

Guillain-Barre Syndrome in Early Pregnancy: Case Report

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Abstract

Guillain-Barre Syndrome is an acute immunodeficient polyradiculoneuropathy of the peripheral nervous system, seen in 1-2 cases in 100,000. Motor involvement is predominant in the peripheral nervous system. Pregnancy does not change the incidence of Guillain Barre Syndrome, but pregnant Guillain Barre cases have a great importance because of their vulnerability to complications. A 23-year-old female patient at the 6th week of pregnancy presented to emergency department with sensory loss and weakness in both lower limbs. The prognosis of Guillain-Barré syndrome diagnosed in early gestational weeks was discussed with literature by presenting a case that resulted in death, despite of plasmapheresis treatment.

Keywords: Pregnancy; Guillain-Barre Syndrome; Plasmapheresis; Mortality; Emergency Medicine

Introduction

Guillain-Barre Syndrome is an acute immune-mediated polyradiculoneuropathy of the peripheral nervous system [1]. The incidence of Guillain-Barre Syndrome in the general population is around 1 to 2 in 100.000 per year. It is the most common cause of acute flaccid paralysis. Clinical findings appear approximately four weeks after the onset of the disease. The incidence of Guillain-Barre syndrome in pregnancy has been reported at the rate of 1.2-1.9/1.000.000 [2]. Conditions related to pregnancy, except preeclampsia, do not increase the risk of Guillain-Barre syndrome [3]. Although the exact etiology of Guillain-Barre Syndrome is unknown, 30% of the cases are associated with C.Jejuni [4]. Also there are some studies show that Zika virus might be related to Guillain Barre Syndrome [5,6].

A 23-year-old female patient at the 6th week of pregnancy was admitted to the emergency department with sensory loss and weakness in both lower limbs. Our case resulted in death despite plasmapheresis treatment. The prognosis of Guillain-Barré syndrome in early gestational weeks and the effect of disease on the pregnancy were discussed with literature.

Case

A 23-year-old female patient at the 6th week of pregnancy was admitted to the emergency department with complaints of ascendant sensory loss and weakness. Her medical and surgical histories were unremarkable. The patient had no known infection history in the past two months.

On physical examination the general condition was considered as moderate. She was conscious, oriented, but had limited cooperation. Vital Findings were; blood pressure:100/70 mmHg, pulse: 110/min, respiratory rate: 18/min, body temperature: 37 °C, sPO2: 96%.

Glasgow coma scale was 15, and the muscle strength was 3/5 at distal and proximal muscles of the upper extremities and 2/5 at distal and proximal muscles of the lower extremities. Deep tendon reflexes of the lower extremity were absent, and left biceps reflex was hypoactive while triceps reflex was absent. The patient had no cooperation for sensory examination. The oropharynx was natural and the gag reflex was normal. On cardiac examination S1 + and S2 + were rhythmic with no additional sound or murmur. Both hemithoraces were equally involved in respiration, there was no rale or rhonchi. She was tender in the suprapubic region and evaluated as having glob vesicale. The patient's urination was limited. The anal sphincters were intact.

Laboratory results: white blood cell count: 13.010/microliter, hemoglobin: 15 grams/deciliter, platelet 374.000/microlitres, C-reactive protein: 2.31 micrograms/deciliter, alanin aminotransferase: 54 international units/liter, asparate aminotransferase: 42 units/liter,

gamma glutamyl transferase: 72 units/liter. Coagulation tests, electrolytes and kidney function was normal. Cranial diffusion magnetic resonance imaging revealed no pathology. Electromyography revealed that sensory and motor fibers were severely affected, and these findings were consistent with acute polyneuropathy. One erythrocyte was seen in the lumbar puncture. Cerebrospinal fluid biochemistry results were in normal ranges except for the elevation of the chlorine value. There was no microbial proliferation in the cerebrospinal fluid culture. The patient was interned with the diagnosis of Guillain Barre Syndrome.

The patient was treated with 6 cycles of plasmapheresis, and Enoxaparin sodium 0.6cc (Clexane; Sanofi Aventis, İstanbul, Turkey) subcutaneously 2 times a day. On the second day of the hospitalization, she was intubated due to developing respiratory distress association with Guillain-Barre Syndrome. We used rocuronium 50 mg (Myocron;VEM, İstanbul, Turkey) and Propofol 50 mg (Propofol-Lipuro %120 ml; B. Braun, Philippines) when intubated. On follow-up, she had fever of 38 °C, and ceftriaxone 1x2 grams a day added to the therapy. On the 10th day, antibioticotherapy was changed to linezolid 600 mg 2 times a day. On the 11th day of the hospitalization, the patient had spotting vaginal bleeding. On the 15th day of admission the patient had missed abortus. Despite of completing the plasmapheresis, the patient died on the 16th day of admission due to acute respiratory distress syndrome secondary to ventilator-associated pneumonia.

Discussion

Guillain-Barre Syndrome can be seen in all ages, mostly in young adults and elderly populations. Guillain-Barre Syndrome can occur in any trimester of pregnancy. In particular, the occurrence increases in the third trimester and postpartum second week. The reason of that seems to be the increasing of the delayed type hypersensitivity reactions and the restoration of the cellular immunity [7]. This is important because of the worsening of the disease in postpartum period [6].

In the study conducted by Auger, *et al.*, it is noteworthy that immunological related diseases increase the risk of illness by 6 times, rheumatic diseases by 7 times, transfusion by 3 times, preeclampsia by 2 times, but there is no data showing that pregnancy increases the risk [3]. 30-35% of cases with Guillain-Barre Syndrome seen in early gestational weeks are CMV positive [8,9]. The etiology of our case is unknown. In the literature, maternal mortality due to Guillain-Barre Syndrome is reported to be 10% and the percentage of mechanical ventilation requirement is above 35% [10]. In the literature, between 1986 and 2018, 7 cases including our case were diagnosed in the first trimester. However, the only case that results in maternal death is our case (Table 1). In a study conducted by Chan Tsui L. and colleagues between 1986 and 2003, 30 pregnant women diagnosed with Guillain-Barre Syndrome were evaluated and only 1 of them died. On the 39th day of the mother's discharge she was found dead at home [11]. Premature termination of pregnancy in cases with rapid and aggressive Guillain-Barre Syndrome should be considered because of the increased risk of complications of the disease in pregnancy. Also in the study conducted by Kemesha Delisser and Joyce Ho, hyperimmune response caused to fetal demise [12]. Early recognition of the disease and early intravenous immune globulin (IVIG) treatment are important for positive outcomes in the survival of the mother. IVIG rapidly neutralizes pathogenic antibodies, resulting in reduced nerve damage and faster clinical recovery [13]. There is no superiority in treatment between plasmapheresis and IVIG in terms of 4-week survival, improvement in disability, duration of mechanical ventilation, and residual disability. In addition, the use of one of the treatments is recommended because the combined use of plasmapheresis and IVIG does not provide additional benefit [14]. Plasmapheresis is effective in the first 2 weeks after the onset of disease in patients who are unable to walk, and it is most effective in the first 7 days after the onset of weakness in the limbs.

Seven cases between 1986 and 2018 were diagnosed with Guillain-Barre Syndrome in the first trimester; Plasmapheresis and IVIG were used together in one of these cases, only plasmapheresis were used in two cases, and only IVIG was used in one case (Table 1). In our case, only plasmapheresis was used. Two treatments can be used together in patients who are not responsive to IVIG or plasmapheresis treatment alone. In one case, a combination of steroid and plasmapheresis was applied [15]. In another patient, IVIG was administered after steroids but no positive effects on pregnancy were detected.

The mean age of cases with Guillain-Barre Syndrome in the early gestational week is 26.7 (minimum 19 years, maximum: 38 years). The age of our case is below the average age in this group. Only one of the cases was able to deliver a healthy birth and 6 pregnancies resulted in intrauterine death or termination of pregnancy. Only our case has resulted in maternal death due to increased respiratory distress. In the studies conducted, the mortality rate of Guillain-Barre syndrome cases ranged from 0% to 13% with an average of 6% [16]. In literature, the mortality rate was found to be 3.33% in patients diagnosed during pregnancy [11]. Guillain-Barre Syndrome in pregnancy has presented as good prognostic outcome in the literature but early diagnosis is important both in terms of mother and baby survival and in terms of morbidity. Pregnancy does not increase the risk of Guillain-Barre Syndrome, but it is important to keep in mind that it increases the vulnerability to disease's complications [11].

Case	Age	Gestation (weeks)	Pregnancy outcome	Maternal outcome	Treatment	Etiology
Zeeman GG (2001)	34	4	Termination of pregnancy	No deficit 2 months later	Steroid and plasmapheresis	-
Nesbitt I, <i>et al.</i> (2000)	19	6	Termination of pregnancy at 9 weeks (due to severe illness and CMV infection)	Discharged from Intensive Care Unit after 86 days, slow recovery afterwards	IVIG	CMV

Brooks H, et al. (2000)	28	7	Cesarean section for genital warts at 38 weeks	Weaned from ventilator 18 weeks after onset	IVIG, then plasmapheresis	-
Mendizabal JE, et al. (1997)	15	10	Missed abortion at 20 weeks	Ambulatory 9 days after admission	Plasmapheresis	CMV
Lupo Julien, et al. (2016)	30	10	Intrauterin death at 25 weeks (due to intrauterin CMV)	4 weeks later, at the time of discharge from hospital, the patient was still unable to walk independently, requiring outpatient rehabilitation.	Steroid,then IVIG	CMV
Kokubun Norito, et al. (2013)	38	6	Missed abortion	Discharged from the hospital on 16 th day,had no sequale on day 59.	The patient refused plasmapheresis treatment.	-
7	23	6	Abortion 2 days before patient died	Exitus on day 16	Plasmapheresis	-

IVIG: intravenous immune globulin; CMV: cytomegalovirus

Table 1: Cases with Guillain-Barre Syndrome diagnosed in the first trimester of gestation published in literature between 1986-2018

Conclusion

In pregnancy, Guillain-Barre Syndrome may present with pain, numbness, reduced or lost sensation before dramatic clinical presentations. Infectious processes have to be questioned (especially upper respiratory tract and gastroenteritis for our country), detailed history should be taken, travels to regions where the incidence of zikavirus and cytomegalovirus transmission are high should be questioned and organic pathologies should be excluded before anxiety is diagnosed in pregnant patients. Early termination of pregnancy in aggressive progressive Guillain-Barre Syndrome in early gestational weeks should be kept in mind.

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