Splenectomy during cesarean section in a patient with severe idiopathic thrombocytopenic purpura (ITP) refractory to the medical treatment

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Idiopathic thrombocytopenic purpura (ITP) is a disease of women of reproductive age and accounts for 5% of pregnancy associated thrombocytopenia. We reported the case of 21-year-old primigravida with ITP who was admitted to our hospital at the 28th weeks of gestation with platelet count of 22x10^9/l. Despite medicament treatment (prednisone, intravenous immunoglobulin /IgG/) there were falls in the platelet count, last at the 33rd week of gestation when platelet count was 15x10^9/l. In consultation with hematologist and surgeon we decided to deliver the patient by cesarean section during which splenectomy will be done. Patient received 60g of intravenous IgG two days before operations which were performed at the 34th week of gestation. Healthy female infant was born with normal platelet count and maternal platelet count at the discharge from hospital was 346x10^9/l.

Key words: idiopathic thromboticopenic purpura /ITP/, pregnancy, splenectomy, caesarean section

INTRODUCTION

Idiopathic thrombocytopenic purpura (ITP) commonly affects women of reproductive age and its incidence in general population is 1-5/10000. In contrast to the more common gestational thrombocytopenia ITP occurs in only 0.1 to 0.2% of all pregnancies and constitutes only 3% of all cases of thrombocytopenia during pregnancy 3,5. In pregnancy medical intervention should be considered whenever the platelet count is between 20-30x10^9/l, or in symptomatic patients with platelet count between 20-50x10^9/l 4. Before delivery platelet count should be maintained above 50x10^9/l which is regarded as safe for vaginal delivery or cesarean section4. Prednisone, 20-30 mg/per day is the conventional initial treatment. Those women with severe form of ITP should be treated with higher dose, and 1mg/kg/day is recommended 5. When symptoms of ITP persists after primary treatment with glucocorticoids the second line of therapy includes intravenous immunoglobulin. Rarely, when medical management fails, in patients with severe ITP defined as platelet count of less than 20x10^9/l, splenectomy is the only definitive treatment3,6,1.

CASE REPORT

A 21-year-old primigravida was admitted to our hospital at the 28th weeks of gestation complicated with idiopathic thrombocytopenic purpura (ITP) with platelet count of 22x10^9/l. This was first hospitalization in our institution, patient was diagnosed with ITP at the 22nd week of gestation in another University hospital where she had puerperal hospitalizations. Platelet count at the admission in this institution was 15x10^9/l, and she was treated with oral prednisone 40mg/daily, and intravenous IgG 25g /400mg/kg/ daily for ten days. Last platelet count at the discharge was 115x10^9/l. In consultation with hematologist we increased prednisone dosage at 80mg/daily, and on the second day she received intravenous IgG 25g /400mg/kg/ for three days. Platelet count on the forth day after admission was 36x10^9/l. Fetal ultrasound revealed normal fetal growth with adequate feto-placental and cerebral circulation. On the fifth day after admission patient developed many petechia and minor purpura on the limbs which did not disappear until childbirth.

Between 30th and 31st weeks of gestation platelet count was in the range of 43-57x10^9/l and prednisone dosage was 60mg/daily (3x20mg), and platelet count until 33rd week of gestation remained stable in the range between 44 and 33x10^9/l. On the 33rd week of gestation there was another fall in platelet count which were than 15x10^9/l, and despite intravenous IgG count were low in next seven days in the range between 28 and 25x10^9/l.
Since there were no improvement in platelet count despite the fact that we applied both modalities of medication therapy, after written informed consent had been obtained, we decided to deliver the patient by cesarean section during which splenectomy will be done. Patient received 60g of intravenous IgG two days before operations which were performed at the 34th week of gestation. Platelet count at the day of operation was 53x10^9/l so there was no need for platelet transfusions. Healthy female infant weighing 2300g was born with Apgar score 8 and 9 at 1st and 5th minute. Baby's platelet count was normal, and baby went home at 3 weeks later. Patient's parameters of the coagulation system were within normal limits, and platelet count day after operations was 141x10^9/l. Maternal platelet count at the discharge from hospital was 346x10^9/l, and 4 weeks after operation 200x10^9/l.

**DISCUSSION**

Idiopathic thrombocytopenic purpura is one of the well studied hematologic disease in which medical as well as surgical treatment strategies have been proposed. Although the role of spleen in ITP has not clearly been identified the benefits of splenectomy in these patients may be the result of a combined effect of elimination of the major source of antiplatelet antibodies production and platelet destruction. The efficacy of glucocorticoid therapy usually does not exceed a complete remission rate of 60%, in case of drug discontinuation the remission rate may decrease to 30%. Splenectomy offers a more definitive approach to the therapy of ITP and should be considered in patients with severe form of ITP (defined as platelet count of less than 20 x 10^9/l), who fail to respond to the medical treatment. Response rate up to 90% following splenectomy are reported.

Because of trans-placental passage of maternal IgG, particularly during the third trimester, fetal thrombocytopenia may also occur. Numerous studies over the past two decades have documented that neonatal thrombocytopenia is both uncommon and unpredictable. Currently there is no proven maternal treatment that could decrease the incidence of fetal thrombocytopenia. Moreover, because of the potential increased risk of neonatal intracranial hemorrhage, the route of delivery is still debated. In the later years cesarean section was recommended for all patients with ITP based on the reported perinatal mortality due to birth trauma and intra-cerebral hemorrhage. However, more recent studies suggest a neonatal mortality of around 0.6%, and there is no evidence that cesarean section is safer for the thrombocytopenic neonate than uncomplicated vaginal delivery.

In our case, the fact that we had patient with severe form of ITP that was refractory to the first and second line of medicament treatment necessitates splenectomy as a definitive treatment option. Delivery was planned when the fetal maturity was achieved, and since conditions were unfavorable for vaginal delivery, cesarean section was done during which we performed splenectomy.

In conclusion, splenectomy during cesarean section is highly effective and safe modality of treatment in the patients with severe form of ITP refractory to the medicament forms of therapy.

**SUMMARY**

Idiopatska trombocitopenijska purpura (ITP) predstavlja oboljenje žena u reproduttovnom periodu i čini oko 5% trombocitopenija u trudnoći.

Prikazali smo slučaj prvorođene stare 21 godinu koja je primljena u našu bolnicu u 28. nedelji gestacije sa brojem trombocita na prijemu 22x10^9/L. I pored medikamentozne terapije (pronizon, intravenski imunoglobulini /IgG/) i dalje su bili prisutni padovi u broju trombocita, poslednji u 33. gestacijskoj nedelji kada je zabeležena trombocitopenija od 15x10^9/L.

U konsultaciji sa hematologom i hirurgom odlučili smo da pacijentkinju porodimo carskim rezom u toku koga će biti uradjena i splenektomija. Pacijentkinja je primila 60 g intravenskih imunoglobulina /IgG/ tokom dva dana pre operacije koja je uradjena u 34. gestacijskoj nedelji. Rezeno je živo žensko dete sa normalnim brojem trombocita, a pacijentkinja je otpuštena iz bolnice sa urednim brojem trombocita / 346x10^9/L/.

**KLJUČNE REČI:** idiopatska trombocitopenijska purpura /ITP/, trudnoća, splenektomija, carski rez

**LITERATURE**


