OLGU SUNUMU CASE REPORT

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Adnexal Torsion Mimicking a Familial Mediterranean Fever Attack in a Pregnant Woman

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ABSTRACT Familial Mediterranean Fever (FMF) is the most common hereditary auto-inflammatory disease that is characterized by recurrent, self-limited serousitis attacks and fever. Differential diagnosis of an abdominal attack of FMF with acute abdomen is difficult. A 31-year-old primigravid with FMF referred to us with right lower abdominal pain at the 25th week of gestation. Her medical history seemed like it was one of the FMF attacks but a salpingo-oophorectomy had to be performed because of the delaying of diagnosis of adnexal torsion in pregnancy. Furthermore, once a patient is diagnosed with FMF, there may be a tendency among physicians to relate each abdominal pain to an FMF attack, whether or not that is the case. Therefore, we suggest that every individual abdominal pain should be considered independent of other pains, even if a patient has been diagnosed with FMF. In this manner, other possible causes of abdominal pain can be taken into consideration.

Keywords: Abdomen, acute; familial mediterranean fever

Familial Mediterranean fever (FMF), the most common hereditary, monogenic autoinflammatory disease, is characterized by recurrent, self-limited attacks of fever and sterile inflammation of serous membranes. Tel Hashomer criteria are commonly used in its diagnosis.[1,2] Attacks of FMF are typically accompanied by leukocytosis and elevated C-reactive protein (CRP) and other inflammatory parameters. They often mimic the clinical signs of an acute abdomen, and they can be indistinguishable from an acute abdominal emergency. Frequent surgical interventions have been reported in FMF patients.[3,4]

FMF attacks can be triggered by common factors such as cold exposure, emotional or physical stress, infections, and menstruation. A typical attack usually lasts 12–72 hours. In a small percentage of patients, the attacks are characterized by afebrile recurrent abdominal pain.[1] Conversely, fever is sometimes the only manifestation of an attack.[5]

Pregnancy is a risk factor for ovarian/adnexal torsion (AT) that is seen most commonly between 10 and 17 weeks of gestation. The diagnosis of AT is often missed during pregnancy due to nonspecific clinical features and uncommon objective findings, especially in the second and third trimesters. In addition, the diagnostic accuracy of ultrasound is limited because the ovaries are displaced from their normal positions.[3]

In this report, we present an unusual case of a pregnant patient with FMF who was misdiagnosed with an FMF attack. The patient underwent salpingo-oophorectomy at 25 weeks of gestation due to a delay in diagnosing AT in pregnancy.

CASE REPORT

A 31-year-old primigravida patient in the 25th week of gestation was referred to our clinic with abdominal pain, nausea, and vomiting that had persisted since the previous day. In her medical history, it was learned that she had experienced periodic abdominal pain and fever attacks since childhood, and that she underwent an appendectomy at the age of 17 years due to severe abdominal pain. There was a family history of periodic fever and abdominal pain attacks; the patient was diagnosed with FMF, and she had been taking colchicine since 19 years of age.

The patient stated that she had discontinued the drug when she conceived because of its possible adverse effects on her fetus, but she had not experienced any attacks during the pregnancy. She also indicated that she had suffered with nausea and vomiting often during the pregnancy. The patient was diagnosed as hyperemesis gravidarum and hospitalized.

The patient stated that she had experienced periodic lower abdominal pain with nausea and vomiting for the past day; she had no vaginal bleeding or discharge. An ultrasound showed a live fetus compatible with 26 weeks' gestation, but no bilateral adnexa were observed. No cervical dilatation was evident on vaginal examination, and tocography revealed no premature contractions. On physical examination, the patient was subfebrile. The abdominal examination was remarkable for tenderness to palpation, exclusively to the right lower quadrant, with voluntary guarding. There was no rebound tenderness. Bowel sounds were within normal limits. The laboratory results showed a slightly elevated white blood cell (WBC) count of 14,000/mm^3 (normal range: 4,000–11,000/mm^3, increasing to 15,000/mm^3 in the second trimester of pregnancy is acceptable) and a high-sensitivity (hs)-CRP level of 17 mg/ L (normal range: 3–10 mg/ L). Hemoglobin, hematocrit, platelet count, and urine analysis were normal.

The patient was hospitalized at 25 weeks of gestation with a diagnosis of acute abdomen, possibly due to FMF attack. Colchicine was started with a dose of 0.5 mg x 2/day.

During the two days since the onset of pain, there was no moderation in the patient's complaints, and her pain even intensified. The patient had a mild fever, and the WBC and CRP levels also continued to rise (WBC count: 17,000/mm^3; hs-CRP level: 22 mg/L).

An ultrasound examination performed the next day showed an enlarged and edematous right ovary; no blood flow could be detected in the right ovary on color and power Doppler ultrasound. Therefore, a laparotomy was performed due to right ovarian torsion at 25 weeks and 3 days of pregnancy. A laparoscopy was not performed due to difficulties that limit laparoscopic surgery in the second and third trimesters. During the laparatomy, extensive adnexal ischemia (Figure 1) was observed. Firstly, detorsion of the twisted adnexa was tried. However, because the necrotized tissue spontaneously ruptured, the right adnexa had to be removed. The patient presented an uneventful postoperative clinical course and had a normal delivery of a healthy infant at 38 weeks of gestation.

DISCUSSION

FMF is usually a childhood-onset disease, mostly between 9 and 16 years of age.[1] Pregnancy does not seem to change the severity and duration of FMF attacks, and the course of pregnancy in patients with FMF is variable. Some patients have an attack-free period, while other women may experience attacks with increased frequency.[6] Colchicine is one of the oldest known drugs used for prophylaxis of FMF attacks and prevention of the development of amyloidosis. Although colchicine crosses the human placenta, there have been no reports of unusual frequency of fetal abnormalities among

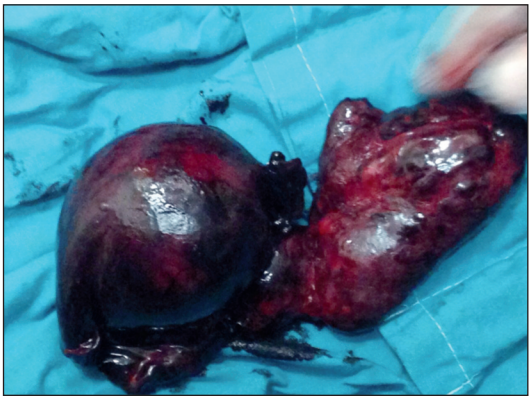

FIGURE 1: Right adnexa with extensive ischemia during surgery.

women taking colchicine before or during pregnancy. Studies have suggested that it does not appear to be a major human teratogen and probably has no cytogenetic effect.[7] The general recommendation is that colchicine should be continued during pregnancy. Researchers are divided on recommending amniocentesis on a routine basis to determine the fetal karyotype.[7] Our current policy is to recommend continuous colchicine before conception and during pregnancy. Where feasible, it is advisable to perform first- and second-trimester combined aneuploidy screening tests.

The incidence rate of AT is unknown in both pregnant and non-pregnant women. A retrospective study indicated that the incidence of torsion during pregnancy is 0.2%.[8] AT is most likely to occur when the ovary is 5 cm in diameter or larger[5] and with benign cysts or neoplasms rather than malignant lesions, possibly because malignant masses are more likely to be fixed in place. Ovarian torsion can also occur in patients with normal ovaries, especially in premenarchal girls, caused by an elongated utero-ovarian ligament.[8] The symptoms and findings of AT in pregnancy are pelvic pain, adnexal mass, nausea, vomiting, low-grade fever, and leukocytosis. Both transvaginal and transabdominal ultrasounds should be obtained in all patients to visualize the abdominal and pelvic structures. An ovarian mass, an enlarged ovary, heterogeneous appearance of the ovarian stroma, multiple small peripheral follicles with non-echogenic stroma, abnormal ovarian location, and decreased or absent Doppler flow within the ovary can be found via ultrasound scanning. Magnetic resonance imaging and computed tomography may be helpful, but they are not used routinely in these cases. A definitive diagnosis of ovarian torsion is made by direct visualization of a rotated ovary at the time of surgical intervention.[8] A laparoscopic intervention is preferred if there is no suspicion of ovarian or fallopian tubal cancer. Detorsion should be the decision if a patient is premenopausal with a viable ovary. The important point is to perform the procedure as quickly as possible, because infection or pulmonary embolism can complicate the detorsion if there is latency.[9] A salpingo-oophorectomy should be performed if the ovary is not viable, if there is a suspected malignancy, or if the patient is postmenopausal. If an oophorectomy is performed, progesterone supplementation is recommended until the 12th week of gestation. Prophylactic tocolysis may also be administered, although there is no evidence to support its use. Confirmation and documentation of the fetal heart rate before and after the surgery is required.[10]

A differential diagnosis of acute abdomen in pregnant patients is one of the greatest challenges for the clinician. Our patient had right lower abdominal pain and the signs of acute abdomen. However, she had undergone an appendectomy four years prior, and she was diagnosed with FMF. In addition, she had discontinued her drugs since finding out she was pregnant. Based on her medical history, the condition seemed like an attack of FMF, but the sonographic findings led us to suspect ovarian torsion. We determined that laparotomy would be safer than laparoscopy at the end of the second trimester. Right adnexal torsion with necrotic tissue was detected, and due to the latency, a salpingo-oophorectomy had to be performed.

In summary, it is very crucial to differentiate FMF attacks from acute abdominal emergencies, and the treatment of pregnant patients with acute abdomen should be individualized. Colchicine

should be continued during conception, pregnancy, and lactation in patients with a history of FMF. Abdominal FMF attacks mimic the clinical presentation of acute abdomen, but FMF symptoms always resolve spontaneously. FMF patients may be admitted to the hospital with a dangerous abdominal emergency without an abdominal FMF attack. Good history taking is an absolutely essential diagnostic tool for achieving a precise diagnosis of FMF and other abdominal occurrences.

A high clinical suspicion and early laparoscopic management correlate with favorable maternal and fetal outcomes, it necessitates a prompt surgical intervention, because any delay leads to irreversible ovarian necrosis, ultimately requiring an adnexectomy. Despite technological advances in ultrasonography, the diagnosis of the torsion is difficult, especially during pregnancy, and it sometimes remains a diagnostic dilemma. Furthermore, once a patient is diagnosed with FMF, there may be a tendency among physicians to relate each abdominal pain to an FMF attack, whether or not that is the case. Therefore, we suggest that every individual abdominal pain should be considered independent of other pains, even if a patient has been diagnosed with FMF. In this manner, other possible causes of abdominal pain can be taken into consideration.

Informed Consent

Informed consent was obtained from the patient for publication of this case report and the accompanying images.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Design: *Ayşe Kırbaş;* ***Control/Supervision:*** *Turhan Çağlar;* ***Analysis and/or Interpretation:*** *Erdinç Sarıdoğan;* ***Literature Review:*** *Erdinç Sarıdoğan, Özgür Kara;* ***Writing the Article:*** *Ayşe Kırbaş;* ***Critical Review:*** *Korkut Dağlar.*

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