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Gastric stromal tumor with pregnancy: A case report

Gebelikte gastrik stromal tümör: Olgu sunumu

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Abstract

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors that develop in the wall of the gastrointestinal tract. There is few given information throughout literature concerning gestation associated with GIST. We report here the case of a patient who was admitted in our hospital for GIST discovered during the second trimester of pregnancy. She was 44 years old, in the fifth months of pregnancy. She was admitted for biliary colic pain. An abdominal examination objectified a distended abdomen with uterine height of 15 cm and right hypochondrium mass of 15 cm. Abdominal ultrasound and magnetic resonance imaging showed a large lesion process in the right hypochondrium region. Endoscopy objectified an aspect of extrinsic compression at the front of the stomach. The patient was operated and an umbilical median incision was performed, exploration showed the presence of a hug mass measuring 25 cm occupying almost all of the right hypochondrium, pushing the liver up and the uterus down, adheres to the stomach at the level of the small curvature with an implantation base of about 3 cm. Histological and immune histochemical study of hepatic process showed a GIST of high risk of malignancy. Only some cases have been reported in the literature on GIST during pregnancy showing the rarity of the pathology that requires multidisciplinary care.

Keywords: Pregnancy, Gestation, Imatinib, Gastrointestinal stromal tumor

Öz

Gastrointestinal stromal tümörler (GIST) gastrointestinal sistemi duvarında gelişen nadir mezenkimal tümörlerdir. Gebelikte GIST ile ilgili literatürde az miktarda bilgi bulunmaktadır. Biz burada gebelik ikinci trimesterde tespit edilen bir GIST olgusunu rapor ediyoruz. Olgu 44 yaşında ve hamileliğin beşinci ayında idi. Biliyer kolik ağrısı ile başvurdu. Abdominal muayenede 15 cm'lik rahim yüksekliği ve sağ hypochondrium 15 cm kitle tespit edildi. Abdominal ultrasonografi ve manyetik rezonans görüntüleme ile sağ hypochondrium bölgesinde büyük bir lezyon gösterildi. Endoskopi ile midenin önünde dışsal bası görüldü. Hasta ameliyata alındı ve orta hat kesi yapıldı. Eksplorasyonda neredeyse tüm sağ hypochondrium işgal eden, karaciğeri yukarı ve rahimi aşağı iten, küçük kurvatür düzeyinde mideye yapışan, yaklaşık 3 cm'lik bir implantasyon tabanı olan 25 cm kitle tespit edildi. Histolojik ve immun histokimyasal çalışmalar, yüksek malignite riskli GIST gösterdi. Literatürde gebelik sırasında sadece olgu sunumu olarak bildirilen GIST, multidisipliner bakım gerektiren nadir bir patolojidir.

Anahtar kelimeler: Hamilelik, Gestasyon, Imatinib, Gastrointestinal stromal tümör

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the digestive tract. They most often occur in the stomach followed by the duodenum. The diagnosis of GIST during pregnancy is very rare. There are less than 21 cases reported in literature about gastrointestinal stromal tumors diagnosed during pregnancy one of them reported in our hospital in 2004 by Cherif and all, here we report the second case of a gastric GIST diagnosed during pregnancy in our hospital [1].

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Case presentation

She is a 44-years-old patient, with an unremarkable medical and family history, mother of 5 children, in the fifth month of pregnancy. She admitted for treatment of moderate hepatic colic pain, dating back to seven months, atypical epigastric pain, epigastric fullness relieved by post prandial vomiting.

Clinical examination found a patient in a good general state, a distended abdomen with uterine height of 15 cm, and mass of the right hypochondrium smooth and painful of 15 cm.

Laboratory tests showed a hypochromic microcytic anemia with a biological inflammatory syndrome.

The morphological assessment including abdominal ultrasound has objectified a hepatic cystic and tissue mass, with thickened and irregular contours.

Hepatic magnetic resonance imaging found a large lesion process in the right hypochondrium region; this lesion is heterogeneous with dual component: a liquefied center (rich in mucin and blood) and very irregular peripheral component with irregular and asymmetric internal contours. This mass is localized in the right hypochondrium region, tacking the liver and coming into contact with the stomach wall (small curvature) (Figure 1).

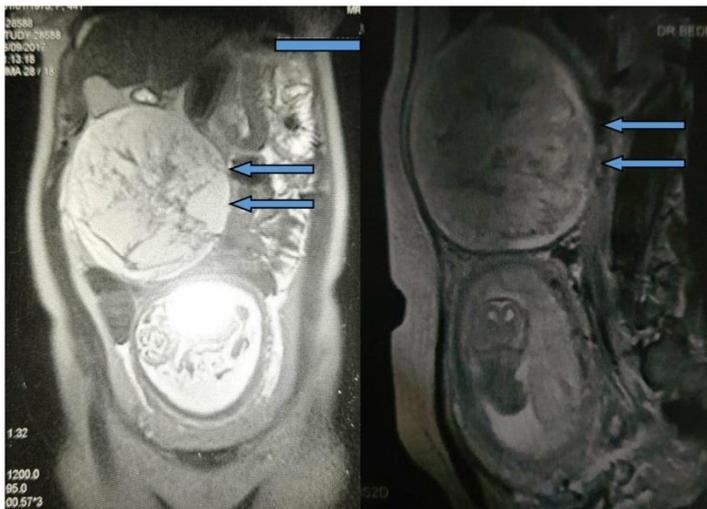


Figure 1: Magnetic resonance images of the stromal tumor and pregnancy.

A pelvic ultrasound has shown an evolutionary single-fetus pregnancy, estimated at 34 weeks of pregnancy. An upper gastrointestinal endoscopy was performed given the gastric compressive aspect of the tumor. It highlighted an aspect of extrinsic compression at the front of the stomach, without mucosa lesions.

After discussing the issue in a multidisciplinary meeting in the presence of Obstetricians, the diagnosis of gastric GIST was suspected. Imatinib is contraindicated in pregnancy so the multidisciplinary decision was to operate the patient during pregnancy and perform an effective tocolysis by the obstetricians during the surgical procedure.

The patient was operated without incident an umbilical median incision was performed, exploration showed the presence of a hug mass measuring 25 cm occupying almost all of the right hypochondrium, pushing the liver up and the uterus down, adheres to the stomach at the level of the small curvature with an implantation base of about 3 cm, after protection of the operative

field by fields soaked with serum the mass was externalized from the abdomen and its implantation base was cut on the stomach with a safety margin of 1 cm then the gastric breach was manually closed (Figure 2).



Figure 2: Left image: view of the tumor before the base section, Right image: view of the tumor after extraction (the red arrow show the base of implantation).

The postoperative follow up were uneventful. The histological study confirmed the diagnostic of GIST. Resection's limits of stomach were healthy and the postoperative decision was to start the imatinib immediately after delivery.

Discussion

The incidence of GIST is approximately 10 to 20 cases per million people and year. The diagnosis must be made early to preserve the prognosis. They are much rarer during pregnancy; few cases have been reported in the literature [2].

The incidence of these tumors in the United States is estimated at 3000 - 4000 cases per year, with a median age of 60 years [3]. The majority of GISTs are localized in the stomach (60%) and small intestine (30%), especially when associated with pregnancy [2]. the remaining 10% are located in the esophagus and rectum [4]. Their occurrence is sporadic in most cases, but there are some familial predispositions, such as neurofibromatosis type I and exceptional familial forms described by Carney and Stratakis or related to a constitutional mutation of KIT or PDGFRA [5].

Association of GIST and pregnancy is rare. Also the challenge is to make the diagnosis early and begin treatments that preserve the pregnancy.

The clinical presentation during pregnancy is not specific: gastrointestinal bleeding, unexplained anemia or abdominal mass. Cases of GIST reported in the literature, show a non-specific symptoms. However, only the histological analysis allows confirming the diagnosis [6].

GISTs are usually well-circumscribed without encapsulation; they grow preferentially on the serosal side of the bowel wall. Macroscopically, the measurement of the maximum diameter of the primary tumor is an important parameter for evaluating the evolutionary potential. It is important to sample the tumor, for the differential diagnosis with other sarcomas (e.g. liposarcoma), and because there may be variations in the proliferation index [7]. Histologically, cell density is generally high and homogeneous, and necrotic alterations, edema and/or bleeding are as most frequent as tumors are large. The cells are fusiform in 70% of cases, most often with a fascicular architecture, suggesting a smooth muscle proliferation [8].

The average diameter of symptomatic tumors is 6 cm against 1.5 cm for asymptomatic tumors. Our patient had a tumor exceeding 25 cm. The cases reported in the literature had tumors diameter ranging between 4 and 23 cm [9].

Useful tests for the diagnosis of GIST depend on the size and location of the tumor. For tumors less than 5 cm and gastric or colorectal localization, the diagnosis is made by endoscopy and confirmed by ultrasonography. For small GIST of the small intestine, the diagnosis is made by enteroscopy. In the case of very large GIST, the gold standard is the abdominal computed tomography or magnetic resonance imaging in pregnancy, which was performed in our case [9].

All GISTs are potentially malignant, and the risk of recurrence after resection can be assessed according to the size and mitotic index. It is likely that other parameters, such as gastric localization, presence of necrosis and the type of mutation, have prognostic value [10].

GIST metastases are localized in the liver in 2/3 of cases and in the peritoneum in one quarter of cases. Lymph node metastases are rare, not justifying their dissection when the diagnosis is suspected. Lung metastasis are also rare, and their occurrence may justify reviewing the diagnosis [6].

Unfortunately, due to the rarity of the disease there are no recommendations for the management of GIST during pregnancy [11]. In review of literature a total of 12 cases were retrieved regarding women who were diagnosed with GIST during the gestational period, eleven out of the twelve pregnant women had laparotomy and/or surgical excision of the tumor, Two patients underwent pregnancy termination at 7 and 15 week of gestation, while on treatment with imatinib, Seven patients started imatinib postpartum such as we describe in our case. Six women remained disease free from 9 to 36 months and two patients with advanced disease showed partial response and stable disease, respectively. There are no reported cases with metastatic involvement of placenta or fetus [12].

The primary treatment of choice for localized or resectable GIST is surgery. The best chance for cure is complete surgical excision of the tumor in clear margins [13-15].

Imatinib mesylate is a tyrosine kinase inhibitor that selectively inhibits BCR-ABL KIT and PDGFR tyrosine kinases and has become the standard of care on adjuvant setting as well as in metastatic disease. The introduction of kinase inhibitors has increased survival of patients with GIST with an approximate median survival of 5 years [16,17].

The safety of imatinib administration during pregnancy is yet to be documented. Current experimental data show teratogenic effects when administered in female rats during the organogenesis period [18,19].

Published data by Pye et al [16], concerning imatinib administration during gestational period in 180 pregnant women revealed that only 50% of them had an uneventful infant delivery, whereas the rest ended either in elective termination or in spontaneous abortion [20,21].

Due to the limited number of cases described throughout the international literature, we cannot reach conclusions concerning the safety of imatinib administration in pregnant women with GIST.

Another significant factor that must be taken into consideration when treating pregnant women with cancer is the time of delivery. During a pregnancy complicated with cancer, the optimal time for delivery is placed between the 35th and the 37th week of gestation.

In conclusion, preterm delivery should be avoided if possible and one must always account for premature neonatal complications. Thus, a multidisciplinary approach involving surgeons, obstetricians and oncologists should take place under the scope of both maternal and fetal health. In addition, the patient's perspective regarding the therapeutic plan should be accounted for.

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