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Case Report

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Uncommon case of guillain-barré syndrome in young female in the first trimester of pregnancy

Abstract

Background: Guillain-Barré syndrome (GBS) is an autoimmune disorder with the estimated incidence between 1.2 and 1.9 cases per 100 000 people annually. It is rare in pregnancy as there is a decrease in the cell-mediated immunity, but it is generally accepted that it carries a high maternal risk.

Aim: In this study, a case of an 18-year-old pregnant woman suffering from GBS who presented with flaccid quadriplegia is discussed in the aspects of diagnosis, treatment, and outcome.

Conclusion: Rapid recognition of GBS as well as resolute and prompt treatment consisting of plasma exchange and intravenous immunoglobulins (IVIG) are necessary to get a good outcome in pregnant women such as the patient discussed.

Keywords: Guillain-Barré syndrome, Immunoglobulin, Plasmapheresis, Pregnancy

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Abbreviations: GBS, Guillain-Barré Syndrome; CSF, Cerebro Spinal Fluid; PT, Prothrombin Time, PTI, Pro Thrombin Index, APTT, Activated Partial Thromboplastin Time; CBC, Complete Blood Count; AIDP, Acute Inflammatory Demyelinating Polyeuropathy

Introduction

Guillain-Barré Syndrome (GBS) is an acute monophasic type of inflammatory demyelinating polyradiculoneuropathy usually associated with symmetrical progressive muscle weakness, areflexia or weakness of deep tendon reflexes, and albumin cytological dissociation (a raised protein level but normal cell count) in the cerebrospinal fluid (CSF). Extra-ocular movement abnormalities, facial weakness, ataxia, respiratory failure, and autonomic dysfunction may also occur. Diagnosis is made based on clinical history, the presence of cytoalbuminologic dissociation in CSF, electro diagnostic investigation results supporting acute neuropathy, and the exclusion of mimic diseases [1,2] Nearly all these cases, however, occurred in the second or third trimester or the first month postpartum. Delayed diagnosis is common in pregnancy or immediate postpartum period because the initial symptoms can be non-specific [3] GBS treatment in pregnancy is either a course of plasma exchange or intravenous immunoglobulins, just as in the rest of general population.4 The relation between GBS and pregnancy or delivery is of special interest due to dramatic GBS symptoms in women facing delivery. The case report of GBS in a pregnant woman in the early stage of pregnancy who presented with unusually dramatic and longstanding course will add to the very limited literature on this subject.

Case report

An 18 years old female gravida I was referred to the Department of Neurology in our hospital in week 9 of pregnancy with a one week history of progressive weakness in the lower limbs and difficulty in walking, which, for the last 3 days, culminated in the loss of ability to walk. The symptoms also included pain in the lumbar area radiating to both the lower limbs, which was preceded by 2 weeks by a flu-like illness. On examination, the patient was found acutely ill, pale, febrile, acyanosed, and anicteric with satisfactory hydration status; her breathing was insufficient, and a cardiovascular examination showed no abnormality. On neurological examination, she was found conscious but restless and apprehensive. All her cranial nerves were found normal. She presented moderate flaccid paraparesis in both the lower limbs, but she was unable to walk. The strength in the upper limbs was normal. Muscle tone was decreased and deep tendon reflexes were lost in lower limbs. There was neither sensory impairment, nor bladder or bowel involvement. Cerebrospinal fluid was abnormal with high protein level (2.2g/l) and normal content of cells (0/ml). The next day after the admission to the ward the patient presented flaccid quadriparesis. The weakness increased the following day and the patient had weak cough and gag reflexes. She was also dyspneic with shallow breathing.

A nerve conduction study showed demyelization and axonal injury in peripheral nerves and nerve roots more distinct in lower extremities. There were prolonged distal latencies in both the peroneal nerves (8.3 and 8.7 ms) and in the left tibial nerve (8.7ms), a decrease of M amplitude response in motor fibres of the perineal nerves (0.3), the left tibial nerve (0.9), and the left ulnar nerve (2.9), conduction block in both the perineal nerves and the left tibial nerve, an F-wave absence in upper and lower limbs nerves, a decrease of sensory response amplitudes in the left ulnar nerve. The clinical state and the nerve conduction study as well as the cerebrospinal fluid analysis suggested GBS. The plasma exchange was started (2000 ml) and plasma was used as a replacement fluid. The anaphylactic shock appeared directly after the procedure was completed, so the patient received hydrocortisone intravenously. These symptoms' intensity was increasing, and so, due to the complications, the patient was given immunoglobulin intravenously (IVIG) (0.4g/kg) for 7 days.

The patient's state deteriorated; she was not able to maintain spontaneous breathing and she could neither walk nor swallow. On the 6th day the patient was intubated and treated in the Department of Anaesthesiology and Intensive Treatment in Regional Hospital

in Poznań not Anaesthesiology and Critical Care Department of the Medical University in Poznan where therapeutic plasma exchange was maintained at the rate of 2000 ml +/- 182 ml of plasma per session. This was in accordance with the guidelines of removing 1-1.5 of plasma in each session. 5% albumin was used as a replacement fluid. The peripheral femoral access was used to carry out the procedure. Proper catheter care was maintained, and lines were flushed with heparin before and after the exchange. No complications occurred during the sessions. The following tests were carried out before and after each procedure: a complete coagulation profile including prothrombin time (PT), prothrombin index (PTI), activated partial thromboplastin time (APTT); complete blood count (CBC) including haemoglobin, platelet count; electrolytes levels (Na+, K+, Ca++); and protein levels. The results remained within normal range throughout the treatment period. Physiotherapy was continued. The patient was discharged from the intensive care unit on day 60 of illness. At the time of discharge, the power in the upper limbs was 3/5 and 2/5 in the lower limbs, and the patient could swallow independently. The patient was transferred to the rehabilitation ward, where she aged for 4 months. Then she was admitted to gynecology where the healthy baby was born by cesarean section in general anesthesia at 37 weeks of gestation. After delivery, the Mather with baby resided in a hospice for social reasons, where proper and comprehensive care, moral support, and psychological counselling were provided. The patient was discharged home after 6 months of the hospice stay with her limbs power assessed 5/5 in the upper and 4/5 in the lower limbs. After 2 years, the follow-up showed no residual weakness, but nerve conduction disturbances have not yet normalization. The baby develops normally.

Discussion

GBS is an autoimmune disorder which is rare in pregnancy as there is a decrease in cell-mediated immunity, which, on the other hand, increases maternal mortality [5-7] Nearly all cases occurred in the second or third trimester or in the first month postpartum and, predominantly, took the form of acute inflammatory Demyelinating polyeuropathy (AIDP), where weakness started in the lower limbs (8). The occurrence of the disorder is very rare in the first trimester of pregnancy, and it also rarely takes the chronic form. Sharma et al. [5] analyzed GBS cases in 47 pregnant women only one of whom was in her first trimester and only one case turned out to be chronic [8] The patient discussed in this paper presented the symptoms in week 9 of pregnancy; moreover, the case turned out to be both chronic and extremely severe. Treatment of GBS during pregnancy is similar to that of non pregnant individuals. Plasmapheresis or IVIG 1g/kg per day for 2 days or 0.4g/kg per day for 5 consecutive days.[4,8-11] is typically used. Close to 10% of patients may deteriorate although the proper treatment, even after an initial phase of improvement. Retreatment over 5 days is recommended in these patients [12] For these patients belonged our pregnant patient in who only after a longer treatment has been an improvement. High dosage IVIG did not have any harmful impact on the course of pregnancy, which supports the other casuistic reports [13-15] This aggressive treatment was safe for patient as well as for the baby.

The failure of the first plasmapheresis was caused by the use of plasma instead of albumin. The case discussed proves that it is more advisable to use albumins than plasma, as the latter results in allergic reaction more often [4,5] which was also observed in the patient discussed in the paper. After albumins were implemented no allergic reaction was observed. The adduced review of a 47 pregnant GBS patients found no treatment related complications with either therapy [8] The patient had Caesarean section because at the time of delivery there was still some muscle weakness. General anesthesia was well tolerated both by the patient and the baby, in accordance with Nesbitt .[16] It is worth stressing, that if muscle strength in a GBS pregnant patient and her general status is satisfactory, the baby can be delivered vaginally. [4,8,13,14]

Conclusion

To conclude early diagnosis and prompt intensive multidisciplinary supportive care - therapy is advantageous in a GBS-complicated early pregnancy, as well as essential for a favorable outcome for both mother and fetus [17] Sometimes the treatment should continue for a long time. Every case of limb paresis in pregnant women should be considered as a possible GBS case, even in the early stage of pregnancy.

Acknowledgments

None.

Conflicts of interest

None.

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