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Case Report

Primary extragastrointestinal stromal tumor of greater omentum misdiagnosed as an adnexal tumor: a case report

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are the most frequently occurring mesenchymal tumors in the gastrointestinal system. These tumors are believed to arise from interstitial cells of Cajal, the gastrointestinal pacemaker cells, due to the presence of tyrosine kinase receptors in their tissue. When tumors with similar morphological and immunohistochemical properties are found outside the gastrointestinal tract such as the mesentery, retroperitoneum, and every now and then the omentum, they are classified as extragastrointestinal stromal tumors (EGISTs). We report a case of a primary epithelioid (EGIST) of the greater omentum, the tumor was diagnosed in women with non-specific symptoms who presented for abdominal pain initially misdiagnosed as an adnexal tumor. This report highlights the need to consider EGIST in the differential diagnosis of abdominal and pelvic tumors and underscores the importance of EGIST-specific studies to evaluate treatment modalities for improved oncological outcomes and quality of life.

Keywords: Extragastrointestinal stromal tumor, Greater omentum, Oncology, Surgery

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) typically develop within the digestive tract, believed to originate from the interstitial cells of Cajal. In the digestive tract, GISTs are most commonly found in the stomach (60-70%), followed by the small intestine (20-25%), colon (5%), and esophagus (5%).[1]

However, a limited number of reports have described GISTs occurring outside of the gastrointestinal tract, known as extragastrointestinal stromal tumors (EGISTs).

The incidence of EGISTs in young adults under the age of 40 is exceptionally rare. The clinical progression of these tumors is typically insidious due to their slow growth rate, often resulting in delayed diagnosis. This uncommon subtype has been documented in only a limited number of case reports.[2]

We present a case of primary epithelioid EGIST in the greater omentum, diagnosed in a woman with non-specific symptoms, including abdominal pain, initially misinterpreted as an adnexal mass.

CASE REPORT

A 78-year-old female patient, with no personal either familial significant past medical history, presented to the emergency department with an abdominal bloating, heaviness, that had been persisting for the past two months without any particular medical or drug history.

Physical examination was non-revealing, a soft abdomen was noted with a palpable mass in the upper right quadrant, which was non-tender on palpation.

Laboratory investigations were normal. However, imaging studies including pelvic ultrasound and a pelvic magnetic resonance imaging (MRI) revealed voluminous

abdominopelvic solid cystic mass, probably adnexal, 104×62×160 mm and peritoneal carcinosis in the right iliac fossa (Figures 1a-d).

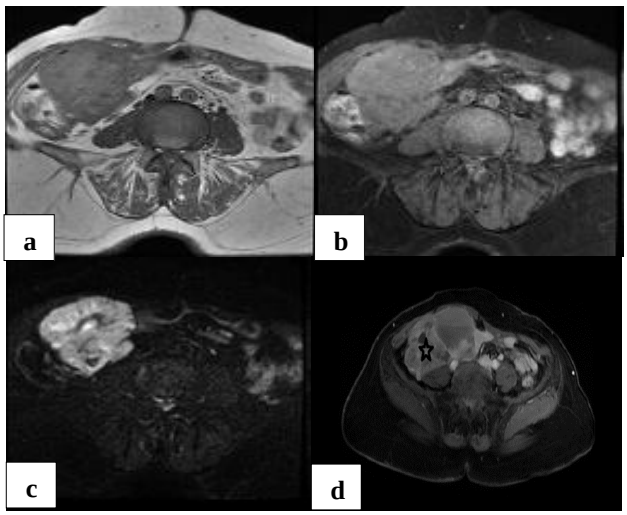

Figure 1: Magnetic resonance imaging (a) low-intensity mass with a high-intensity surface on T1-weighted imaging (T1WI); (b) heterogeneous high-intensity mass on T2-weighted imaging (T2WI); (c) diffusion-weighted imaging (DWI); and (d) a decreased gadolinium enhancement area on the posterior surface of tumor.

Given these radiographic findings, an exploratory laparoscopy was performed. Intraoperatively, we identified Friable right large mass with irregular contours in the right iliac fossa, the origin of which is difficult to identify, multiple nodules of carcinosis in the epiploic area nodules in the stomach, nodules in digestive tract with a Fagotti score ≥8. Biopsy of the mass, carcinosis nodules and epiploon nodules were performed.

Due to inconclusive histopathological examination results we opted for an exploratory laparotomy. Intraoperatively we identified a 16 cm mass with irregular, heterogeneous contours, highly vascularized and highly suspicious, which appears to be dependent on the greater omentum, forming an adherent magma with the intestine and greater omentum. With several nodules of peritoneal carcinosis (Figure 2).

We performed a cautious adhesiolysis and a cautious resection of the mass (Figure 3). The two ovaries appeared macroscopically free of any abnormalities.

Histopathological examination of the resected specimen confirmed the diagnosis of GIST.

The tumor was composed of bundles and fascicles of spindle-shaped cells, with nuclei that were blunt-ended and showed prominent cytoplasmic vacuolation. Immunohistochemical staining demonstrated strong membranous positivity for DOG1 and CD117/C-kit expression in GIST, with cytoplasmic and membranous labelling.

The postoperative course was uneventful with no complications. The patient was discharged home on the second postoperative day.

Following a multidisciplinary consultation and considering the metastatic nature of the tumor, it was decided to continue lifelong imatinib therapy.

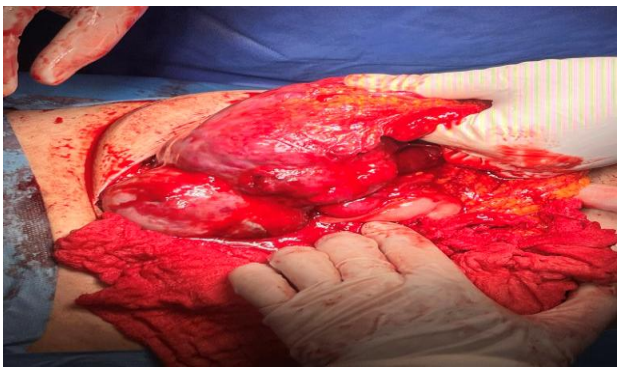

Figure 2: Intraoperative images showing the tumor attached to the greater omentum with no signs of connection with adjacent organs.

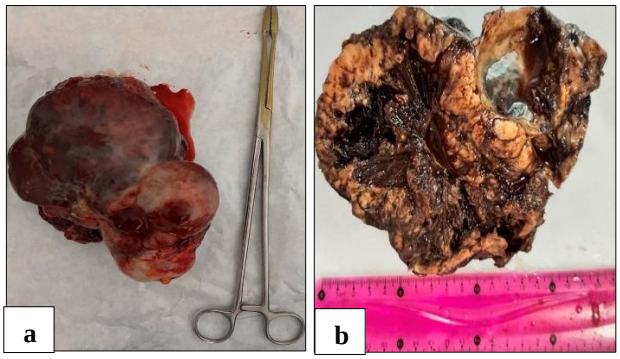

Figure 3: Post-surgical excision of EGIST (a) the mass measuring 18×10 cm, and (b) internally, the tumor was divided by fibers and filled with necrotic tissue.

DISCUSSION

EGISTs arise outside the gastrointestinal tract but share histological features similar to GISTs. Unlike GISTs, whose clinical, pathological, and prognostic characteristics are well established in literature, EGISTs remain poorly understood. Their incidence, histogenesis, and histological predictors of prognosis have yet to be clearly defined.[3]

GISTs are malignant mesenchymal tumors arising in the wall of the digestive tract, most often in the stomach and small intestine, more infrequently in the rectum, colon and oesophagus. Histologically, GISTs have the phenotype of interstitial cells of Cajal.[4] Although they are the most common mesenchymal tumors of the digestive tract, they

are thought to account for only around 1% of all GI malignancies.[5]

EGISTs mainly affect the omentum, mesentery and retroperitoneum. Rare cases involve the pancreas, gallbladder, liver, abdominal wall, pharynx and posterior mediastinum.[6]

EGISTs show similarities to GISTs, but research has identified distinct variations in their characteristics. GISTs originate from interstitial Cajal cells in the intestinal wall, whereas EGISTs are derived from either pluripotent stem cells or Cajal-like cells found outside the gastrointestinal tract. The physical size and primary location of EGIST determine its clinical presentation. Pain in the abdomen and the presence of an abdominal mass are the most common symptoms, while gastrointestinal bleeding due to mucosal erosion is uncommon. As a result, EGIST is often diagnosed at a more advanced stage, often with metastases, or mistakenly diagnosed as a pelvic mass, as in our patient's case.[7,8]

CT is the usual diagnostic tool for large tumors, and more rarely for EGIST of limited diameter. The diagnosis of EGIST is confirmed by histology.

In terms of histology, these tumors can be divided into three main types: the mixed type (10%), the spindle cell type (70%) and the epithelioid type (20%). In our case, histological examination of the resected specimen showed that the tumor was composed of fascicles and bundles of spindle cells. These cells showed marked cytoplasmic vacuolization and blunt-ended nuclei.[9]

The CD117 antigen, a byproduct of the protooncogene c-KIT and a characteristic of GIST with almost universal expression (>95%), has been identified thanks to immunohistochemistry. But later on, a more sensitive and specific marker known as DOG-1 (found on GIST 1), or anoctamin1, was found and turned out to be a useful tool. It is especially helpful in confirming the diagnosis in this subgroup because it is robustly expressed in more than 99 percent of GISTs, including the majority of KIT-negative cases. There have also been reports of positivity for CD34 (70%) smooth muscle actin (30–40%) S-100 (5%), desmin (2%), and cytokeratins (2%).[4,9]

Surgery is the main treatment for non-metastatic EGIST of the greater omentum, although there is no consensus on adjuvant therapy. In these cases, chemotherapy and radiotherapy have limited efficacy. Given the risk of recurrence, particularly in large tumors with Kit alterations, Imatinib mesylate may be considered as part of the treatment plan. Treatment decisions must be individualized according to patient factors and tumor characteristics.[10]

The patient has undergone successful open surgery for tumor resection. After surgery, treatment with imatinib was initiated and continued for six months. Follow-up assessments at showed significant clinical improvement, with no evidence of recurrence.

CONCLUSION

As shown by our patient's distinct presentation, our case report underlines the rarity of GIST in the greater omentum. Even though they are rare, EGISTs should be included in the differential diagnosis of intra-abdominal masses. Further investigation and collaborative initiatives are needed to expand our knowledge of EGISTs located in the greater omentum, in order to pave the way for better diagnostic and treatment strategies in similar cases.

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