SEZARYEN ESNASINDA RASTLANTISAL OLARAK SAPTANAN GASTROİNTESTİNAL STROMAL TÜMÖR: OLGU SUNUMU VE LİTERATÜR TARAMASI

Incidental Diagnosis of a Gastrointestinal Stromal Tumor during Cesarean Section: A case Report and rReview of the Literature

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ÖZET

Gastrointestinal stromal tümörler (GIST'ler) gastrointestinal sistemin interstisyel Cajal hücrelerinden gelişen en yaygın mezenkimal tümörlerdir. GIST'lerin görüldüğü yaş dağılımı göz önüne alındığında, gebelik veya puerperium sırasında GIST tanısı çok nadirdir. 26 yaşında primigravid bir kadın acil servisimize doğum sancısı ile başvurdu. Sezaryen operasyonu sırasında, ince bağırsağın antimezenterik duvarından kaynaklanan 7 cm'lik bir kitle saptandı. Hastaya genel cerrahi ekibi tarafından sezaryen sonrası 38. günde ince barsak rezeksiyonu ve parsiyel omentektomi operasyonu uygulandı. Patolojik incelemede 9x6x6 cm infiltratif gastrointestinal stromal tümör saptandı. Medikal onkolojinin önerisiyle Imatinib tedavisi başladı. Hasta halen tıbbi onkoloji kliniği tarafından takip edilmektedir. Sonuç olarak, sezaryen sırasında batın içinin eksplorasyonu asemptomatik kitlelerin tespiti açısından önemlidir.

Anahtar Sözcükler: *Gastrointestinal Stromal Tümör; Gebelik; Sezaryen*

ABSTRACT

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors that develop from the interstitial Cajal cells of the gastrointestinal tract. Given the age distribution of ocurrence, a diagnosis of GISTs during pregnancy or puerperium is very uncommon. A 26-year-old primigravid woman was admitted to our emergency department with labor pain. During cesarean a 7 cm mass was found incidentally, which originated from the antimesenteric wall of the small intestine. The patient underwent a small bowel resection and partial omentectomy on the 38th day after cesarean section by the general surgical team. Pathology revealed a 9x6x6 cm infiltrative gastrointestinal stromal tumor. Imatinib therapy was started with medical oncology's recommendation. The patient is still being followed by the medical oncology clinic. In conclusion, exploration of the abdomen during a cesarean section is important for the detection of asymptomatic masses.

Keywords: *Cesarean Section; GISTs; Pregnancy*

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INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors that develop from the interstitial Cajal cells of the gastrointestinal tract, and are usually present during the fifth to seventh decade of life (1,2). Given the age distribution of ocurrence, a diagnosis of GISTs during pregnancy or puerperium is very uncommon (3). These tumors may be asymptomatic and are discovered incidentally during a radiologic scan for another purpose. Moreover, non-specific symptoms of pregnancy may be confused with symptoms from the tumor, resulting in the difficult diagnosis of tumors in pregnancy (4). GISTs during pregnancy and puerperium that were reported in the literature are listed in Table 1 (5-19). Here, we describe our experience in the unique case of a GIST detected incidentally during the cesarean section of a 26-year-old pregnant patient, which was asymptomatic during the pregnancy.

CASE PRESENTATION

A 26-year-old primigravid woman was admitted to our emergency department with labor pain. Four years ago, she had a myomectomy operation. Ultrasonographic examination showed a viable, intrauterine 39-week pregnancy. Upon vaginal examination, her cervical dilation was 3-4 cm and cervical effacement was at 60%. As she was in active labor and had a history of myomectomy, the patient was scheduled for a cesarean section; the patient gave birth to a live 2800gr female baby. During the cesarean section, neither acid nor blood were detected in abdomen. Bilateral ovaries and tubes were natural in appearance. Bleeding control was performed following the suturation of the lower segment's transverse incision. Then, a 7 cm mass was found incidentally, which originated from the antimesenteric wall of the small intestine (Figure 1). The general surgery team was consulted during surgery. It was reported that the lesion was suspected to be malignant; segmental intestinal resection was necessary, and that the operation would be planned after the necessary approvals and preoperative examinations. During cesarean section mass resection was not performed because there was no informed consent form including the possibility of small bowel resection and ileostomy.

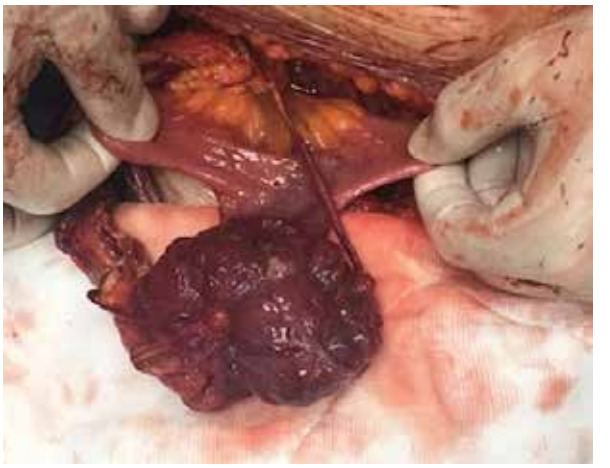

Figure 1. The wiev of the GIST in small bowel which was found incidentally during cesarean section

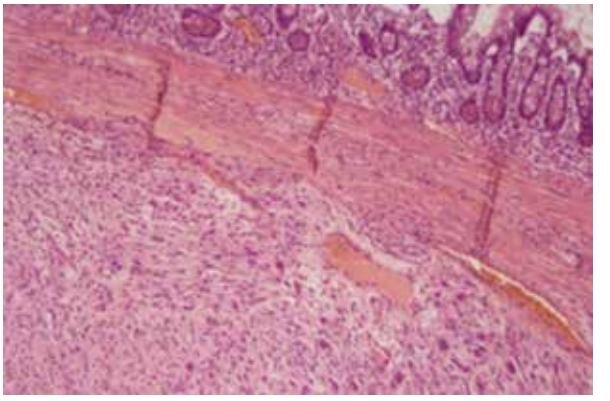

Figure 2. The tumor was composed of spindle and epithelioid cells (Hematoxylin and eosin staining x10)

The cesarean section of the patient was completed and uneventfully discharged on the third postoperative day. The patient received a follow-up from the general surgeons. The patient was informed about the planned surgery and was expected to decide on the operation. After the patient decided on the operation, preoperative upper gastrointestinal tract endoscopy, colonoscopy and radiologic scanning of the abdomen and thorax with computed tomography (CT) were performed.

Table 1. GISTs during pregnancy and puerperiumthat were reported in the literature

Cases	Age	Presentation	Site	Size	Surgery	Pathology
Benjamin Michael Stubbs et al. (5)	31	Lethargy, sensation of abdominal mass	transverse colon	17x14x12	Intestinal resection	CD117, CD34, a smooth muscle actin (+) desmin (-)
Michael varras et al. (6)	28	Acute abdomen	small intestine	13x10x9	Intestinal resection	CD117, CD34, a smooth muscleactin (+) desmin, S-100, cytokeratin (-)
Cuerva-González MJ et al. (7)	39	Gıs bleeding, ex fetus	small intestine	15x12	Intestinal resection	CD117, c-kit (+)
S. Scherjon (8)	25	Sensation of abdominal mass	small intestine	20	Intestinal resection + splenectomy	CD117, SM Actin (+) CD34, S-100 (-)
Covoney S. (9)	42	Incidentally	extradigestive stromal tumor(retroperitoneum)	11	Resection	N. A
Emma T. Igras et al. (10)	42	Incidentally	duodenum	12	Intestinal resection	CD117, DOG1 (+) CD34, PDGFR, desmin, S100 (-)
N. Haloob et al. (11)	31	Lethargy, dizziness	small intestine, colon	10,6x6,8x10,9	Intestinal resection	CD117, CD34 (+)
PHILIP T. V. et al. (12)	32	Abdominal pain	stomach	13x9	Intestinal resection	CD34, vimentin (+), smooth muscle actin, e2, progesterone, cytokratin, desmin S-100 (-)
S. Lanzafame et al (13)	29	Abdominal pain	stomach	4x3x2	Wedge resection	CD 34, CD 117, EGFR, PgR (+) desmin, a smooth muscle actin, S-100, GFAP, HER-2, estrogen receptor (-)
U. Parampalli et al (14)	28	Upper gis bleeding	stomach	5,7	Partial resection	CD117, CD34 (+) smooth muscle actin, S-100, desmin (-)
Jove Oblitas WDC et al. (15)	28	Upper gis bleeding	stomach	8	Intestinal resection	CD117, Ki 67 (+) CD34 (-)
Iliass Charif et al. (16)	42	Abdominal pain	stomach	16x21x23	Intestinal resection	CD 117, CD 34, PS100, AML (+)
Ilay Gozukara et al. (17)	21	Abdominal pain	extradigestive stromal tumor	17x17x10	Resection	c-kit, CD34, Desmin(+)
S. Mahdaoui et al. (18)	38	Sensation of abdominal mass	extradigestive stromal tumor (omentum)	17x12x12	Did not performed	N. A
Neerja Goel et al. (19)	25	N.A (primary tumor)	stomach, colon (primary tumor)	15x16x8	Intestinal resection (primary tumor	Smooth muscle, CD117(+) primary tumor
Present case	26	Incidentally	small intestine	75x55	Intestinal resection + partial omentectomy	CD117, DOG1 (+), CD34, SMA, S-100 (-)

The patient's abdominal CT revealed a 75 x 55 mm mass with lobulated contour, heterogeneous contrast enhancement after administration of intravenous contrast agent, and necrotic components with no enhancement in the left lower quadrant, adjacent to the ileal anses. Surgery was performed after obtaining the informed consent form, including the possibility of ileostomy. The patient underwent a small bowel resection and partial omentectomy on the 38th day after cesarean section by the general surgical team. There was no metastasis detected in the preoperative radiologic scanning and intraoperative exploration of the abdomen.

Intraoperative pathologic result of the frozen section is 'infiltrative gastrointestinal stromal tumor and any tumor was detected at the surgical margin'. Final pathology revealed a 9x6x6 cm infiltrative GIST and frozen section result was confirmed. The mitotic rate was less than 5 mitoses per 50 high power fields (HPFs). The tumor's cell type was mixed epithelioid and spindle cells (Figure 2). Cytological atypia was significant. Immunohistochemical staining of the tumor tissue demonstrated strongly positive reactivity to CD 117 and DOG-1, but negative reactivity to CD 34, SMA, and S-100. Imatinib therapy was started with medical oncology's recommendation. The patient is still being followed by the medical oncology clinic. When the patient started imatinib therapy, she was recommended not to breastfeed her baby. A one-year follow-up the patient is alive and recent radiologic imaging shows any local recurrence or secondary metastases.

DISCUSSION

GISTs are the most common mesencyhmal tumors of the gastrointestinal tract. Although the tumor's localization is commonly in the stomach (50-60%) and small intestine (20-30%), it can be detected anywhere in the gastrointestinal tract such as the large bowel or esophagus. Rarely, stromal tumors can occur outside of the GI tract such as the mesentery, omentum, and retroperitoneum (20, 21).

Patients with GISTs usually have non-specific symptoms (i.e., early satiety, bloating) and these symptoms may also occur in pregnancy. Because of this, the diagnosis of GISTs during pregnancy is difficult. Also, GISTs may cause intra-abdominal hemorrhage due to tumor ulceration, upper and lower gastrointestinal bleeding, abdominal mass sensation due to the size of the mass, and gastrointestinal obstruction (4). Intra-abdominal hemorrhage, gastrointestinal bleeding, and intestinal obstruction in pregnancy increase the risk of morbidity and mortality for both the fetus and pregnant patient. Cuerva Gonzales MJ., et al. reported the case of a patient who was diagnosed with a gastrointestinal stromal tumor during her pregnancy due to massive lower gastrointestinal bleeding, and the fetus died (7). In some cases, GISTs are asymptomatic and are detected incidentally during radiological imaging or an abdominal operation for another purpose. In their cases, Covoney S. and Emma T. Igras et al. diagnosed GISTs in pregnancy incidentally during the routine ultrasonographic examination of a fetus (9, 10). Our case is a unique example of a GIST that was detected incidentally during a cesarean section. The tumor originated from the small intestine and was approximately 50 cm proximal to the cecum. The patient's diagnosis, who had no symptoms during pregnancy, was incidentally detected during her cesarean section.

The prognosis of GISTs is highly associated with their histological type, size, mitotic index, localization, and metastasis. Poor prognostic factors include mixed type histology, small intestinal localization, a tumor size of >5 cm, and >5/50 HPF mitotic index. In our case, mixed type cell morphology, small intestinal bowel localization, and a tumor size of 7 cm were poor prognostic indicators (22, 23).

The primary treatment of GISTs is surgery. Resection of the tumor with a clear surgical margin is the preferred surgical procedure (24). Imatinib mesylate, which selectively inhibits BCR-ABL, KIT, and PDGFR tyrosine kinases, is the standard adjuvant treatment for patients with metastatic disease and poor prognostic factors (25, 26). Moreover, breast feeding is not recommended for patients under imatinib treatment and for one month after the last imatinib dose, due to its potential for serious adverse reactions in the infant (27). In this case, because of the poor prognostic factors, imatinib

treatment was started by medical oncologists after the surgical resection of the tumor, and breast feeding was stopped during imatinib treatment.

CONCLUSION

This is the first reported GIST case in the literature that was incidentally detected during a cesarean section. Exploration of the abdomen during a cesarean section is important for the detection of asymptomatic masses. Additionally, our case demonstrates the management of an incidental GIST that was detected during a cesarean section with a multidisciplinary team including obstetricians, gynecologists, general surgeons, pathologists, and medical oncologists.

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