Primary Extragastrointestinal Stromal Tumor of the Sigmoid Mesocolon with Metastatic Spread to Greater Omentum: Case Report

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ABSTRACT

A 71-year-old female complained of abdominal pain, weight loss and abdominal distension. Gynecologic examination revealed a hardly movable, palpable mass in the lower abdomen, reaching the umbilicus. An abdominal ultrasound and computed tomography (CT) scan suggested a large abdominal mass with the possible origin in the left ovary and without significant lymph node enlargements. The patient subsequently underwent complete evacuation of tumor tissue, omentectomy and total abdominal hysterectomy and bilateral salpingo-ovariectomy. Immunohistochemical examination revealed strongly positive staining of tumor cells for CD117. The final pathologic diagnosis was a primary extragastrointestinal stromal tumor (EGIST) of the sigmoid mesocolon with omental metastasis. The differential diagnosis of the tumor presented in the lower abdomen should consider the EGIST as well.

Key words: immunohistochemistry, CD117, metastasis

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumor of the gastrointestinal (GI) tract. GISTs express c-kit protein also known as CD117, which is considered as a highly specific marker that differentiates it from other mesenchymal tumors¹. Neoplasms with histology and immunohistochemistry similar to GISTs may occur outside the GI tract, for example in the soft tissue of the abdominal cavity (omentum and mesentery) or in the retroperitoneum². GIST that arises primarily outside the GI tract is termed extragastrointestinal stromal tumor (EGIST). While the histogenesis, prognostic parameters and outcomes of GISTs are widely known, pathogenesis, incidence and prognosis of EGISTs have not yet been completely defined. We report the results of the macroscopic and microscopic examinations, including immunohistochemical studies, of an EGIST of the sigmoid mesocolon.

Case Report

A 71-year-old woman presented with abdominal pain and pressure of 3 months duration, accompanied by anemia and a weight loss of 4 kilograms. There was no family history of malignant diseases. Physical examination revealed a hardly movable, palpable mass in the lower abdomen, reaching the umbilicus. The abdominal ultrasound discovered a solid, highly vascularized mass (25x15 cm) with numerous shunts, located in the pelvic cavity (behind the uterus) and abdomen. The resistive index
Discussion

The GISTs are the most common mesenchymal tumors of the GI tract with an incidence estimated at 7 to 14 per 1 million in the general population. These tumors usually occur in the stomach (60–70%) and small intestine (20–35%), with rare occurrence in the colon and rectum (5%), esophagus (<2%) and appendix. Some GISTs are unrelated to the tubular gastrointestinal tract and they are termed EGISTs. EGISTs are usually of the gastrointestinal stromal tumor (GIST) type, which is defined by virtue of any degree of association with the tunica muscularis propria or the muscularis propria. EGISTs are more common in the alimentary tract, with a predilection for the stomach (50%) and small intestine (20–35%).

EGISTs have been reported in four published series from 1999 to 2008. The EGISTs are the most common mesenchymal tumors of the GI tract with an incidence estimated at 7 to 14 per 1 million in the general population. These tumors usually occur in the stomach (60–70%) and small intestine (20–35%), with rare occurrence in the colon and rectum (5%), esophagus (<2%) and appendix. Some GISTs are unrelated to the tubular gastrointestinal tract and they are termed EGISTs. Incidence of EGISTs is not defined yet. EGISTs are probably rarer than previously reported.

Most cases of EGISTs are likely to represent more extensive extramural growth with eventual loss of contact with the muscle layer of the gut. EGISTs should be defined by virtue of any degree of association with the muscularis propria (supported by desmin immunoreactivity), but not by localization of the bulk of the tumor.

EGISTs arising in the mesentery of sigmoid colon are extremely rare. A total of 99 omental and mesenteric EGISTs have been reported in four published series from 1999 to 2008. EGISTs have various clinical behavior, and the parameters used for predicting the prognosis of GIST may not be completely suitable for EGIST evaluation. Miettinen et al. examined seven cases of mesenteric EGISTs and nine cases of omental EGISTs. Mesenteric EGISTs appeared more aggressive (higher mitotic activity, frequent malignant behavior).

Reith et al. noted that EGIST arising within the abdominal cavity (40 cases) and the retroperitoneum (8 cases) expressed CD117 (100%), CD34 (50%), neuron-specific enolase (44%), smooth muscle actin (26%), desmin (4%), and S-100 protein (4%). High cellularity, mitotic activity (>2 mitoses/50 HPF) and the presence of necrosis were factors indicative of a potentially aggressive clinical course for EGIST. Our patient displayed two high-risk features (mitotic activity >2/50 HPF and presence of necrosis). Tumor size, which is commonly used in GISTs as a prognostic factor, is not applicable to EGISTs. EGISTs appear to have enough space to grow and they are often large size, presenting clinical symptoms only after a long time. Since the preoperative diagnosis based on clinical and radiological data is difficult, the patients

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Fig. 1. Surgically removed tumor mass.
usually undergo a surgical operation for the generic diagnosis of "abdominal mass" or suspected gynecological tumor\(^\text{1,11}\). Our patient had nonspecific imaging features on abdominal ultrasound and CT scan that could mimic the ovarian tumor.

An aggressive surgical approach is the most effective treatment\(^\text{12}\). Lymphadenectomy is not required\(^\text{13}\).

**Conclusion**

This case draws our attention to the importance of considering EGISTs in the differential diagnosis of a tumor mass in the lower abdomen. Especially surgeons as well as diagnostic pathologists should be aware of this possibility. In most cases a preoperative diagnosis is not possible.

**REFERENCES**


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**PRIMARNI EKSTRA GASTROINTESTINALNI STROMALNI TUMOR MEZENTERIJA SIGMOIDNOG DEBELOG CRVJA S METASTAZAMA U VELIKOM OMENTUMU: PRIKAZ SLUČAJA**

**SAŽETAK**

Žena u dobi od 71 godinu žalila se na bol u trbuhu, nadušnost i gubitak tjelesne težine. Ginekološkim pregledom palpirala se slabo pomična tumorska masa u donjem trbuhu koja je sezala do pupka. Abdominalnim ultrazvukom i kompjutoriziranim tomografijom prikazana je velika tumorska tvorba u trbuhu koja vjerojatno potječe od lijevog jajnika. Povećani limfni čvorovi nisu prikazani. Kirurškim je zahvatom u potpunosti odstranjeno tumorsko tkivo, učinjena je omentektomija i totalna abdominalna histerektomija s obostranom adneksektomijom. Imunohistokemijskim bojanjem dokazana je jaka pozitivnost tumorskih stanica na CD117. Konačna dijagnoza je bila primarni ekstragastrointestinalni stromalni tumor (EGIST) mezenterija sigmoidnog debelog crijeva s metastazama u velikom omentumu. Diferencijalno dijagnostički kod tumora donjeg abdomena uvijek treba uzeti u obzir EGIST.