Science Journal of Clinical Medicine
2021; 10(4): 143-146
http://www.sciencepublishinggroup.com/j/sjcm
doi: 10.11648/j.sjcm.20211004.21
ISSN: 2327-2724 (Print); ISSN: 2327-2732 (Online)

Essential Thrombocytosis Detected in Pregnancy: A Case Report

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To cite this article:
Behshad Pazooki, Ailar Ahangari. Essential Thrombocytosis Detected in Pregnancy: A Case Report. *Science Journal of Clinical Medicine*. Vol. 10, No. 4, 2021, pp. 143-146. doi: 10.11648/j.sjcm.20211004.21

Received: October 22, 2021; **Accepted**: December 11, 2021; **Published**: December 24, 2021

Abstract: Essential thrombocytosis is a myeloproliferative disease of unknown reason that causes progressive platelet overproduction, which this high counts of platelets appear to be associated with intravascular thrombosis and related event. There are several medications to prevent thrombosis formation in these patients, such as hydroxyurea, and aspirin. We do not have much information about the duration of the medication. Our case was a 28-year-old woman without any history of medical problem, who gave birth recently, presented to the emergency department with epigastric abdominal pain, nausea/vomiting, loss of appetite, chills and diarrhea. On initial examination, huge splenomegaly about 20 cm below the edge of the ribs in the midclavicular line were detected. All lab data were within normal ranges. Pregnancy behaves as a trigger for undiagnosed essential thrombosis in this patient. She underwent anticoagulant therapy which still continuous. It seems that pregnancy became a triggering factor for ET in our case to show up with the symptoms like thrombosis. There is lack of evidence about duration of treatment. We don't know when we should stop the anticoagulant therapy or we should continue the medications lifelong. This issue should be considered as the basis of future studies.

Keywords: Essential Thrombocytosis, Pregnancy, Anticoagulants, Myeloproliferative

1. Introduction

Essential thrombocytosis is a myeloproliferative disease of unknown cause involving multipotent hematopoietic progenitor cells in the bone marrow [1, 2] that causes progressive platelet overproduction, which this high counts of platelets appear to be associated with intravascular thrombosis and related events but very high platelet counts can initially be associated with bleeding. However, several studies show that the incidence of thrombotic events in these patients is higher than bleeding [3, 4]. There are several medications to prevent thrombosis formation in these patients, such as hydroxyurea, [5] and aspirin [6]. However, there is a lack of evidence about the duration in which treatment should be continued. We do not have much information about this issue, although researchers have found many links between this disease and some genes, such as the Philadelphia chromosome [7] and lq +, duplication or triplication of q21 to q32 that has been seen on the first chromosome, indicating a meaningful connection [8, 9]. We are going to present a case, which in that, pregnancy played a role as trigger for undiagnosed Essential Thrombocytosis. The treatment will be discussed; however, there is controversies and lack of evidence about sufficient medication use.

2. Case Report

A 28-year-old woman without any history of medical problem presented to the emergency department of Valiasr Hospital, Imam Khomeini Complex with epigastric abdominal pain since 5 days ago. 25 days before the recent visit, she gave birth by cesarean section, during which the patient had no problems and discharged in good condition after delivery. 1 day after the onset of the pain that led the patient to the hospital, nausea, and vomiting and loss of appetite also began. Other accompanying symptoms were chills and diarrhea during this period. On initial examination,

the physician noticed huge splenomegaly about 20 cm below the edge of the ribs in the midclavicular line. The rest of the physical examinations were completely normal. This patient had no other past medical history about excessive bleeding and family history of sickle cell disease, thalassemia, or coagulation-related diseases. In addition, the habitual history of patient was fully negative. The vital signs in arrival time were as follows: blood pressure, 125/95 mmHg, pulse rate, 95 beats/min, regular; temperature, 36.4°C, O_2 saturation, 99%. The results of the lab tests were as mentioned in Table 1, which all were completely within normal range.

Table 1. *Lab data.*

Variable	Finding
WBC	7800 / mm^3
Haemoglobin	11.2 gr/dl
Platelets	210000 mm^3
Na	134 mMol/L
K	4.2 mMol/L
BUN	31 mg/dl
Cr	0.8 mg/dl
SGOT	11 IU/L
SGPT	12 IU/L
LDH	458 IU/L
ALP	223 IU/L
BS	119 mg/dl
CRP	43 mg/dl
CPK	67 IU/L
Amylase	50 IU/L
Lipase	31 IU/L
beta2 glycoprotein IgG	0.4 U/ml
beta2 glycoprotein IgM	1.5 U/ml
anti Cardiolipin IgG	0.3 U/ml
anti Cardiolipin IgM	0.9 U/ml
C3	110 mg/dL
C4	31 mg/dL
viral markers were nonreactive	

* BUN: Blood Urea Nitrogen, Cr: Creatinine, SGOT: Serum Glutamate oxaloacetate transaminase, SGPT: Serum Glutamate pyruvate transaminase, LDH: Lactate Dehydrogenase, ALP: Alkaline Phosphatase, BS: Blood Sugar, CRP: C Reactive Protein, CPK: Creatine Phosphokinase.

At the initial examination of the patient, due to the possibility of perforation of the internal viscera, Chest X-ray was performed. No Pneumoperitoneum was detected, but cardiomegaly was reported. Echocardiography was performed which shows a mild pleural effusion suggestive of infection. The results showed no infection. Abdominal ultrasound was performed, which showed massive splenomegaly measuring 143 x 226 mm^2. In addition, a tubular lesion was seen in the medial region of the spleen, suggesting splenic vein thrombosis, and no lesion was seen in the spleen and other intra-abdominal organs. Due to clinical suspicion of mesenteric ischemia, the patient underwent diagnostic laparoscopic surgery, during which no mesenteric ischemia was seen. Small amount of ascites fluid was reported as a clear liquid. Due to the absence of ischemia, the surgery was stopped. In the next step, the patient underwent an abdominopelvic CT scan, which showed huge splenomegaly (210 mm) with multiple hypodense and wedge-shaped areas with a maximum size of 38 x 78, indicating a spleen infarct. Abundant collateral vessels were seen in the umbilical cord, suggesting chronic splenic vein thrombosis, and several filling defects were seen in the SMV vein and the beginning of the confluence vein, which appeared to be a subacute process. Other veins reported to be normal. Due to splenomegaly and normal platelet count in the initial lab data, the patient was examined for peripheral blood smear, which showed the white blood cell count as 7000 and there was no leukoerythroblastosis or blast. The cells are normochromic and normocytic and there is no teardrop. The platelet count was 200,000 and no aggregation was seen. So far, the patient underwent bone marrow aspiration and biopsy, which is the result reported to be normal. The patient also underwent genetic testing for BCR-ABL and JAK 2 (F617), which confirmed the genetic mutation that predisposed patient to venous thrombosis and splenic infarction. The patient discharged after improving general condition and advised to take Warfarin 5 mg/day. During the following weeks, the patient was revisited several times, the patient's pain had resolved, she has no sign of any other thrombosis and lab test in her follow up visits reported to be normal. Also, the prior pleural effusion dissolved. After ending the breastfeeding, we changed Warfarin to Rivaroxaban 20 mg/day. In the follow up lab tests, the amount of platelets reported to be 700000.

3. Discussion

In general, thrombocytosis occurs reactively or autonomously [10]. In a reactive way, platelets count rises as an acute phase reactant. This increase in number happens due to rised inflammatory cytokines and has no impacts on the hematopoietic stem cell [11] but is different in the autonomous group. For example, in Essential thrombocytosis (ET), the proliferation of megakaryocytes increases and as a result, the number of platelets produced in the bone marrow and presented in blood increase. The first signs of this disease are venous and arterial thrombosis [10, 12]. However, it is difficult to diagnose definitively which can be done by rolling out other diseases (reactive thrombocytosis, myeloproliferative disorders (polycythemia Vera, chronic myelocytic leukemia, or myelofibrosis)) [13]. The difference between the reactive and autonomous groups is important because in the reactive group the platelet count decreases rapidly by eliminating the underlying cause and the effects of high platelet count on thrombosis or bleeding disappear. But in the autonomous group, there is a need for long-term antiplatelet therapy [14]. Researches has shown that ET is an independent risk factor for thrombosis, and JAK2 V617F gene mutations occur in more than half of patients with ET, making them more susceptible to thrombosis [15]. Given that, the two main risk factors for thrombosis are a previous history of thrombosis and age over 60 year [16, 17]. Studies show that people with thrombosis who are less than 60 years old and suffer from ET are more likely to have a JAK2 mutation [18]. Splenic infarction is very rare among patients with ET and has been reported in several case reports around the world. However, it is important to note that the incidence of ischemia in these patients is not limited to the

spleen and can lead to thrombosis in cerebral, coronary, ocular, and other central and peripheral veins and arteries, which means peripheral gangrene, acute cerebral and cardiac stroke and ocular problems [19].

In our case, there was a splenic infarction, which can be detected in 99% of cases with a liver-spleen scan, and also in 75% cases with abdominal CT scan [20] in reported patient, splenic infarction was diagnosed by abdominal CT scan. Supportive treatment and proper hydration, continuous follow-up, and the use of analgesics should be considered. The symptoms should disappear after 7-14 days. In cases where symptoms do not disappear or complications such as abscesses, cysts and bleeding occur, surgical treatment is preferred, as initial surgical treatment can reduce mortality and morbidity [21-23]. Due to the presence of ET in this patient, the possibility of surgical complications (bleeding and Ischemia and mortality) is much more than other cases, so we tried to manage this patient in a non-surgical treatment approach. We hydrated the patient and prescribed analgesics to reduce pain under close monitoring.

In evidence, the number of people with spleen infarct due to ET is small, as a case report, Keun-Tae Kim MD and colleagues diagnosed a splenic infarction in a 46-year-old man who presented with abdominal pain similar to our case. At the initial time of the diagnosis, he had higher number platelets than our case. The patient reported three vascular lesions during 8 years, splenic vein thrombosis, heart attack and brain stroke, which may be due to the high platelet count in this patient [24]. Of course, the presence of a previous vascular lesion has been shown to be a risk factor for subsequent thrombotic lesions, as in previous researches.

Another study on patients with thrombocytosis under 40 years of age found that the most common cause of thrombocytosis were ET and secondary thrombocytosis. Out of 20 patients with ET, only 2 presented thrombotic events. It shows individuals, as well as all patients whose thrombocytosis background was not secondary to a malignancy, had a good prognosis. This indicates that with proper management of these patients, accidents are rare and the prognosis is appropriate [25].

Another study by Valerio De Stefano et al on patients with ET who also had a mutation in the JAK2 V617F gene, showed that this mutation, especially in its homozygous form, was an independent cause of thrombosis. According to the incidence rate of splenic infarction in some cases, splenic infarction can be a sign of high incidence of other vascular events in other areas, both symptomatic and asymptomatic, which signifies the importance of pharmacologic treatment and reducing other risk factors of vascular accidents [26].

Another study by MING FANG et al in the management of pharmacologic therapy in an aortic thrombosis in a patient with ET stated that, given the evidence of previous studies on the appropriate therapeutic response to surgery and the possible complications of surgery in these patients, The patient was managed with medications. So far, the results were satisfactory and the thrombosis was resolved [27] which is similar to our management in this patient. Other risk factors should be considered too.

Also, a study conducted by Masayuki Oki et al. On a 65-year-old woman with ET who had extensive aortic thrombosis and splenic infarction, found that only medications in this patient resulted in complete cure of the patient's symptoms and removal of the aortic thrombosis. Shrinkage of the infarct area and the spleen itself in the patient, indicates the need for surgery in these patients which is probably less than others [28].

4. Conclusion

In the presented patient, ET was not diagnosed due to the absence of symptoms or complications. It seems that pregnancy became a triggering factor for ET to show up with the symptoms like thrombosis. We may consider this effect to evaluate it more. In addition, there is lack of evidence on about duration of treatment. We don't know when we should stop the anticoagulant therapy or we should continue the medications lifelong. This issue should be considered as the basis of future studies. It is suggested to design and perform clinical trials or doing systematic reviews in order to answer this question: "How long should we continue the anticoagulant therapy?".

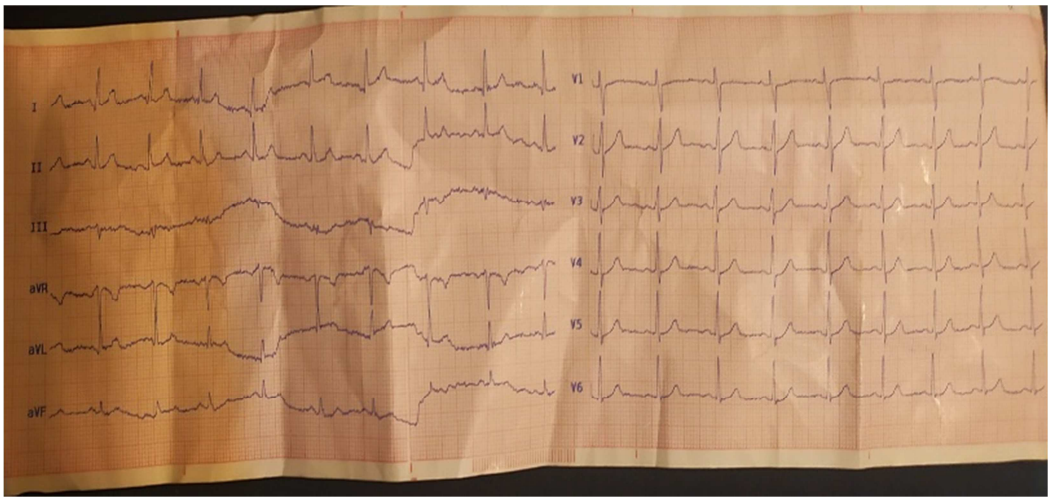

Figure 1. *The ECG of the Patient.*

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