AN UNUSUAL PRESENTATION OF HUGE GASTRIC CYSTIC GASTROINTESTINAL STROMAL TUMOR

Büyük Mide Kistik Gastrointestinal Stromal Tümörünün Olağandışı Bir Sunumu

Nihan TURHAN[1], Didem ERTORUL[1], Elbrus ZARBALIYEV[2], Şafak COŞKUN[3], Dilek YILMAZ[4]

[1]Şehit Prof. Dr. İlhan Varank Sancaktepe Training and Research Hospital, Department Of General Surgery İstanbul, Türkiye
[2]Department of General Surgery, Gaziosmanpaşa Hospital, Yeni Yüzyil University, İstanbul, Türkiye
[3]Marmara University Hospital, Department Of General Surgery, İstanbul, Türkiye
[4]Şehit Prof. Dr. İlhan Varank Sancaktepe Training and Research Hospital, Department Of Pathology, İstanbul, Türkiye

Nihan TURHAN, Uzm. Dr. (0000-0002-8672-3853)
Didem ERTORUL, Op. Dr. (0000-0003-4595-5938)
Elbrus ZARBALIYEV, Op. Dr. (0000-0002-4595-8314)
Şafak COŞKUN, Uzm. Dr. (0000-0001-8443-1497)
Dilek YILMAZ, Dr. (0000-0003-4193-4558)

İletişim:
Dr. Elbrus ZARBALIYEV
Department of General Surgery, Gaziosmanpaşa Hospital, Yeni Yüzyil University, İstanbul, Türkiye

Geliş tarihi/Received: 09.08.2022
Kabul tarihi/Accepted: 30.07.2023
DOI: 10.16919/bozoktip.1159004

Bozok Tıp Derg 2023;13(3):90-93
Bozok Med J 2023;13(3):90-93

ABSTRACT

Gastrointestinal stromal tumors (GISTs) are nonepithelial, mesenchymal tumors arising from the interstitial cells of Cajal. The diagnostic tool for the GISTs is tyrosine kinase growth factor receptor (c-KIT / CD117) expression. They most commonly occur in the stomach. Here we reported a rare case of GIST, presenting as a large cystic mass connected to the main tumor with a peduncle. A 44-year-old female patient presented to the emergency department with abdominal pain and intestinal obstruction. Emergency laparotomy revealed a cystic mass of approximately 25 cm in diameter. The pathological diagnosis was gastrointestinal stromal tumor with cystic degeneration. Tumor contained areas of necrosis, high mitotic index, c-KIT positive and was large in diameter. Imatinib mesylate treatment was given. Cystic GISTs have poor prognostic factors and patients should be followed closely in the postoperative period.

Keywords: *Gastrointestinal Stromal Tumor, Intraabdominal Mass, Gastric Tumors*

ÖZET

Gastrointestinal stromal tümörler (GİST'ler), interstisyel Kajal hücrelerinden kaynaklanan epitelyal olmayan, mezenkimal tümörlerdir. GİST'ler için tanı aracı tirozin kinaz büyüme faktörü reseptörü (c-KIT/CD117) ekspresyonudur. En sık midede görülürler. Bu çalışmada tümöre pedikül ile bağlı büyük bir kistik kitle olarak ortaya çıkan nadir bir GİST olgusunu sunduk. 44 yaşında kadın hasta karın ağrısı ve barsak tıkanıklığı şikayetleri ile acil servise başvurdu. Acil laparotomide yaklaşık 25 cm çapında kistik kitle saptandı. Patolojik tanı kistik dejenerasyonlu GİST idi. Tümör nekroz alanları içeriyordu. Yüksek mitotik indeks, c-KIT pozitif ve çap olarak büyüktü. Imatinib Mesilat tedavisi verildi. Kistik GİST'ler kötü prognostik faktörlere sahiptir ve postoperatif dönemde hastalar yakın takip edilmelidir.

Anahtar Kelimeler: *Gastrointestinal Stromal Tümör, İntrabdominal Kitle, Mide Tümörleri*

INTRODUCTION

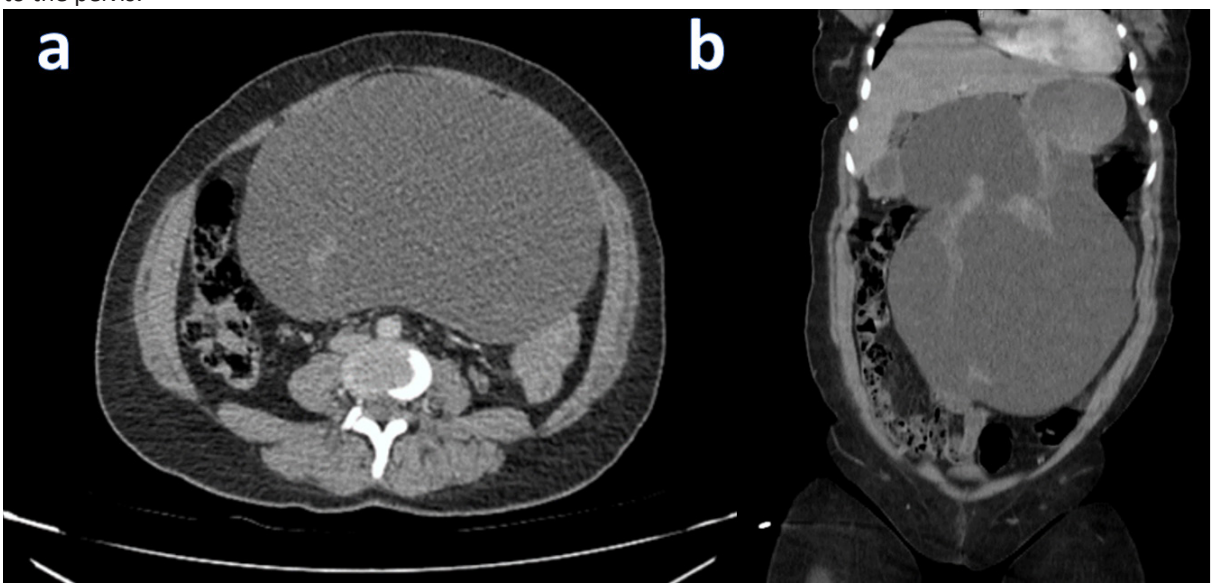
Gastrointestinal stromal tumors (GISTs) are nonepithelial, mesenchymal tumors arising from the interstitial cells of Cajal. These cells provide connection between myenteric and muscular layers in the gastrointestinal tract. Most of GISTs arise from muscularis propria and show extraluminal growth (1). GISTs are most commonly occur in the stomach (60-70%) (2). While stomach is the most common site for GISTs, pedunculated cystic GIST of stomach with extragastric growth presentation is very rare (3-5).We report a female patient with a giant gastric cystic GIST.

CASE REPORT

A 44-year-old female patient presented to the emergency department with complaints of abdominal pain, swelling, nausea, and multiple bilious vomiting for the past few days. The patient was evaluated in the emergency room with the signs of intestinal obstruction. She was dehydrated, conscious, cooperative and oriented. Vital signs were blood pressure: 110/70 mmHg, pulse: 98/min, fever: 36.8 °C, respiratory rate: 20/min. On physical examination of the abdomen, there were distention, tenderness and defense. There were fullness filling the entire abdomen on palpation and dullness on percussion. There was intestinal content coming from the nasogastric tube inserted in the patient. In the rectal examination, there were no palpable mass and blood.

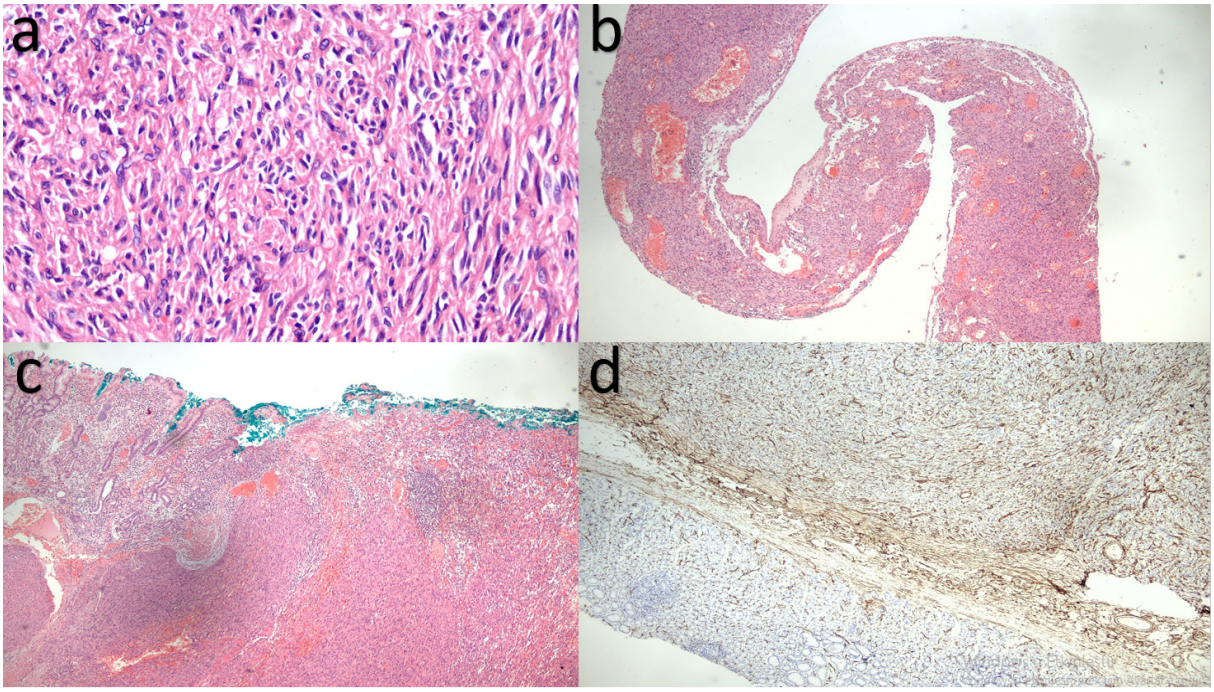
Hb was 9.7 g/dL (11.00-15.00), CRP: 93.9 mg/l (0-5). Leukocyte, thrombocyte, liver function tests, blood electrolytes, blood urea nitrogen and creatinine values were within normal limits. Abdominal computed tomography (CT) showed a septated cystic mass in the midline of the abdomen with a diameter of 25x16 cm, thought to be of mesenteric origin (Figure 1a, b). The patient was operated on urgently with those findings. The abdomen was entered by laparotomy with a midline incision. During the exploration, it was observed that there was a cystic mass of approximately 25 cm in diameter, located in the midline, extending from the stomach to the pelvis. This mass was associated with a mass of approximately 2 cm in diameter arising from the posterior corpus wall at the greater curvature of the stomach. Although the cystic mass had some adhesions with the surrounding omentum, there was no invasion to other surrounding organs. When the exploration was continued, it was observed that there was another mass of 2-3 cm in size, which was thought to originate from the stomach wall in the pre-pyloric part of the stomach. The cystic mass was removed en bloc by distal gastrectomy, including two palpable solid masses. Histopathological sections showed two tumoral masses, one of which showed cystic degeneration in the gastric wall. No infiltration into the surrounding tissue was observed. The cystic tumor was connected with a pedicle consisted of tumor cells. The resected tumors were 24.5x19x8 cm and 3x2x1.5 cm in size. The tumor cells had eosinophilic cytoplasm and round nuclei with spindle and epithelioid cell morphology. There was significant superficial ulceration in submucosa and mucosa layers of cystic mass. Mitosis of 20 / 50 HPF was noted (Figure 2 a, b, c). Immunohistochemistry showed CD 117, CD 34, DOG 1, SMA positive, Desmin & Myogenin negative (Figure 2d). Ki 67 proliferation index was 1%. Lateral, proximal and distal clean surgical margins were more than 1 cm. The diagnosis was gastrointestinal stromal tumor with cystic degeneration. The patient, who had no problems in the postoperative follow-up, was discharged on the 7th postoperative day with good recovery. No tumor was detected in any other area in the upper and lower gastrointestinal system endoscopy and positron emission tomography (PET-CT) performed in the postoperative period. As the patient's tumor was c-KIT positive, big in size and high mitotic index, the patient was started on Imatinib treatment.

Figure 1.
a- CT scan of 25x16 cm diameter cystic mass in the midline of abdomen
b- CT scan of 25x16 cm diameter septated cystic mass in the midline of abdomen extending from the stomach to the pelvis.



Bozok Tıp Derg 2023;13(3):90-93
Bozok Med J 2023;13(3):90-93

Figure 2.
a- GIST with spindle and epithelioid cell morphology H&E x200,
b- GIST with wall appearance in areas of cystic degeneration H&E x40,
c- GIST showing superficial ulceration in submucosa and mucosa H&E x40,
d- GIST cells stained positive with



The patient is followed in the 10th postoperative month without any additional problems. A written statement was obtained from the patient for this report.

DISCUSSION

GISTs are cellular spindle cell, epithelioid or pleomorphic mesenchymal tumors of gastrointestinal tract, first defined in 1983. Before that time, they were classified as smooth muscle tumors like leiomyoma, cellular leiomyoma, leiomyoblastoma and leiomyosarcoma (6). GISTs arises from Cajal cells of the gastrointestinal system. The diagnostic tool for the GISTs is tyrosine kinase growth factor receptor (c-KIT / CD117) expression. Only a small subgroup of GISTs which lack KIT expression are presented in literature. Other diagnostic criteria include positivity for CD34 (70%), smooth muscle actin expression (20-30%), S100 protein expression (10%) and negativity for Desmin (only 2-4% are positive). DOG1 positivity is considered clinically significant in diagnosing GIST (1, 5, 7). GISTs incidence is 1.5/1.000.000/ year. GISTs are more common in elderly ages and in males (8). GISTs are usually small masses located in the submucosal area and are diagnosed incidentally during examinations or surgeries performed for reasons other than symptomatic presentations. The most common complaints in symptomatic GISTs are compression symptoms due to mass compression effect. Although the need for emergency surgery is rare (<15%) for GISTs, intestinal obstruction, gastrointestinal system bleeding, tumor rupture, intussusception are surgical emergencies (1, 9). Intestinal obstruction is in the form of extraluminal compression or intussusception due to the extraluminal exophytic growth of the submucosal tumor (9). The Armed Forces Institute of Pathology (AFIP) classification system. discovered that in addition to tumor size and mitotic rate, anatomical location and the total area for mitotic counting (5 mm^2) is important for prognosis (10). High mitotic activity (mitosis of > 5 / 50 HPF), big tumor diameter (greater than 5 cm),

and c-KIT (CD 117) expression are associated with malignant behavior of GISTs. Apart from these, positive surgical margins, tumor rupture, tumor necrosis, peripheral organ invasion and distant metastasis are considered among the poor prognostic factors (10).

It has been reported that esophageal and gastric GISTs have a better prognosis than colorectal and small bowel GISTs. Gastric GISTs presenting as a pedunculated cystic mass, on the other hand, have a poor prognosis due to their large size, high number of mitoses, and areas of tumor necrosis (4, 5, 11). Although stomach (60-70%) is the most common site for GISTs, they can occur from esophagus to anus in every organs in the gastrointestinal tract (1, 7). In the literature, there are few case reports of GIST presenting as a large cystic mass connected to the main tumor with a peduncle (5). It has been suggested that aggressive tumor growth, congestion, intratumoral hemorrhage and necrosis are effective in the formation of large cystic spaces in the tumor and formation of cystic GIST (11, 12).

The most effective treatment for GISTs is surgery. These tumors generally do not invade the surrounding tissue. The tumor should be removed en bloc. For pedunculated GISTs complete surgical resection (R0) should be provided by wedge resection of the organ which is in continuity with the mass. The recommended approach is R0 resection with wide margins of about 1–2 cm (1, 4, 13).

The efficacy of chemotherapy and radiotherapy in the treatment of GISTs is controversial. The recognition of c-KIT positive patients has an impact on the treatment protocol. Imatinib mesylate is approved for use in GISTs, as it inhibits the tyrosine kinase activity of the c-KIT receptor (1). The cystic GIST of our patient contained areas of necrosis, high mitotic index, c-KIT positive and was large in diameter. Imatinib mesylate treatment was started because of the poor prognostic factors.

Cystic GISTs should be kept in mind in differential diagnosis of intra-abdominal cystic tumors. The frequency of endoscopic and radiological follow-up of GIST patients after treatment should be done individually, taking into account prognostic parameters. Poor prognosis of gastric cystic GISTs should not be forgotten, and patients should be followed closely in the postoperative period.

REFERENCES

1. Miettinen M, Lasota J. Gastrointestinal stromal tumors--definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. Virchows Arch. 2001;438(1):1-12.

2. Miettinen M, Sarlomo-Rikala M, Lasota J. Gastrointestinal stromal tumors: recent advances in understanding of their biology. Hum Pathol. 1999;30(10):1213-20.

3. Naitoh I, Okayama Y, Hirai M, Kitajima Y, Hayashi K, Okamoto T, et al. Exophytic pedunculated gastrointestinal stromal tumor with remarkable cystic change. J Gastroenterol. 2003;38(12):1181-4.

4. Cavallaro G, Sadighi A, Polistena A, Rossi V, Cristaldi M, Paparelli C, et al. Pedunculated giant GISTs of the stomach with exophytic growth: report of two cases. Int J Surg. 2008;6(6):e80-2.

5. Wang L, Liu L, Liu Z, Tian Y, Lin Z. Giant gastrointestinal stromal tumor with predominantly cystic changes: a case report and literature review. World J Surg Oncol. 2017;15(1):220.

6. Sripathi S, Rajagopal K, Srivastava RK, Ayachit A. CT features, mimics and atypical presentations of gastrointestinal stromal tumor (GIST). Indian J Radiol Imaging. 2011;21(3):176-81.

7. Bülbül Doğusoy G. Gastrointestinal stromal tumors: A multicenter study of 1160 Turkish cases. Turk J Gastroenterol. 2012, 23(3):203-11.

8. Mehta D. Commentary to: Atypical Presentation of Gastrointestinal Stromal Tumours-A Case Report by Kalpana Raja et al. Indian J Surg. 2014;76(5):425.

9. Akyüz C, Torun M. Mide Gastrointestinal Stromal Tümörünün Neden Olduğu Rekürren Gastroduodenal İntussepsiyon. Clin Exp Health Sci 2017;7:178-80.

10. Miettinen M, Lasota J. Gastrointestinal stromal tumors: review on morphology, molecular pathology, prognosis, and differential diagnosis. Arch Pathol Lab Med. 2006;130(10):1466-78.

11. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. Radiographics. 2003;23(2):283-304, 456; quiz 532. doi: 10.1148/rg.232025146.

12. Gurram RP, Gnanasekaran S, Midha K, Biju P, Kalayarasan R. Atypical Presentation of Gastrointestinal Stromal Tumor as Multiple Intra-Abdominal Cysts: A Case Report. Cureus. 2020;12(5):e7999.

13. Hatipoğlu E, Ergüney MS, Ertem M. Evaluating the laparoscopic approach to proximally located benign gastric tumors. Laparosc Endosc Surg Sci. 2020;27(1):9-15.