

The Role of Nursing in Ruptured Gastrointestinal Stromal Tumor in Adolescents and Young Adults: A Case Report

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Abstract

Gastrointestinal stromal tumors (GISTs) are rare life-threatening forms of cancer representing 0.1%-3% of all gastrointestinal (GI) malignancies. Nursing has become a critical component in handling patients with gastrointestinal tumors. This case report investigates the role of nursing in ruptured GISTs in adolescents and young adults (AYA).

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Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy

Abstract

Gastrointestinal stromal tumors (GISTs) are rare life-threatening forms of cancer representing 0.1%-3% of all gastrointestinal (GI) malignancies. Nursing has become a critical component in handling patients with gastrointestinal tumors. This case report investigates the role of nursing in ruptured GISTs in adolescents and young adults (AYA). It focuses on a 17-year-old female who was admitted to the emergency department (ER) due to the sudden onset of generalized abdominal pain, nausea, and vomiting. After an initial examination, the patient underwent an urgent laparotomy. During this procedure, an unexpected ruptured cystic outgrowth was found originating from the distal ileum. Resection and an anastomosis of the affected section were performed, and a pelvic drain was inserted. GISTs are malignant mesenchymal tumors of the gastrointestinal tract. In rare cases, the tumor may rupture and result in hemoperitoneum; thus, it is challenging for healthcare workers to detect and manage it.

Keywords: gastrointestinal stromal tumor, small intestine, nursing

Background

Gastrointestinal stromal tumors (GISTs) are typically discovered in the gastrointestinal tract and arise in the interstitial cells of Cajal (Joensuu, 2006). The disease mainly affects the stomach (60%-70%) and the small intestines (20%-25%). Its estimated incidence is 10-15 per million (Soreide et al., 2016), with a median age of 65 years at the time of diagnosis. Less than 10% of individuals are diagnosed prior to age 40 (Miettinen et al., 2005; Nilsson et al., 2005). GISTs rarely occur in children and adolescents; they account for only 1.4% to 2.7% of all GIST cases (Casli & Blay, 2010).

Based on statistical analyses, adolescents and young adults (AYA) and older adult (OA) patients differ in terms of the (mutational status, syndrome, and localization of GISTs). AYA most commonly exhibit an acute presentation, thus resulting in more emergencies for AYA compared to OA (18% vs. 9%). Moreover, the overall five-year survival rate is 85% (IJzerman et al., 2020). Mutations in the proto-oncogene commonly drive GISTs, thus affecting more AYA (Nagini, 2012). GISTs have genetic code profiles that change in the genetic structure in *PDGFRA* (6%), in *KIT* (69%), in NF1 associated (4%), in SDH deficient (8%), and in *ETV6-NTRK3* gene fusion (2%). Genetic research has revealed that sporadic genetic changes, including other gene mutations, are divided into four succinate dehydrogenase SDH units (Meyers et al., 2005).

For the clinical presentation, many GISTs are asymptomatic and identified incidentally. Symptoms depend on the size and location of the tumor (Cichoz et al., 2008). In adolescents and young adults (AYA), some patients have chronic anemia (hemoglobin: <8 g/dl) and associated symptoms such as fatigue and paleness, as well as abdominal pain, vomiting, and

abdominal distention (Rink & Godwin, 2009). In rare cases, the patient has a ruptured GIST and spontaneous hemoperitoneum, requiring immediate intervention (Sugawara et al., 2003).

Adolescents and young adults (AYA) have different characteristics, treatment responses, and psychosocial needs, and these aspects should be addressed in research (Fero et al., 2017). Moreover, a ruptured cyst is a life-threatening circumstance that requires immediate attention (Bucher et al., 2008). However, research on nursing in relation to AYA with GISTs is limited. To fill in this gap in the literature, this case report focuses on ruptured gastrointestinal stromal tumors in AYA from a nursing perspective.

Case Presentation

A 17-year-old female arrived in an emergency department (ED) with the sudden onset of generalized abdominal pain. She was experiencing aching; its frequency was intermittent, and its severity was moderate but aggravated by walking. Her pain score was 6 out of 10 on the numerical pain scale. She was also experiencing nausea and vomiting, both of which began in the morning. The patient was single, a high school student, and unemployed. She had a history of recent anemia detected in another clinic and was prescribed an iron tablet (100mg/day). However, she stopped taking it six months ago and did not follow-up on her condition.

Her vital signs, the body temperature was 37C, her blood pressure was 95/60 mmHg, her respiratory rate was 25 breaths/min, and her radial pulse was 100 beats/min and regular. She weighed 47 kg, and her height was 156 cm. She did not know her previous weight. During the examination, she was conscious, oriented. The abdominal examination showed generalized abdominal tenderness mainly in the lower abdominal area with guarding with decreased bowel sounds. The patient's skin was pale and somewhat cold. The early diagnosis for the patient was a ruptured ovarian cyst versus a ruptured viscus.

Investigations

Lab investigations showed a hemoglobin concentration (HGB) was 5.9g/dl [NR, 12.9-14.2g/dl], the white blood cell (WBC) count was $9.67 \times 10^3/\text{ul}$ [NR, $3.7-10.1 \times 10^3/\text{u l}$], and the platelet count was $708 \times 10^3/\text{ul}$ [NR, $155-366 \times 10^3/\text{u l}$]. A hepatic test showed normal value of aspartate aminotransferase (AST) and alanine aminotransferase (ALT). A renal function exam indicated a blood urea nitrogen (BUN) level of 3 mg/dl [NR, 7-18mg/dl], a creatinine level of 0.46mg/dl [NR, 0.55-1.3mg/dl] and a serum albumin level of 3.10q/dl [NR, 3.4-5q/dl]. The test for C reactive showed a level lower than 6 mg/L [NR, less than 10 mg/L]. Her iron level was 14 uq/dl [NR, 50-175uq/dl], her iron binding capacity level was 185 uq/dl [NR, 250-450 uq/dl], and her ferritin level was 2.34 mg/ml [NR, 13-400 mg/ml].

An abdominal ultrasound showed that the right adnexal soft tissue mass measured 5.5 x 4.5 cm, indicating a mild to moderate free pelvic abdominal turbid collection. Based on the ultrasound, the patient underwent an urgent exploratory laparotomy. During the laparotomy, an unexpected ruptured cystic outgrowth was found that originated from the distal ileum (Figure 1). It had an indurated bowel wall area underneath it as well as a moderate amount of blood-free fluid. Consequently, resection and an anastomosis of the affected segment were performed, and a pelvic drain was inserted. The ruptured cyst was sent to the histopathology lab, and its examination revealed a gastrointestinal stromal tumor (GIST) and a mixed tumor of ileum. The tumor size was 6.5 x 3.5 x 2 cm. For the unifocal tumor, the mitotic rate was 36/5 MM2, with a histological grade 2, which is a high grade. The mitotic rate was >5/5MM2 with a risk assessment of high risk. The mucosal resection margins were free of tumors. However, tumor cells were seen on the ruptured cyst's wall, and the cyst contents indicated pathological stage 3.

Treatment

The treatment of the case involved many aspects. In the ER, the patient received intravenous fluids to prevent hypovolemic shock. She was given two units of packed RBCs to correct her hemoglobin level prior to the operation. Intra-operation, because of the ruptured tumor, the hemoperitoneum was managed through laparotomy via resection, an anastomosis, and the placement of a surgical drain. On the first day post-operation, the patient received two units of packed RBCs while in intensive care. To prevent complications related to wound infection, she was given antibiotic prophylaxis—specifically, ceftriaxone 1g, a daily IV for five days, and analgesics including pethidine 50 mg, IM, 12 hours for the first 24 hours and paracetamol 1,000 mg, IV, eight hours for the first seven days. Since the patient was a teenager and had a serious and rare illness, she was provided with good professional psychosocial support from healthcare workers and her family, and this aspect was an important part of her treatment.

Based on the literature, the basic method and the best way to manage GISTs is complete surgical resection (Bucher et al. 2006, 2008; Van der zwan & Dematteo, 2005). Research conducted in the United States on 392 AYA diagnosed with GISTs showed that when 332 (84%) of the AYA population underwent surgical management, their GIST-specific survival (GSS) and overall survival (OS) rates improved (Fero et al., 2017). Other research has shown that Imatinib, which is a drug that inhibits the kinase enzyme activity of a Kit, is an effective first-line medical therapy for post-operative therapy, malignant metastasis, and recurrent GISTs (Cavnar et al., 2021; Han et al., 2015; Griffin et al., 2005). Imatinib improves the survival rate; thus, the median survival rate was seven years, and the lowest survival rate was less than one year (Fauske et al., 2020).

Outcome and Follow-up

Because the patient had a sudden GIST rupture, surgical intervention was necessary. A multidisciplinary approach was used to achieve optimal care and to reach the desirable outcomes. First, immediately after the operation, the patient was shifted to the intensive care unit (ICU) for close monitoring. The plan was to continue replenishing the blood volume with two units of packed RBCs, but a hypokalemia occurred as a complication of dehydration. She was given intravenous fluids (Dextrose 5% normal saline with 40 mcq of KCL 100ml/hr) for 24 hours. A Foley's catheter was attached pre-operation to monitor her kidney functions; after 24 hours, the output was 2,400 ml of clear, yellow, and the catheter was disconnected. Thereafter, the patient was moved to the surgical ward to continue her management. The plan was to continue monitoring the patient. This monitoring included a focus on her surgical drain outcomes, which were as follows: 300 ml bloody exudate on the first day, 250 ml serous on the second day, 300 ml serous on the third day, 100 ml serous on the fourth day, 80 ml on the fifth day, 50 ml serous on the sixth day, and less than 20 ml on the seventh day, when the device was disconnected.

The wound dressing was changed on the second and seventh days. The wound was clean, with no signs of infection. The patient was referred to a social worker to give appropriate support to the patient and her family. Another type of medication was mentioned during the treatment part. An abdominal CT scan with contrast was performed on the fourth day to check metastasis. The scan revealed a multiple hepatic focal

soiled lesion visible on the right lobe hepatic segment, though bowel thickening was not detected. On the ninth day, the patient was discharged, and no surgical complications were perceived. She was given an urgent referral for the oncology center as well as instructions regarding diet, exercise, medication, and wound care. Regarding the follow-up, the patient assessment indicated she was high risk. Therefore, she was instructed to follow-up and to have a CT scan every four months for two years. She was also told to continue treatment in the tertiary center that specialized in oncology.

Discussion

In this case report, we discussed the rare case of ruptured gastrointestinal stromal tumors in adolescents and young adults. Specifically, we detailed the case of a 17-year-old female who was admitted to the ER with simple nausea, vomiting, and generalized abdominal pain. Based on the patient's history as well as an examination and an ultrasound, the working diagnosis was a ruptured ovarian cyst versus a ruptured viscus. A surgical intervention occurred to manage the hemoperitoneum. Post-surgery, the patient had no complications and was discharged on the ninth day post-operation. To manage the same cause, an experienced multidisciplinary team is necessary. This team should include histopathologists, radiologists, surgeons, and oncologists, as well as gastroenterologists, nuclear medicine specialists, and clinical nurse specialists, who play a vital role. The role of nursing starts with the arrival of patients in the ER; the symptoms vary depending on the patient, the size of the tumor, and the location of the tumor. Patients may have non-specific symptoms, including upper gastrointestinal bleeding, anemia, abdominal pain/discomfort, and a palpable mass (Judson et al., 2017). In some adolescents and young adults (AYA), fatigue and weakness may occur due to iron deficiency anemia. Moreover, if the tumor originates in the intestine, constipation may occur (Rink & Godwin, 2009).

In the present case, the patient had a hemoperitoneum, which stemmed from the ruptured cyst. The clinical guidelines for nursing require the immediate application of an intravenous fluid to prevent shock and the observation of the patient's condition. When replenishing the blood volume, the nurse should prepare the patient for diagnostic procedures, such as an abdominal B-ultrasound or a CT scan, that detect GISTs. Thereafter, the nurse should prepare for an emergency operation and communicate with operating room staff. Research has indicated that most patients are not diagnostic prior to surgery (Han et al., 2015). This finding is consistent with the present case, which featured an abdominal B-ultrasound. The results helped to verify the working diagnosis, but the accurate diagnosis was made after the laparotomy and histopathologic sample results.

In the early post-operative period, the nurse has to monitor the circulatory, respiratory and urinary systems of the patient for the early detection of life-threatening complications. It is important to prevent complications related to a compromised immune system as well as hospitalization due to illnesses such as pneumonia. A nurse should also monitor a patient's pain and anxiety levels after the surgical procedures (Griffin et al., 2005). After that, the nurse must focus on patients' early mobilization and should help with the incision treatment (Han et al., 2015). In the later period, the nursing role involves helping with the gradual restoration of body functions such as self-adherence to a diet and medications, monitoring a patient's behavior, providing a health education seminar, and offering the patient appropriate psychosocial support (Griffin et al., 2005).

In conclusion, a ruptured GIST in adolescents and young adults is an emergency situation that requires immediate intervention from expert nursing staff and a physician. An active examination, diagnosis, and treatment may help increase the AYA's chance of survival. Further studies are needed to focus on the risk factors, early detection of the disease, and long-term follow-up.

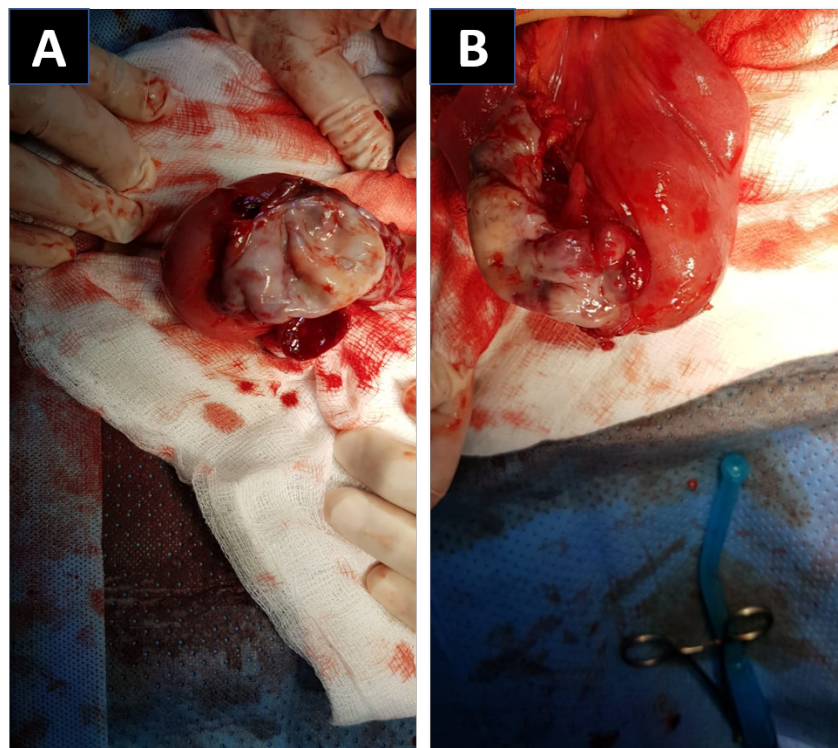


Figure 1 (A, B): Ruptured cystic outgrowth from the bowel wall.

Figures:

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