BULBAR ONSET MOTOR NEURON DISEASE: A CASE REPORT

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ABSTRACT

Bulbar onset motor neuron disease is a progressive neurological disorder, which leads inexorably via weakness of limb, bulbar and respiratory muscles to death from respiratory failure three to five years later.[1]

KEYWORDS: Motor neuron disease, amyotrophic lateral sclerosis.

INTRODUCTION

MND’S are a group of progressive neurological disorders that destroy motor neurons, the cells that control essential voluntary muscle activity such as speaking, walking, breathing, swallowing. It is a rare but devastating illness which leads to progressive paralysis and eventual death. The epidemiological method may provide important clues to the causes of MND by demonstrating temporal and geographical trends in distribution.[2] The initial symptoms of motor neuron disease often affect certain areas of the body before eventually becoming more widespread. In case of bulbar motor disease, problems initially affect the muscles used for speech and swallowing; increasingly slurred speech (dysarthria) is usually the first sign of this type of motor neuron disease, increasingly difficult to swallow (dysphagia). Presently I’m reporting a case of bulbar onset motor neuron disease.

CASE REPORT

A 52-year old female presented to neurology department with progressive dysarthria of 2 years duration and worse since March 2017. She was apparently normal till 2 years ago, when she developed slurring speech and which was gradually progressing. No associated dysphagia, no limb weakness, no sensory changes. On clinical examination the was patient was found to be conscious, obeying and with mild bifacial LMN weakness, left palatal palsy,
jaw opening weakness and neck flexors are weak. Trapezius weakness present, tongue fasciculation’s along with brisk jaw jerk and facial reflexes.

Complete blood count, liver function tests, renal function tests were conducted. A slight increase in the count of eosinophil’s-7.3percentage (1-6), and decrease of serum creatinine-0.57 mg/dl (0.6-1.1), globulin-1.9g/dl (2-3.5) and albumin-6.1 g/dl (6.4-8.3) are noted. Pulmonary function test done showed moderate obstruction. The patient was diagnosed with bulbar onset amyotrophic lateral sclerosis and started with EDAVARONE therapy. We planned to give 6 cycles of EDAVARONE therapy, patient underwent first cycle and admitted for second cycle. First cycle 14 Injections, EDAVARONE 60mg as I.V injection over 60min/day for 14 days given, followed by a 14-day drug-free period. Subsequent treatment cycle of EDAVARONE 60mg on 10 of 14 days was planned followed by 14-day drug-free period. Patient seems to be healthy and was discharged with medications Vitamin E (Evion LC) to build muscle strength and Remylin D for neurological pains.

**RESULTS**

Patient with bulbar onset ALS usually present with dysarthria can develop almost simultaneously with bulbar symptoms like jaw opening weakness, facial reflexes and in the vast majority of cases will occur within 1–2 years. Paralysis is progressive and leads to death due to respiratory failure within 2–3 years for bulbar onset ALS cases.[4]

**DISCUSSION**

Amyotrophic lateral sclerosis is a rapidly progressive, invariably fatal neurological disease that attacks the nerve cells responsible for controlling voluntary muscles. Amyotrophic lateral sclerosis, or ALS, is called Lou Gehrig's disease. Only about 5 to 10 percent of all ALS cases are inherited in the family's genes. Early symptoms of ALS are subtle and may include twitching, cramping, or stiffness of muscles; muscle weakness affecting an arm or a leg; slurred and nasal speech; or difficulty chewing or swallowing. No one test can provide a definitive diagnosis of ALS, although the presence of upper and lower motor neuron signs in a single limb is strongly suggestive.[3]

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