CASE REPORTS

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Gastrointestinal Stromal Tumor in Pregnancy: A Rare Case with Review of Literature

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors that develop in the wall of the GI tract. The aim of this study was to report a case with GIST in pregnancy in Western Iran. A 39-year-old pregnant woman (G5 P4) was admitted with a chief complaint of abdominal pain and epigastric fullness on April 12, 2016. Sonography showed gestational age of 15w+1d and a solid cystic intra-abdominal mass extending from the epigastrium in the midline to the left upper quadrant. On April 20, abdominal MRI showed a solid mass measuring 24x15x12.5cm with smooth and well-defined borders. On May 29, the pathology report showed a gross large mass measuring 24x19x10cm attached to a segment of the stomach measuring 20x8cm with heterogeneous cystic hemorrhagic cut surface of the tumor. Microscopic pathology reported compatible with gastrointestinal stromal tumor, group 3b (Intermediate risk) according to Miettinen risk classification. In conclusion, GIST in pregnancy is very rare and therefore, there is little diagnostic information about its risk factors, but CD117+/ CD34+ can be an important diagnostic sign for pregnant GIST.

Keywords: Gastrointestinal stromal tumor, Pregnancy, Case report

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Introduction

Gastrointestinal stromal tumors (GISTs) are rare[1] and have been diagnosed as a biologically distinctive tumor type, different from smooth muscle and neural tumors of the GI tract[2]. These tumors are mesenchymal tumors that develop in the wall of the GI tract[3] and their diagnosis during pregnancy or puerperium is extremely rare[3-5]. Nowadays, diagnosis of GISTs is based on histopathological features, including the immunohistochemical (IHC) staining of CD117, DOG-1, CD34, SMA, desmin and S-100[6]. The incidence of GISTs is approximately 10 to 20 cases per million people per year. More than 90% of these tumors occur in patients over 40 years of age (fourth and eighth decades) and the median age is 63 years;[7,8] therefore, they are rarely diagnosed in pregnancy[8]. GISTs occur anywhere in the intestine; the most common site is the stomach (50-60%), followed by the small intestine (20-30%), large bowel (10%), the esophagus (5%), and only 5% elsewhere in the abdominal cavity such as the mesentery, omentum or retroperitoneum[9]. Herein, we report a case with GIST in pregnancy in Western Iran.

Case report

A 39-year-old pregnant woman (gravida 5 para 4 living4) was admitted to the gynecology and obstetrics service with chief complaint of abdominal pain and epigastric fullness on April 12, 2016. She had referred to a gynecologist with a sonography on April 11, 2016 that showed gestational age of 15w+1d and a solid cystic intra-abdominal mass extending from the epigastrium in the midline to the left upper quadrant. In her past medical history, the patient had 4 NVD (normal vaginal delivery), appendectomy 7 years ago and hypertension 4 years ago (maximum 170/100 mmHg). The patient was discharged on April 16, for further outpatient work-up with abdominal MRI requested. On April 20, abdominal MRI showed a solid mass measuring 24x15x12.5cm with smooth and well-defined borders with multiple cystic changes on the left side of the abdominal cavity with no connection to liver, spleen, kidney or pancreas of probable mesenteric or ileal origin. Leiomyosarcoma or leiomyoma was suggested on MRI and lymphadenopathy was noted in retroperitoneum. The lab data were within normal limits for WBC, platelets, urea, Cr, Na, K, carcinoembryonic antigen (CEA) and carbohydrate antigen 125(CA125). Serum α-fetoprotein (AFP) was 39.1 ng/dL and beta-hCG titer was 15835 mIU/mL. The patient consulted with an onco-surgeon. On May 8, 2016, radical subtotal gastrectomy was performed for the mass in the lesser curvature of the stomach and retroperitoneal lymphadenectomy was performed for staging of the tumor. On May 9, sonography of the uterus showed a live fetus with variable presentation, posterior fundal placenta, appropriate amniotic fluid for age and gestational age of 18w+5d according to femur length. The patient was discharged on May 16, 2016 with good conditions. The pathology report on May 29, 2016 showed a gross large mass measuring 24x19x10cm attached to a segment of the stomach measuring 20x8cm with heterogeneous cystic hemorrhagic cut surface of the tumor (**Figure 1**). Microscopic pathology reported compatible with GIST, group 3b (Intermediate risk) according to Miettinen risk classification10 (**Figure 2**). Mitotic rate was less than 5/50 high power field. Tumor location was in the stomach with free surgical margins and all nine isolated lymph nodes. IHC report on August 6, 2016 confirmed histologic diagnosis with CD117(c. KIT) and myeloid stem cell antigen (CD34) positivity (**Figure 3**), but neuron-specific enolase (NSE) and alpha-smooth muscle actin (SMA) negativity. The patient insisted on continuing the pregnancy. The patient and fetus were in good conditions on August 6, 2016.

Discussion

This study reports a case of GIST in pregnancy (a 39-year-old woman) with high serum AFP in whom

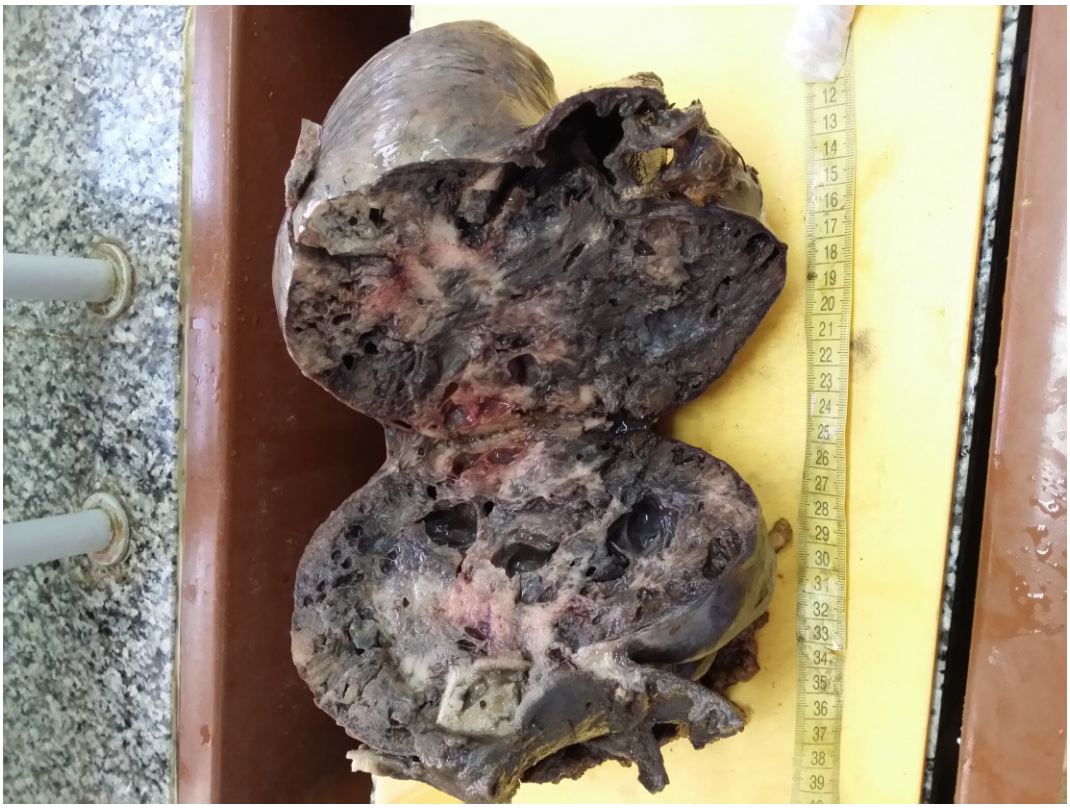

Figure 1: Gross view of large mass measuring 24x19x10cm attached to a segment of the stomach

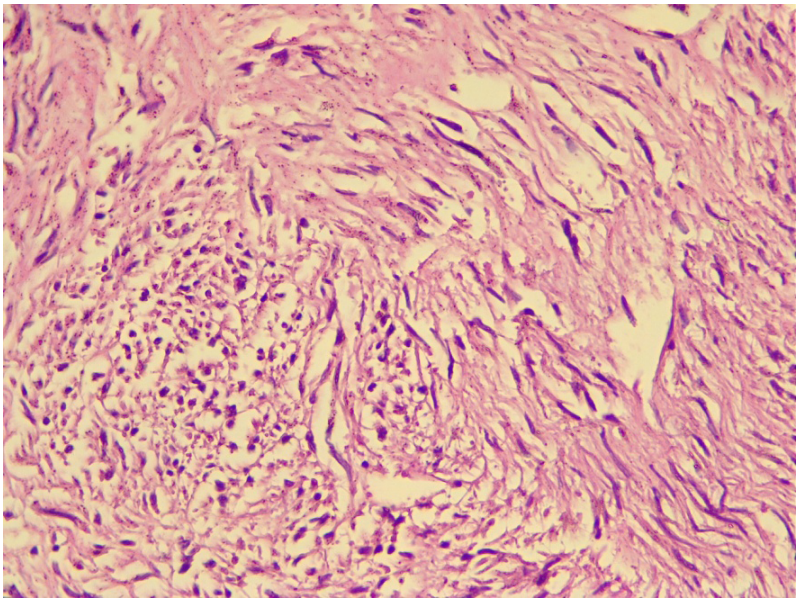

Figure 2: GIST(x400 magnification), Hematoxylin & Eosin staining

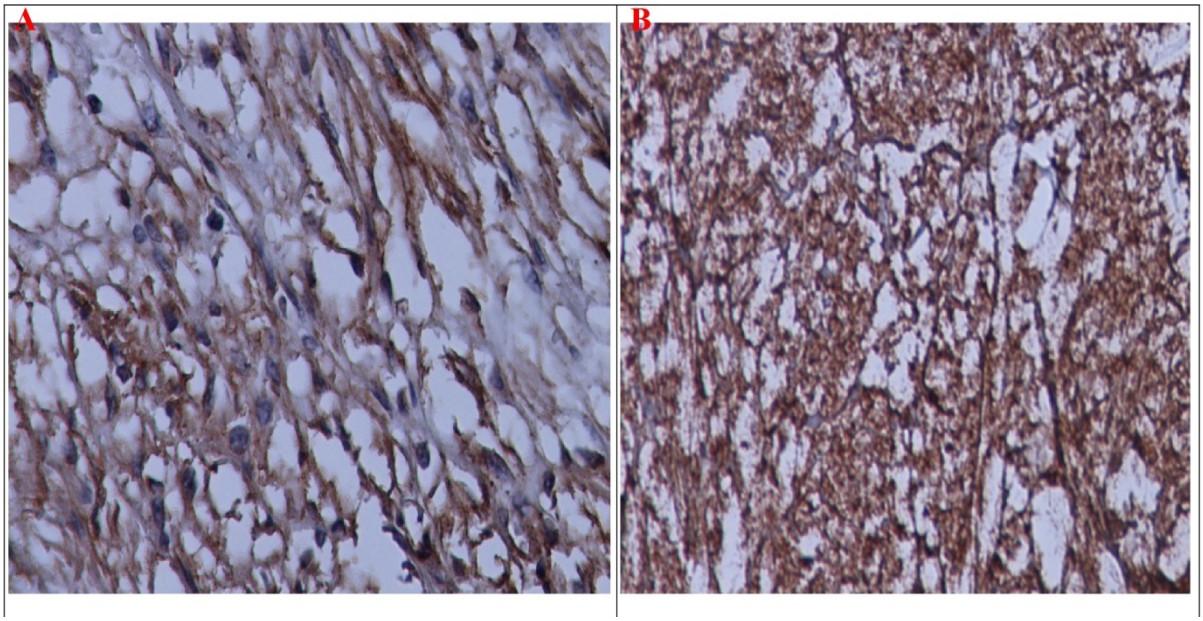

Figure 3: Immunohistochemical positive staining in GIST (x400 magnification), (A) CD117, (B) CD34

CD117 and CD34 were positive, while NSE and SMA were negative. Also, the size of the tumor and its mitotic rate were 24x15x12.5cm and less than 5/50HPF, respectively, and tumor location was in the stomach (intermediate risk) with free surgical margins (0/9 lymph node involvement). Approximately 80% of GISTs have CD117 positivity[3,11-13]. Therefore, CD117 cannot be specific for GISTs[14]. Riddle et al.[13] reported that CD117 and CD34 positivity are the immunohistochemical signs of GISTs. Out of 10 patients with GIST in one study, all had CD34 positivity[12]. Two case reports of GIST[15,16] showed that Immunohistochemical staining for CD117/CD34 was, weakly+/+ and +/+, respectively, and also, SMA was checked in two cases that were negative[15,16] while NSE was slightly positive[15]. One study[12] reported a total of 12 patients diagnosed with GIST: the male-to-female ratio was 1:1 and mean age was 68.2 ± 7.0 years. A systematic review between January 2000 and December 2014 on 13550 GIST patients[1] showed that the median age at diagnosis was mid 60s across most studies (range, 10 to 100 years) and sex distribution was equal across studies. The age/sex of patients in two GIST cases[15,16] was 65 years/male and 67/female, respectively. The cases presented in **Table 1** show that the presentation of GIST in pregnant women is between 25 to 42 years. Also, CD117 in all checked cases and CD34 in most cases are positive, while SMA is negative in most checked cases.

Scherjon et al.[12] reported that CD117 is positive in approximately 85%–90% of GIST cases. CD34 is positive in 60%–70%. SMA may be present in 20%–40% of the cases. SMA positivity has been often in the opposite direction with CD34 expression[12]. Lin et al.[9] showed that the levels of tumor markers, such as CA19-9, CA 125, CEA and AFP, were normal in GIST case that CEA and CA 125 were normal in this case.

Conclusions

GISTs are rare and GIST during pregnancy is ex-

Table 1: Cases of GIST in pregnancy

Reference	Age of pregnancy	CD117	CD34	SMA	NSE	Site	Risk
3	28	P	P	P	-	Small intestine	High
5	29	P	P	N	-	Stomach	Low
8	31	P	P	P	-	Large bowel	High
11	32	-	P	N	-	Stomach	Low
17	25	P	P	P	-	Small intestine	High
18	28	P	P	N	-	Gastro-esophageal	High
19	42	P	N	-	-	Retroperitoneum	Low
The present case	39	P	P	N	N	Stomach	Inter

Abbreviations: P, positive; N, negative

tremely rare. Therefore, there is little diagnostic information on its risk factors. Neverhteless, CD117+/CD34+ can be an important diagnostic sign for pregnant GIST. Also, the researchers can in future studies focus on prognostic factors in pregnant GIST patients.

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