser sac a large solid tumour was found located posterior to the gastric wall and anterior to the pancreas (Fig. 2). There was a clear plane of dissection without invasion of either organ and a complete resection of the mass was performed with safety (Fig. 3). It was assumed that the tumour originated from tissues of the lesser omentum. Neither metastatic liver lesions nor lymphadenopathy were observed. The postoperative course was uneventful and the patient was discharged after 6 days.

Histological examination revealed spindle cells with severe pleomorphism (Fig. 4) and high mitotic activity (mitotic count of 8 mitoses/50 high-power fields). Immunohistochemically, the tumour cells showed strong positivity for c-kit (CD117) (Fig. 5), DOG-1 and human haematopoietic progenitor cell antigen (CD34). Immunostains for desmin and smooth muscle actin (SMA) were negative. The expression of Ki67 protein was 5%. Considering tumour's size, its morphology and high mitotic index, the estimated risk of recurrence after surgery was high and subsequently the patient was referred to a medical oncologist for further management. Molecular analysis of c-kit (exons 9,11,13,14, 15, 16, 17)