The Journal of Obstetrics and Gynecology of India (May–June 2023) 73(3):282–284
https://doi.org/10.1007/s13224-023-01743-1

CASE REPORT

An Unusual Presentation of Gastrointestinal Stromal Tumor as an Ovarian Mass: A Diagnostic Dilemma

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Received: 30 June 2022 / Accepted: 26 February 2023 / Published online: 17 March 2023

Introduction

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors of the gastrointestinal tract arising from interstitial cells of Cajal and account for 1–3% of all gastrointestinal (GI) malignancies [1]. They commonly arise from the stomach (50–70%) and small intestine (20–30%), including the duodenum, jejunum, and ileum. Only few cases of extra gastrointestinal stromal tumors (EGISTs) in the omentum and the mesentery have been reported in the literature [2].

GIST usually presents with nonspecific clinical symptoms and may sometimes present as a pelvic mass, especially in females. Differentiating a GIST from a pelvic mass by radiological studies is challenging. Thus, preoperative diagnosis of GIST may be difficult, especially in the case of an atypical pelvic mass.

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Case Report

A 42-year-old female, P5L5, came to the outpatient department with complaints of lower abdomen pain for 3 months and heaviness in the right lower abdomen for 1 month. The pain was insidious in onset, dull aching, progressive, radiating to the lower back, and was not relieved on taking analgesics. She had history of menorrhagia for 6 months, underwent endometrial biopsy and was on medical management outside. However; poor compliance to treatment was there. She had no history of ovarian, breast, or gastrointestinal cancers in the family.

On examination, her general condition was fair and was hemodynamically stable. She was obese (BMI-31.8 kg/m^2). On palpation; tenderness was present in the right iliac fossa. However, there was no palpable lump or organomegaly. On per speculum, the cervix was found to be hypertrophied, and few nabothian follicles were present. On bimanual pelvic examination, uterus was anteverted, bulky in size; mobile and right fornix fullness was felt. A vague cystic mass separated from the uterus was felt and the left fornix appeared free. On per rectal examination, rectal mucosa was free.

On routine evaluation, she was B positive with Hb of 13.2 gm/dl. All other investigations including renal function test, tumor markers; CEA (2.66 ng/dl), CA-125 (11. 9ul/ml), Beta-HCG (0.4 mU/ml), and alpha-fetoprotein (12 ng/ml) were within normal limits. The Pap smear was satisfactory and was negative for intraepithelial malignancy. Her transabdominal ultrasound revealed well-defined heteroechoic area of size 7.9 × 4.8 cm noted in the right iliac fossa in the peritoneal cavity. Another similar lesion m/a 4.2 × 3.1 cm was noted adjacent to the above lesion. The uterus was normal in shape and measured 12.6 × 6.8x 5.4 cm. The left ovary was normal; however, the right ovary was not visualized. CECT whole abdomen was suggestive of a large well-defined enhancing lesion in the right adnexal region suggestive of right ovarian neoplasm without any significant lymphadenopathy as shown in Fig. 1.

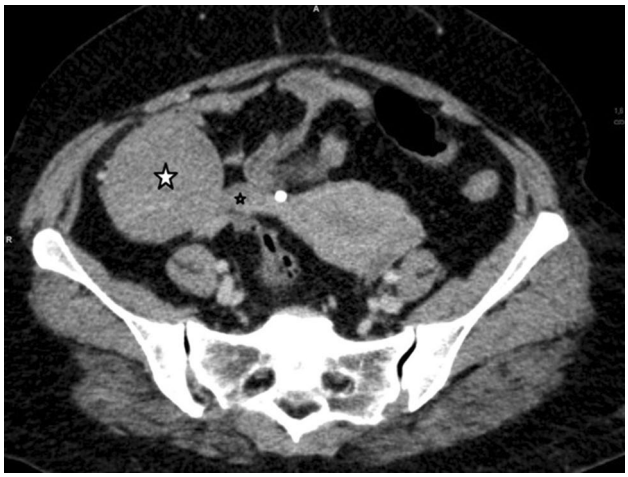

Fig. 1 CECT whole abdomen axial section of pelvis showing mildly enhancing soft tissue density solid mass in right adnexal adjacent to right ovary

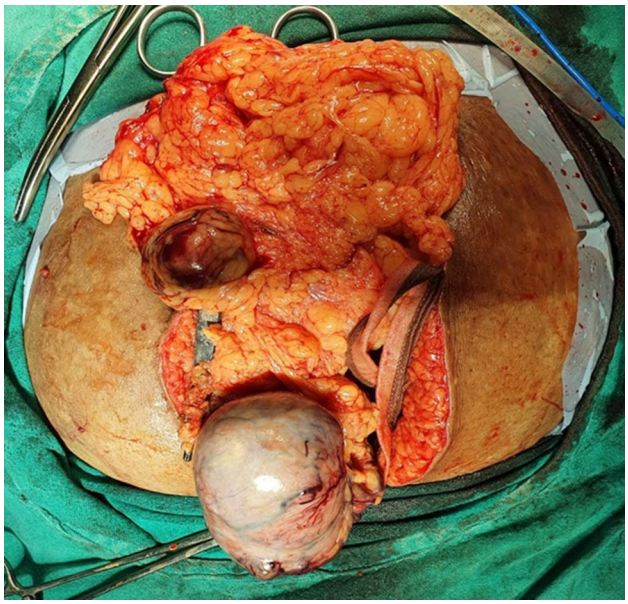

Fig. 2 Intraoperative image showing both omental masses

In concurrence with surgeons, an exploratory laparotomy was planned with a preoperative diagnosis of right ovarian tumor. The abdomen was opened through a midline vertical incision. A large mass of approximately 8 × 6 cm was noticed in the omentum lying in the right iliac fossa adherent to right ovary as shown in Fig. 2. Another adjacent omental mass of app 4 × 3 cm was seen. Gross systemic inspection and palpation of organs revealed a bulky uterus with both ovaries firm and mildly enlarged. Pouch of Douglas, cecum, ascending colon, hepatic flexure, transverse colon, splenic flexure, descending colon, and sigmoid colon appeared free. On the anterior surface of the liver, a nodule 3 × 3 cm was felt in the segment VIII which was widely resected and sent for frozen section examination. On retroperitoneal dissection, no abnormal lymph nodes were seen. Both the omental masses were resected and sent for frozen section examination which revealed features favoring malignancy. This was followed by total Omentectomy. Resection of the liver nodule, pelvic peritoneal deposits were done. A decision of total abdominal hysterectomy with bilateral salpingo-oophorectomy was taken since there was intraoperative diagnostic dilemma as the omental mass was adherent to the right ovary and frozen section reported presence of nonspecific malignant cells.

Histopathological examination revealed multifocal extra gastrointestinal stromal tumor, mixed type. A high grade in nature, its margins were partially capsulated with focal capsular deposits & margins were involved focally. Peritoneal deposit and liver also showed presence of tumor cells. Uterus with the cervix and both the ovaries were free of tumor cells. However; uterus had focal adenomyotic changes. Tumor cells were positive for CD117, VIMENTIN and negative for CD 34, S100, Pan ck, desmin, and Epithelial membrane antigen (EMA).

The postoperative period was uneventful. After multidisciplinary tumor board discussion which included histopathologist, radiologist, radiation oncologists, GI surgeons, and medical oncologists, the patient was started on imatinib due to the large size and high grade of tumor. On follow-up, she remains asymptomatic and disease-free at the end of 6 months and was advised for further follow-up visits every 6 months.

Discussion

GISTs are the most common mesenchymal tumor of the gastrointestinal tract. They arise from the intestinal cells of Cajal and express the CD117 antigen (c-Kit) and gain functional mutations responsible for the growth of these tumors. They usually occur after 40 years of age with a slight male predilection. Clinically, most of the patients are asymptomatic although large lesions may present with symptoms such as vague abdominal pain, bloating, abdominal fullness, hematemesis, or melena. Sometimes, they may be just an incidental finding during laparotomy or other radiological imaging. Differential diagnosis of GIST includes leiomyoma, ovarian tumor, schwannoma, solitary fibrous tumor, and mesenteric cyst.

Most patients exhibited normal serum levels of tumor markers including CA-125 levels. GISTs do not have typical features of ultrasound; thereby making contrast-enhanced CT (CECT) the imaging of choice for the evaluation of such masses. GISTs range in size from 1 mm to very large

tumors, occasionally measuring greater than 20 cm. They are well-circumscribed, intramural lesions, centered within the muscularis propria and on cut sections appear fleshy, tan-pink, which may show cystic degeneration or hemorrhage. Microscopically GISTs are classified into three morphological types: spindle cell type (70%), epithelioid type (20%), and mixed type (10%). On immunohistochemical staining, 95% are CD117 (c-kit) positive, 70% are CD34 positive, and 40% stains are positive for α-smooth muscle actin (α-SMA) and negative for desmin and S100 protein.

Complete surgical resection of the primary tumor remains the gold standard treatment for localized primary GIST. Lymphadenectomy is not routinely required for GIST unless lymph node metastasis is clinically suspected. In patients with low-risk GIST, the complete surgical resection is enough, and postoperatively regular clinical follow-up is recommended. GIST with intermediate-risk/and or high risk needs further treatment with a tyrosine kinase inhibitor to prevent recurrence and metastasis. Oral Imatinib 400 mg once daily is the drug of choice in such cases.

Preoperative diagnosis of GIST is difficult because of its nonspecific symptoms and clinical presentation, which occurred in this case as well. A study by Ijeri et al. reported a series of ten patients who presented with a presumptive diagnosis of an ovarian mass in the gynecology department and were finally diagnosed to have a GIST [3]. Shrestha S et al. reported a case where a large GIST was diagnosed as a malignant ovarian tumor preoperatively, however, later on, a diagnosis of jejunal GIST was made [4].

Conclusion

Whenever there is a disparity between clinical presentation, tumor markers and imaging; diagnostic laparoscopy and biopsy are the options. Frozen section, histology, and immunohistochemistry remain the gold standard for definitive diagnosis in cases with atypical presentation. This case also highlights that chemotherapy can cure lymphomas, and lymphadenectomy in such tumors is done only for enlarged nodes.

Acknowledgments None.

Funding No funding was done.

Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Approval All procedures followed were in accordance with the ethical standards of responsible committee on human research (institutional and national) and with the Helsinki declaration of 1975, as revised in 2008.

Informed Consent Written informed consent was obtained from the patient, as well as relatives, for participation in study and publication.

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