Turk J Gastroenterol *2015; 26: 56-9*

Gastrointestinal stromal tumor and leiomyoma of the ileum mimicking adnexal mass: A report of two cases

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ABSTRACT

Adnexal masses are formations seen in women of all ages; they most often include cystic elements. Medical history, physical examination, different imaging methods, and tumor marker determinations must be used together for preoperative evaluation of an adnexal mass. Both benign and malignant tumors of the small intestine are more rarely encountered than malignant tumors of other gastrointestinal system components; although advanced imaging methods and other diagnostic techniques are used, they do not always allow these tumors to be differentiated from adnexal masses. We report here on two cases operated on with the preliminary diagnosis of an adnexal mass, in which the presence of a gastrointestinal stromal tumor and a leiomyoma of the ileum, respectively, was established.

Keywords: Adnexal mass, small intestine tumor, GIST, ileal leiomyoma

Case Report

INTRODUCTION

Adnexal masses are formations seen in women of all ages; they most often include cystic elements (1). History, physical examination, different imaging methods, and tumor marker determinations must be used together in the preoperative evaluation of an adnexal mass (2). The primary imaging method used to evaluate adnexal masses is ultrasonography (USG). Especially after menopause, cases in whom USG characterizes tumor morphology as solid or complex, accompanied by elevated CA-125 levels (>35 U/mL), must be recognized as being at high risk for an ovarian malignancy.

Malignant tumors of the small intestine are less frequent than other tumors of the gastrointestinal system (GIS) (3). Gastrointestinal stromal tumors (GIST), which make up 1% of all GIS tumors, are rare (4). They are most frequently encountered in the stomach, and their second most frequent location is the small intestine (5). Leiomyomas are the most frequently observed benign tumors of the small intestine (6). Often, these generally asymptomatic tumors are only diagnosed post-mortem. Their most frequent location is the jejunum.

The patients presented here were both operated on with a preliminary diagnosis of adnexal tumor following preoperative physical examination and imaging diagnostics; however, the operation showed the presence of GIST in one case and leiomyoma of the ileum in the other.

CASE PRESENTATIONS

Case 1

A 54-year-old woman, postmenopausal for 10 years, was admitted to our clinic with a random finding of an adnexal mass at a routine examination. Her surgical history included an operation for breast cancer. Routine blood chemistry, complete blood count and tumor markers were within normal limits in this asymptomatic patient. Serum CA-125 level was 32.5 U/mL. A hard, immobile mass in the pelvic median line, with irregular contours, was palpated at physical examination. A heteroechogenic mass of 75x100 mm, appearing to be solid-cystic with irregular borders and thick septa, was observed in the left adnexal area on USG examination. It was determined to be an adnexal mass via magnetic

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Received: June 29, 2012 **Accepted:** October 02, 2012
 • Available online at www.turkjgastroenterol.org • DOI: 10.5152/tjg.2015.3833

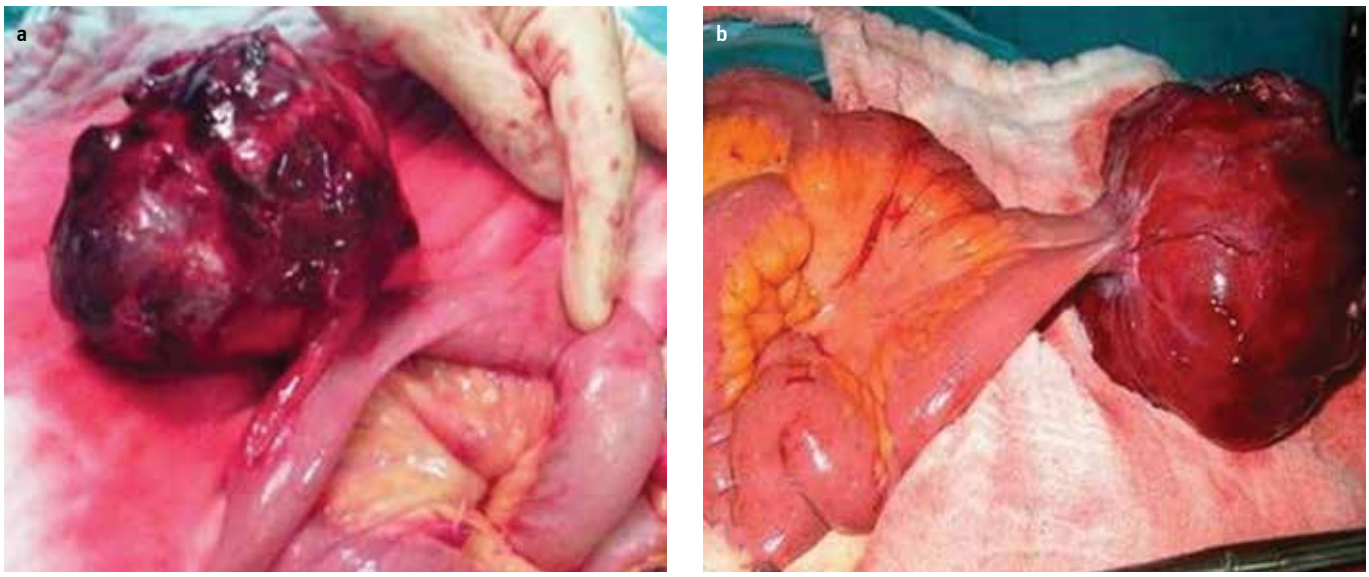

Figure 1. a, b. Laparatomic appearance of the GIST **(a)** and leiomyoma **(b)** of the ileum.

resonance imaging, similar to the USG examination results. Laparatomy showed a solid tumor 11 cm in diameter arising from the small bowel loops, mainly ileal, adhering to surrounding tissues and to the left adnexal area (Figure 1a). The uterus and ovaries appeared to be atrophic. The small intestine was anastomosed termino-terminally and the mass was excised. On macroscopic examination, the maximum diameter of the tumor was 11.2 cm, and it was mainly located in the muscularis propria but extended to the serosal surface as well. On histologic examination, the tumor was composed of spindle and epithelioid cells, with a hypercellular appearance, containing 5 mitoses per 50 high-power fields (HPF). The immunhistochemical profile of the tumor was CD117(+), CD34(-), desmin(-), smooth muscle actin(-), S-100(-), ki-67 proliferation index 4%(+). With these histopathological and immunohistochemical findings, the diagnosis was a high-risk GIST (Miettinen-Lasota AFIP criteria). No metastasis was found in whole-body screening. The patient, who was in good general condition after the surgical procedure, was started on imatinib mesylate Gleevec® (Novartis Pharma Stein AG, Stein, Switzerland) treatment. The patient will be followed up at 3-month intervals with CT scans.

Case 2

A 69-year-old women, postmenopausal for 25 years, was admitted to our clinic with an adnexal mass that was randomly found during a routine examination. The personal and family history of this asymptomatic patient was unremarkable. Tumor markers, CBC, and blood chemistry were within normal limits, except for a CA-125 level of 65 U/mL. A hard, immobile mass measuring 15 cm in diameter was palpated in the right adnexal area at physical examination. A 20 cm, irregularly shaped, septated heteroechogenic mass with solid and cystic fields, appearing to arise in the right adnexa, was observed by transvaginal US and abdominal CT-scan (Figure 2 a,b). Laparatomy showed a solid tumor 13.5 cm in diameter arising from the small bowel loops, mainly ileal, adhering to surrounding tissues and to the left adnexal area (Figure 1b). The uterus and ovaries appeared to be atrophic. Ileoileal anastomosis and excision of the small intestine were performed. On histopathologic examination, the tumor showed perpendicularly oriented fascicles of spindle cells with bright eosinophilic cytoplasm and blunt-ended nuclei. The immunohistochemical profile of the tumor was desmin(+), smooth muscle actin(+), CD117(-), CD34(-), S-100(-), ki-67 proliferation index <1%(+). The diagnosis was leiomyoma of the small bowel. No additional postoperative treatment was given.

DISCUSSION

The presence of solid components, thick septa, papillary projections, and non-hyperechogenic (vascularized) tissue at USG examination of the adnexa indicate a high risk for patients (7). An evaluation considering both transvaginal USG appearance and CA-125 levels can effectively predict malignancy. Symptoms such as abdominal distension and abdominal or pelvic pain more frequent than 12 times monthly, or frequent urination and loss of appetite, call for a careful evaluation for the presence of ovarian cancer (8,9).

Ovarian cells produce and secrete CA-125. In addition, the peritoneum is a source of CA-125. CA-125 has been used as a tumor marker for ovarian cancer since the description of high concentrations of CA-125 in 80% of patients with epithelial ovarian cancer. However, this elevation is not specific and has also been observed in many physiological and pathological conditions: during menses, pelvic inflammatory disease, uterine myoma, endometrial cancer, gastro-intestinal tumors, and other cancers (10). Therefore, as in the second case, an elevated CA-125 level has been detected in patients with gastrointestinal tumors.

Malignant tumors are less frequently seen in the small intestine than in other sectors of the GIS (3). GIST is the third most

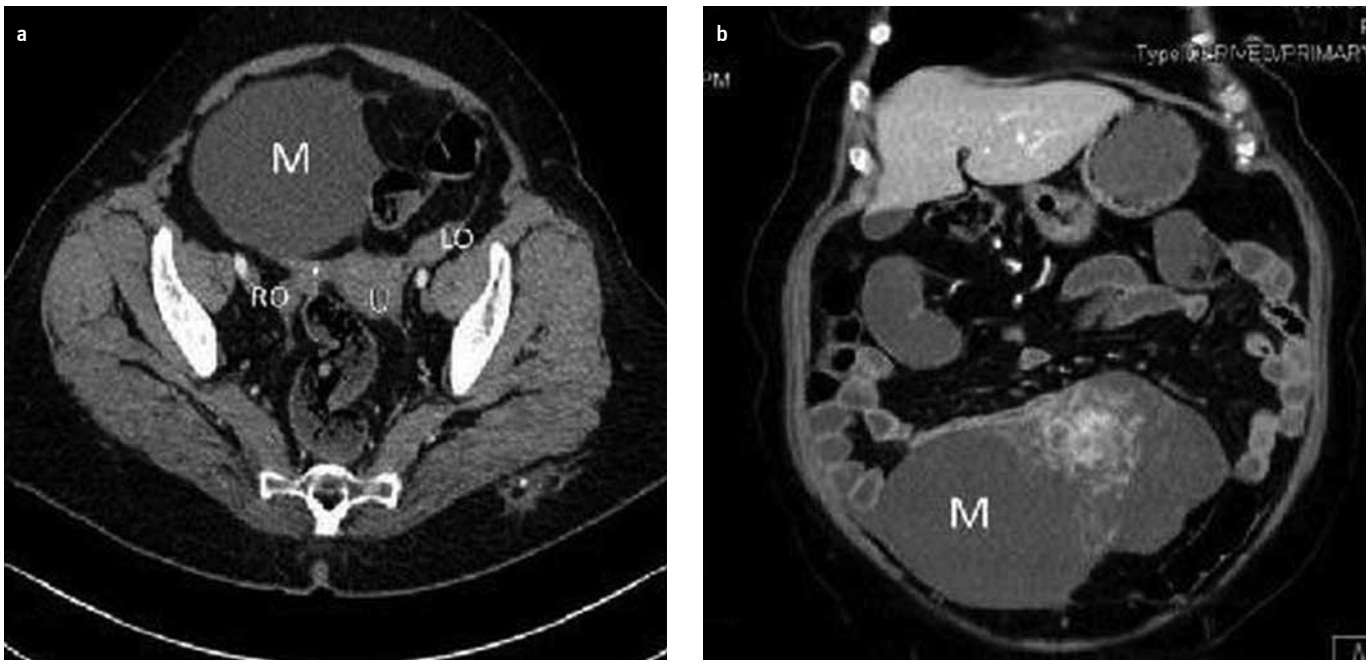

Figure 2. a, b. Transvers section CT. 10x15 cm thin-walled mass with cystic dansity in the right adnexa most likely originated from the right ovary **(a)**, and coronal reconstruction reaching up to the long axis 20 cm cystic mass with a solid components are shown **(b)**. M: Mass; U: Uterus; LO: Left ovary; RO: Right ovary.

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frequent tumor of the small intestine, following carcinoid and adenocarcinoma (11,12). It is relatively rare, making up only 1% of all GIS tumors. This tumor of mesenchymal origin arises in Cajal's cells (4). Its incidence is 1-2/100,000. Approximately 20-30% of cases are malignant in character. It is rarely seen before the age of 40 (5). GIST may present with massive gastrointestinal hemorrhage or an acute abdomen but may also be asymptomatic. The most frequent primary site of GIST is the stomach (60-70%), followed by the small bowel (20-30%), colon and rectum (5%), and esophagus (5%) (5). Recurrence is frequent, reaching approximately 50% in 5 years following complete resection of the primary tumor (13). Malignancy grade depends on tumor size and mitosis frequency. The potential malignancy is deemed to be low for tumors smaller than 5 cm with a mitotic count <5/50 HPF and high if the tumor is larger than 10 cm with a mitotic count >10/50 HPF (12,13). Currently, adjuvant postoperative treatment with tyrosine kinase inhibitors is recommended for high-risk patients. Imatinib mesylate, a selective tyrosine kinase inhibitor, targets the c-KIT proto-oncogene and platelet-derived growth factor-alpha receptor.

Leiomyomas are the most frequent benign tumors of the small intestine. They are asymptomatic in general. When symptoms are present, they most frequently consist of abdominal pain (54%), hemorrhage (45%), and acute intestinal obstruction with or without intussusception. Leiomyoma occurs equally frequently in both sexes and peaks in the fifth and sixth decades of life. Localization is significant for leiomyomas, the most frequent location being the jejunum (44%), followed by the ileum (37%) and duodenum (19%) (14).

Imaging investigations were performed in both reported cases with a focus on adnexal disease. The adherence of the mass to the uterus and its appendages at physical examination causes it to be interpreted as an adnexal mass or a uterine fibroid (especially a pedunculated one). Neither of these two cases was symptomatic; only the second had an elevated CA-125 level. The possibility of non-gynecological disease should be kept in mind in patients with normal tumor markers, as in our first case, or those with an unusual clinical assessment. In conclusion, both benign and malignant tumors of the small intestine are rare, diagnosed late, and even when advanced imaging methods and other diagnostic techniques are used in preoperative evaluation, are not always differentiated from adnexal masses.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author contributions: Concept - L.A., B.S.H.; Design - L.A., B.S.H., R.F., D.S.; Supervision - H.Y., M.C.T., M.S., L.A.; Resource - L.A., R.F., B.S.H., H.Y., D.S.; Materials - L.A., B.S.H., R.F., M.S., D.S.; Data Collection&/or Processing - L.A., B.S.H., R.F., M.S.; Analysis&/or Interpretation - L.A., B.S.H., R.F., M.S., M.C.T.; Literature Search - L.A., B.S.H., R.F.; Writing - L.A., B.S.H., M.C.T.; Critical Reviews - L.A., M.S., H.Y., M.C.T.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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