CASE REPORT

Successful Weaning After Plasmapheresis Performed in a Patient with Myasthenic Crisis Provoked by Listeria Meningoencephalitis

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Abstract

We report the development of acute respiratory failure in a myasthenic patient who was on immunosuppressive treatment. The patient was hospitalized due to myasthenic crisis provoked by listeria meningoencephalitis. He had to be intubated and mechanical ventilation was started due to acute hypercapnic respiratory failure. The patient could not be weaned from the ventilator despite intravenous immunoglobulin treatment and treatment for listeria infection. After 8 courses of plasmapheresis, he recovered and was successfully weaned from the ventilator.

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Key words: *Myasthenia gravis, myasthenic crisis, listeriosis, listeria meningoencephalitis, respiratory failure, weaning.*

Introduction

Infection due to *Listeria monocytogenes* mostly effects immunocompromized hosts (1,2). Although, viruses usually cause encephalitis and bacterial infections are associated with meningeal inflammation, *Listeria monocytogenes* is one of the few bacterial microorganisms that may present as a meningoencephalitis and most frequently as a rhombencephalitis (1,2). Listeria rhombencephalitis causes respiratory failure in 41% of the cases, most probably due to the involvement of autonomic respiratory centers (1,2). Here we report a case of listeria meningoencephalitis without brainstem involvement provoking myasthenic crisis, acute hypercapnic respiratory failure and difficulty in weaning from the mechanical ventilation in a patient with myasthenia gravis. To the best of our knowledge, two cases of listeria meningitis in myasthenia gravis were reported previously (3), and this is the first report of listeria meningoencephalitis in a myasthenic patient.

Case Report

A 34-year-old man with myasthenia gravis was admitted to our Medical Intensive Care Unit because of a fever, headache, nausea, vomiting, confusion and left hemiparesis

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over a 3-day period. Myasthenia gravis was diagnosed when he was 18, for which thymectomy was performed in the same year. Since then, he has been using prednisone (10-60 mg qd adjusted according to the severity of his symptoms). Nine months ago, azathioprine (50 mg qd) was added to the treatment because of bulbar symptoms involving dysphagia, dysphonia and shortness of breath. One month prior to this admission, he was hospitalized in another center due to fever and diarrhea and treated with ciprofloxacin for 10 days with the probable diagnosis of enteric fever. However, one week after being discharged from that hospital, fever had recurred and his present symptoms developed.

On admission to our center, his physical examination revealed a temperature of 39°C, confusion, neck stiffness with other meningeal irritation signs. On his neurologic examination, ocular movements were found to be restricted to all directions but other cranial nerve functions were normal. Additionally, left hemiparesis with positive babinski sign was present. Cranial computed tomography was unremarkable. The cerebrospinal fluid (CSF) examination revealed an opening pressure of 300 mm-H_2O, a protein concentration of 18 g/L and a glucose level of 7 mg/dL. Microscopic examination showed 400 x 103/mL white blood cells, 50% of which were neutrophils. Electroencephalograhy demonstrated slowing of the background activity, prominently on the right side. Azathioprine was discontinued and prednisone (100 mg/d) was started. Ceftriaxone was started pending the results of blood and CSF cultures. However, fever of the patient persisted, respirations became shallow, his consciousness deteriorated and he had to be orotracheally intubated and mechanical ventilation was started. The repeated CSF examination revealed 70% lymphocytes. Brain magnetic resonance imaging demonstrated gadolinium-enhancing lesion on the right frontoparietal lobe and meningeal thickening (Figure 1a and 1b), and no brainstem lesion was found. On the third day after his admission, *Listeria monocytogenes* was grown in the cultures of CSF and blood, so, ceftriaxone was discontinued and ampicillin (2 g q4h) was started. Within a few days, his consciousness improved, and the results of the repeated CSF examination demonstrated the disappearance of lymphocytosis and improvement in glucose levels to 77 mg/dL.

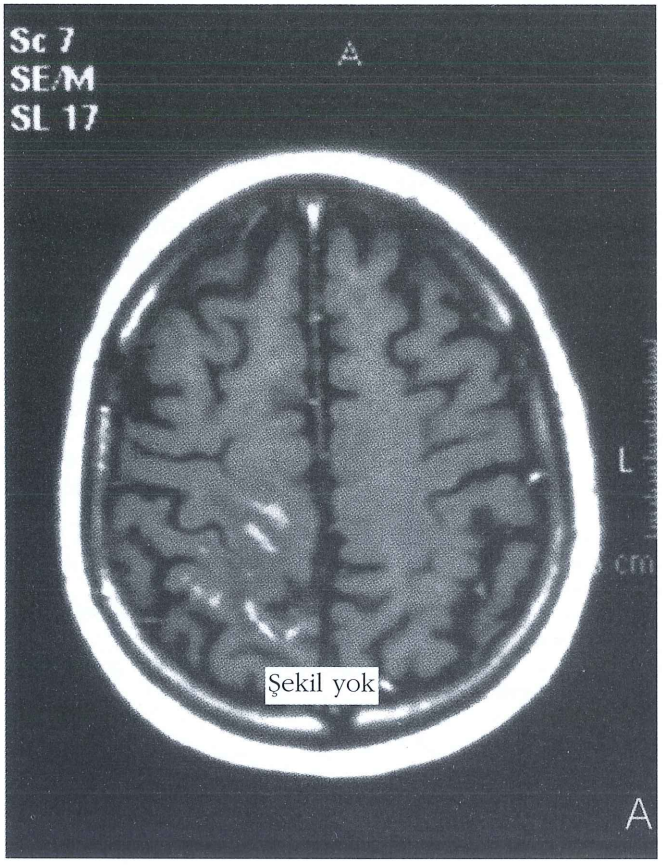

Fig. 1a. *T1 weighted axial image after gadolinium injection shows hypointense lesion on the right frontoparietal lobes, and nodular and linear contrast enhancement of the lesion.*

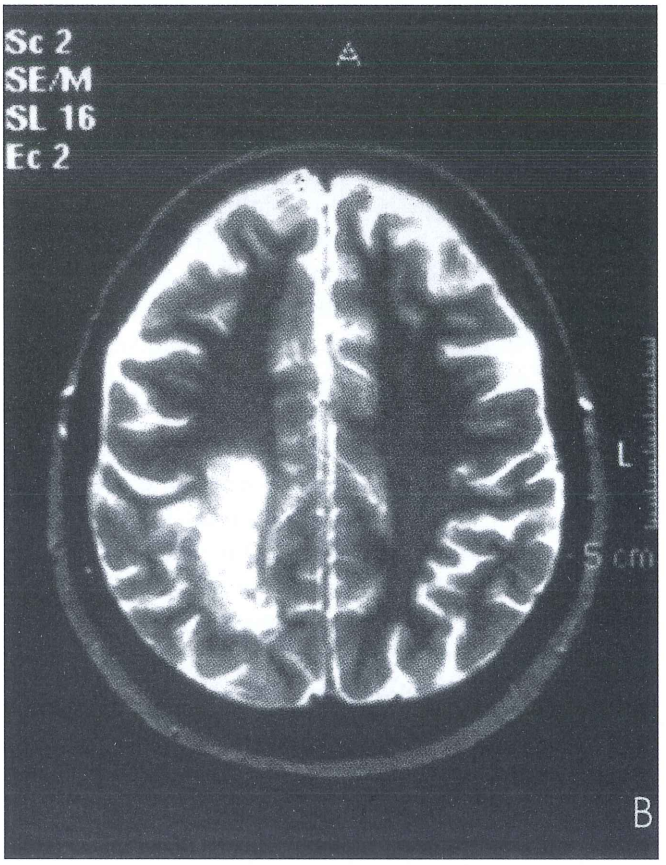

Fig. 1b. *T2 weighted axial image demonstrates large hyperintense lesion on the right fronto-parietal lobes.*

However, he could not be weaned from the ventilator. This was presumed to be due to the respiratory muscle weakness induced by myasthenic crisis. On the 22nd day, tracheotomy was performed. Despite treatment with intravenous immunoglobulin at a dosage of 0.4 g/kg qd for 5 days, his bulbar symptoms and generalized muscle weakness continued for about a week after treatment. Daily plasmapheresis was performed as 8 courses (40 mL/kg qd). During the plasmapheresis treatment, daily weaning trials were performed. Just after the 3rd course of plasmapheresis, the patient was put on pressure support ventilation (Siemens servo 300 ventilator, Sweden) with pressure levels adjusted to achieve a tidal volume above 350 mL and a respiratory rate below 30/min. If the patient did well, the pressure support was decreased 2-3 cmH2O every 2-3 hours. If the symptoms and signs of respiratory distress occurred, the pressure support level was increased to the previous level. When the pressure support level was decreased to 8 cm H_2O, he underwent a t-piece trial. For about 4 days, the duration of t-piece trials were prolonged, and finally, on the 35th day after the beginning of mechanical ventilation, he no longer needed mechanical ventilation. Prednisone, 15 mg, on alternative days and pyridostigmine, 60 mg q4h, were continued. After an uneventful hospital course, he was discharged with only mild left hemiparesis.

Discussion

Listeria monocytogenes causes central nervous system infections mostly in immunocompromized individuals (1,2). On the other hand, these infections are rarely seen in the patients with myasthenia gravis (3). However, combined treatment of azathioprine and prednisone in our thymectomized patient resulted in increased susceptibility to listeria infection.

The development of acute hypercapnic respiratory failure requiring mechanical ventilation and the difficulty in weaning from the ventilator observed in this patient could be due to several causes. Our patient was using prednisone which could predispose to the development of steroid-induced myopathy of the respiratory muscles. Steroid-induced myopathy has been reported to be the cause of difficulty in weaning from the ventilator in a myasthenic patient (4). In this report (4), successful weaning was accomplished after discontinuation of steroid treatment. We do not think, this was the case in our patient because, he was weaned after plasmapheresis treatment, when he was still on steroids during the weaning trials. Brain stem involvement in listeria infection can cause respiratory failure (1,2) and even Ondine's curse syndrome has been reported during the course of brain stem encephalitis due to *Listeria monocytogenes* (5). In our patient the cause of respiratory failure was not brain stem encephalitis because of lack of signs of brain stem involvement and negative neuroradiological findings. The only reason for the development of respiratory failure and difficulty in weaning from mechanical ventilation in this case could be the development of respiratory muscle weakness due to myasthenic crisis provoked by infection. Treatment by plasmapheresis and appropriate antibiotics led to successful weaning. Presence of diaphragmatic weakness (but not diaphragmatic fatigue) was demonstrated in the patients with respiratory failure due to Guillain-Barré Syndrome and myasthenia gravis (6).

Listeria monocytogenes should be considered as a likely pathogen of central nervous system infection in an immunocompromized patient. If this patient is myasthenic, who is prone to the development of respiratory failure, early appropriate empiric antibiotic treatment together with plasmapheresis could be life saving.

References

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