Cesarean Section Combined with Splenectomy for Refractory Immune Thrombocytopenic Purpura

Wen-Jui Lee, Chien-Nan Lee, Woei Tsay, Fon-Jou Hsieh, Song-Nan Chow*

Department of Obstetrics and Gynecology, and 1Department of Internal Medicine, National Taiwan University Hospital and College of Medicine, National Taiwan University, Taipei, Taiwan.

*Correspondence to: Dr. Song-Nan Chow, Department of Obstetrics and Gynecology, National Taiwan University Hospital, No. 7, Chung-Shan South Road, Taipei 10020, Taiwan. E-mail: snchow@ha.mc.ntu.edu.tw

Received: June 16, 2003
Revised: June 16, 2003
Accepted: June 16, 2003

SUMMARY

Objective: The major treatment options for maternal immune thrombocytopenic purpura (ITP) are corticosteroids or intravenous immunoglobulin. If the patient fails to respond to medical treatment, splenectomy should be considered.

Case Report: A 27-year-old pregnant woman, gravida 2, para 1, was referred to our hospital because of ITP at 35 weeks' gestation. Steroid therapy had been prescribed (75 mg/day prednisolone) since 31 weeks' gestation, but no response was observed. The patient was admitted to our ward due to premature rupture of membranes at 36 weeks' gestation, and cesarean section was performed as she had had a previous cesarean section. After delivery of the infant, splenectomy was performed as definitive therapy for her ITP. The patient's initial post-operative course was uneventful; the platelet count on the follow-up visit 6 weeks later was 109,000 cells/µL.

Conclusions: ITP should be managed with steroid therapy or intravenous immunoglobulin. When these fail, splenectomy may be the only remaining option. [Taiwanese J Obstet Gynecol 2004;43(1):35-37]

Key Words: autoimmune disorder, cesarean section, immune thrombocytopenic purpura, splenectomy

Introduction

Immune thrombocytopenic purpura (ITP) is an autoimmune disorder resulting in IgG-mediated accelerated platelet clearance. The incidence is 1 to 2 per 10,000 pregnancies [1]. In the pregnant patient, the antibodies cross the placenta, with the potential to cause profound neonatal thrombocytopenia. However, the fetal or neonatal platelet count cannot be reliably predicted from the maternal platelet count, maternal platelet antibody level, or a history of maternal splenectomy for ITP [2–5]. Data from several reports indicate that 10% of infants born to women with ITP are born with platelet counts below 50,000 cells/µL.

Treatment of ITP depends on the severity of thrombocytopenia, drug response, and patient compliance.

The major treatment options for maternal ITP are corticosteroids or intravenous immunoglobulin (IVIg) [6]. Splenectomy is usually reserved as second line therapy and only performed if there is failure to respond to an adequate trial of medical management or an inability to tolerate steroid treatment [7]. We report the case of a patient with ITP who failed to respond to steroid therapy and who had a cesarean delivery coupled with splenectomy as definitive therapy for her ITP.

Case Report

A 27-year-old woman, gravida 2, para 1, was referred to our hospital because of ITP at 35 weeks' gestation. During pregnancy, at 12 weeks' gestation, she had experienced bleeding gums from brushing her teeth. Platelet counts at 19 weeks and 23 weeks of gestation were 53,000 cells/µL and 45,000 cells/µL, respectively. At 31 weeks of gestation, steroid therapy (75 mg prednisolone/day) was prescribed. Despite this treatment, her platelet count decreased to 26,000 cells/µL at 34 weeks' gestation. The platelet count at referral, at 35 weeks' gestation, was 20,000 cells/µL. Her blood
pressure was normal. Laboratory results included a negative antibody test against human immunodeficiency virus, a negative antinuclear antibody test, normal liver function studies and electrolytes. No sign of complement activation was seen. ITP was suspected, and, because of the limited response to steroid therapy, splenectomy was suggested by the hematologist.

The patient was admitted to our ward due to premature rupture of membranes at 36 weeks' gestation. At the time of admission, her platelet count was 17,000 cells/µL. Hemoglobin concentration was 11.6 g/dL, and prothrombin time and activated partial thromboplastin time were normal. Determination of the fetal platelet count was unnecessary because cesarean delivery was planned due to previous cesarean section. Transfusion of 12 units of platelets was performed soon after admission.

Under general anesthesia, a low segment transverse cesarean section was performed. A female infant weighing 3,460 g was delivered. The infant had Apgar scores of 8 and 9 at 1 minute and 5 minutes, respectively. After delivery of the infant, a splenectomy was performed on the mother through a midline abdominal incision, and another 12 units of platelets were transfused during the surgery. A rubber drainage tube was placed in the subphrenic space. There were no intraoperative complications. Estimated blood loss was 1,100 mL. Maternal platelet count was 110,000 cells/µL 6 hours after surgery. Transfusion of six units of platelets was performed on postoperative days (PODs) 1 and 2. Maternal platelet count was 101,000 cells/µL on POD 4. The drainage tube was removed on POD 6, and she was discharged from our hospital on POD 7. The dose of prednisolone was gradually reduced and was discontinued 19 days after the surgery. Medical treatment with azathioprine 50 mg/day was started at the same time. Six weeks after surgery, the platelet count was 109,000 cells/µL.

After delivery, petechiae over the infant’s trunk, extremities, buttocks, and inguinal area were seen, but the infant’s platelet count was 245,000 cells/µL. The infant’s initial post-delivery course was uneventful, and no specific treatment was necessary. The petechiae disappeared without treatment 2 days later. The infant was discharged on day 8.

Discussion

ITP is an autoimmune disorder characterized by a low platelet count and mucocutaneous bleeding. The decreased platelet count is due to autoantibodies directed against target antigens on platelets, especially the glycoprotein IIb-IIIa complex on glycoprotein Ib. The antibodies serve as opsonins, accelerating platelet clearance by phagocytic cells in the reticuloendothelial system. Thrombocytopenia, defined as a platelet count below 100,000 cells/µL, is diagnosed frequently during pregnancy, affecting up to 8% of pregnant women. Gestational thrombocytopenia accounts for 75% of thrombocytopenia cases at term [8–10]. The platelet count of a patient with gestational thrombocytopenia is rarely below 70,000 cells/µL, and returns to within a normal range within 2 months after delivery [11]. ITP is present in 0.01–0.02% of women at the time of delivery [12]. ITP and gestational thrombocytopenia are diagnoses of exclusion and difficult to differentiate. The differential diagnosis of severe thrombocytopenia during pregnancy includes systemic lupus erythematosus, HIV infection, antiphospholipid antibodies, and HELLP syndrome (hemolysis, elevated liver enzymes, low platelet count). All of these conditions were ruled out in this patient.

Treatment is reserved for patients with platelet counts below 10,000 cells/µL or counts of 10,000–30,000 cells/µL with evidence of impaired hemostasis, or in preparation for delivery or invasive procedures [13]. The first line of therapy is prednisolone, at a dose of 1 mg/kg body weight per day. Those who fail to respond are treated with IVIg (0.4 g/kg/day for 5 days or 1 g/kg/day for 2 days), which typically results in improvement lasting several weeks [6,11]. If the patient fails to respond to either drug or their combinations, splenectomy may be recommended as second line therapy. In our patient, steroid therapy was initiated, but with no response. The extremely high cost of IVIg forced consideration of a more definitive therapy such as splenectomy. Splenectomy is the treatment of choice in the non-pregnant patient. Traditionally, splenectomy during pregnancy has been associated with high fetal and maternal mortality [14]. Therefore, splenectomy was performed after the cesarean section in our patient.

Maternal platelet counts do not accurately reflect fetal platelet counts [9]. Although 10% of neonates born to women with ITP are born with platelet counts below 50,000 cells/µL, only 4% are born with platelet counts below 20,000 cells/µL. The risk of intracranial hemorrhage (ICH) or other major bleeding complication is less than 1% [15]. There is no evidence that treating maternal thrombocytopenia will improve fetal platelet count. Routine fetal scalp monitoring or percutaneous umbilical vein blood sampling are not currently recommended due to inaccuracy of the fetal platelet counts and the potential morbidity of the procedures [13]. There are no studies showing that the risk of ICH is reduced by cesarean section [16]. As the frequency of severe hemorrhagic complications in the newborn is

...
low, it is recommended that the mode of delivery be based on obstetric indications.

Thrombocytopenia also alters the selection of anesthesia. Many anesthesiologists suggest the avoidance of regional anesthesia in patients with a platelet count of less than 100,000 cells/µL [17]. Patients with ITP are generally refractory to platelet transfusion due to the presence of platelet antibodies, and, therefore, platelet transfusion is not indicated unless there is severe and life-threatening bleeding, or before splenectomy or cesarean section. A platelet count of more than 50,000 cells/µL should be the target of transfusion, and that target can usually be achieved with six units. Unnecessary transfusion of platelet concentrates in the absence of hemostatic failure may stimulate more autoantibodies and worsen maternal thrombocytopenia [7].

In conclusion, treatment for ITP is usually not recommended if the platelet count is more than 50,000 cells/µL. In patients with platelet counts below 10,000 cells/µL, or counts between 10,000–30,000 cells/µL with evidence of impaired hemostasis, or in preparation for delivery or invasive procedures, the initial treatment of choice is steroids, followed by IVIg. Splenectomy may be the only remaining option when medical treatment has failed.

References