Gastrointestinal Stromal Tumor Presenting as a Mass on Pelvic Sonography

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- GI, gastrointestinal
- GIST, gastrointestinal stromal tumor

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal gastrointestinal (GI) tumors but are rare overall among other GI neoplasms. They are commonly asymptomatic and found incidentally during laparoscopy, surgical procedures, or radiologic studies. When symptomatic, they tend to present as GI bleeding, an abdominal mass, or abdominal pain. They may occur anywhere in the GI tract, most commonly in the stomach and small intestine. Histologically, they are described as tumors that are immunohistochemically positive for CD34 and CD117. It has been reported that GIST can present as a pelvic mass and has been found incidentally on sonography. We report a case of a GIST that was found on pelvic sonography and presented as severe pelvic pain during menstruation in a young woman.

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Case Report

A 36-year-old white woman, gravida 3, para 3, presented to the gynecologist’s office after lower back pain and severe pelvic pain during menstruation developed. Her medical history was notable for endometriosis and cervical adenocarcinoma treated with cervical conization 6 years previously. Abdominal examination was unremarkable, without masses or ascites noted. Vaginal, cervical, and uterine examination findings were grossly normal. Digital rectal examination revealed a 1-cm nodule on the right uterosacral ligament and a fullness higher in the pelvic cul-de-sac.
Pelvic sonography was performed with a Voluson 730 Expert system (GE Healthcare, Milwaukee, WI) with a 5- to 9-MHz broadband transvaginal probe. The solid adnexal mass was located left of the midline in the cul-de-sac. It appeared separate from the cervix and left ovary and measured 6.4 × 3.3 × 6.8 cm (Figure 1). After B-mode evaluation was done, the power Doppler mode was activated to evaluate vascularization of the mass. Pulsed Doppler ultrasound was used to interrogate identified blood vessels to obtain a flow velocity waveform. The pulsatility index and resistive index were calculated automatically (Figure 2). Next, color Doppler imaging was combined with a static 3D volume sweep. The volume data set was rendered with the glass body mode. Numerous blood vessels with multiple branching points were visualized (Figure 3). Abdominal and pelvic computed tomography with contrast showed a 7.7 × 4.3-cm mass within the cul-de-sac of unknown etiology extending left of the midline (Figure 4). There was no obvious adenopathy, metastasis, or ascites noted.

Figure 1.
Transvaginal sonogram showing a pelvic mass with dimensions of 6.4 × 3.3 × 6.8 cm.

Figure 2.
Transvaginal sonogram with Doppler imaging used to evaluate the vascularity of the mass.
Figure 3.

Transvaginal sonogram with Doppler imaging showing numerous blood vessels with multiple branching points in the pelvic mass.

Figure 4.

Abdominal and pelvic computed tomography with oral and intravenous contrast material showing the bladder, uterus, small bowel, and pelvic mass (A) with the mass extending left of the midline (B).

The patient was counseled on various treatment options and follow-up. We recommended exploratory laparoscopy with possible laparotomy. On laparoscopy, the palpable pelvic mass proved to be a solid, multilobulated neoplasm arising from the antimesenteric surface of the distal ileum, 2 ft proximal to the ileocecal junction. A small-bowel resection was performed, and a side-to-side functional end-to-end enterenterostomy was created. A 3.0 × 3.0-cm segment of the small bowel was sent for histologic evaluation.

Pathologic examination revealed a brown-tan hemorrhagic lobular mass arising from the serosal surface measuring 8.0 × 6.0 × 2.0 cm that was 1.5 cm from each of the margins. On microscopic examination, there was a spindle cell proliferation arranged in short bundles and interlacing arrays. There was patchy edema and hemorrhage without necrosis and fewer than 5 mitoses per 50 high-power fields. A battery of immunostains was applied, and the tumor cells were strongly immunoreactive with CD117 (Figures 5 and 6) and CD34 and negative for calponin, CD56, desmin, smooth muscle actin, and S-100 protein (not shown), an immunohistochemical profile strongly consistent with a diagnosis of a GIST. The patient
returned to the clinic after 6 weeks with appropriate wound healing. No chemotherapy or immunotherapy was deemed necessary at that time.

Figure 5.

Histopathologic sample stained with hematoxylineosin showing typical spindle cell morphologic features of a GIST.

Figure 6.

Histopathologic sample stained with the CD117 (c-Kit) immunostain with virtually all cells positive.

Discussion

Gastrointestinal stromal tumors constitute a subset of GI mesenchymal tumors of varying differentiation. Previously, these tumors were classified as GI leiomyomas, leiomyosarcomas, leiomyoblastomas, or schwannomas as a result of their histologic findings and apparent origin in the muscularis propria layer of the intestinal wall. With the advent of immunohistochemical staining techniques and ultrastructural evaluation, GISTs now are
recognized as a distinct group of mesenchymal tumors. In the present classification, GISTs account for approximately 80% of GI mesenchymal tumors.

Grossly, GISTs are well-demarcated spherical masses that appear to arise from the muscularis propria layer of the GI wall. Intramural in origin, they often project exophytically, intraluminally, or both, and they may have overlying mucosal ulceration. Larger GISTs nearly always outgrow their vascular supply, leading to extensive areas of necrosis and hemorrhage.

GISTs are rare tumors with an incidence of 5000 to 6000 cases in the United States, which constitutes less than 3% of all GI malignant neoplasias. Specifically, they represent only 20% of small-bowel malignant neoplasms, 1% to 2% of gastric malignancies, and less than 1% of malignancies involving the esophagus, colon, and rectum. About 50% to 70% of GISTs occur in the stomach, 33% in the small bowel, 5% to 15% in the recto-colon, and only 1% to 5% in the esophagus. They are multicentric in less than 5% of cases. These tumors have a wide clinical spectrum at presentation. They range from incidentally detected, asymptomatic, benign GISTs to large malignant tumors, which frequently cause the patient to seek medical attention. Previous articles have described the evaluation of malignancy, diagnosis, and prognostic factors of GIST. The National Institutes of Health convened a GIST workshop in April 2001, and a consensus approach for the diagnosis of GIST was reached. The most important prognostic factors include tumor size, mitotic rate, and site. Gastric tumors were found to be less aggressive than intestinal tumors. Small-intestinal tumors were found to have a worse prognosis and esophageal tumors the best, but those conclusions were not reliable.

It was found that small intestinal GISTs of greater than 5 cm, with more than 5 mitoses per 50 high-power fields, or both have high risk for metastasis. This tumor was considered high risk for aggressive behavior mainly because of its size of 8.0 cm and therefore was surgically removed.

The mainstay in treatment of a GIST is surgical resection. However, imatinib mesylate (Gleevec; Novartis Pharmaceuticals Corporation, East Hanover, NJ) has been found to be beneficial for both induction and adjuvant therapy in advanced cases. The adnexal mass described in this case measured greater than 5 cm in diameter and contained numerous branching blood vessels with elevated diastolic flow (low resistive index). These findings are associated with malignancy in adnexal masses diagnosed by pelvic sonography. These morphologic characteristics and vascular patterns prompted definitive surgical therapy with good results.

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Footnotes

- Received July 13, 2006, from the Indiana University School of Medicine, Indianapolis, Indiana USA (M.K.); Department of Obstetrics and Gynecology (R.K.) and Division of Maternal and Fetal Medicine, Department of Obstetrics and Gynecology (A.K.H.), St Vincent Women’s Hospital, Indianapolis, Indiana USA; and AmeriPath Indiana, Indianapolis, Indiana USA (W.J.). Revision requested August 14, 2006. Revised manuscript accepted for publication August 15, 2006.

- Received July 13, 2006.
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