Miller Fisher variant of Guillain-Barré Syndrome on a child: Case report

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RESUMO

Introduction: Guillain-Barré syndrome is an acute polynuropathy characterized by peripheral nerve inflammation and demyelination, probably secondary to an immune response against myelin antigens. GBS is clinically diagnosed and supported by complementary exams (cerebrospinal fluid investigation and electrophysiology). It is a monophasic disease that rarely presents relapse and is characterized by rapid, progressive and ascendant muscle weakness, associated with a protein-cytological dissociation of the cerebrospinal fluid. In the present work, we describe a case report of Miller Fisher syndrome, a rare subtype of GBS characterized by the clinical triad: ophthalmoplegia, ataxia and areflexia. Case report: M.F.Z. patient, female, 5 years old, from and residing in Campo Grande, MS. Father reported that child began presenting flu-like symptoms three weeks ago, lasting for three days and symptomatic improvement after use of medication. Two weeks ago, she started with right eyelid ptosis, being referred to evaluation with a neurologist at the Santa Casa which presented unaltered CT scan results. Patient worsened after three days with decreased strength in the lower limbs and upper limbs and difficulty walking. On examination M.F.Z. was conscious without sensory changes, distal paresis in upper and steeper in the lower limbs with deep areflexia and right eyelid ptosis. Father denied previous diseases of the child and family. Gestational maternal history and birth without complications. After 10 days of evolution, elevated cerebrospinal fluid protein (110.70 mg/dL) was found and the child was diagnosed with Miller Fisher syndrome after the exclusion of other conditions. The underlying disease of the patient was treated with IV Ig administered at a dose of 2 g/kg, and 0.5/kg/day for 4 days. The patient showed regression of motor deficit, though still with a slight ptosis at hospital discharge. Discussion: Miller Fisher syndrome is extremely rare in children and is a diagnostic challenge at those ages. It presents bimodal trend with a peak of involvement between the 2nd and 3rd decade and another between the 5th and 7th decade of life. However, 20% of all cases occur in children under the age of 10 years. Miller Fish syndrome may be associated with infectious, autoimmune and neoplastic diseases. It was included as GBS subtype due to the presence of albumino-cytologic dissociation and spontaneous recovery. Despite the weakness not being the main feature, patients have moderate weakness of limbs and decreased proprioception, without sensory loss. The diagnosis is supported by the clinical manifestations and electrophysiological and cerebrospinal fluid analysis tests. The test can have unaltered results during the first week of the disease. Typical changes are found from the tenth day of evolution. There are two forms of treatment: plasmapheresis and intravenous human immunoglobulin, which acts by modulating the immune system. International results of randomized trials demonstrated equivalent efficacy of both these treatments. The outcome is usually good. After reaching the summit, muscle strength returns first proximally then distally, and may have improved motor deficits within 6 months after the end of treatment. Approximately 4 to 15% of patients die and 20% are left with some residual damage, even when treated.

References


