AMYOTROPHIC LATERAL SCLEROSIS: A DISARRANGEMENT OF THE NEUROMUSCULAR’S MACHINERY

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RESUMO

Introduction: Among the motor neuron diseases, Amyotrophic Lateral Sclerosis (ALS) is the most common disease and the most devastating, characterized by the death of lower and upper motor neurons or corticospinal.1,2 Studies show that there are geographical variations in the incidence of ALS3,4. Objective: To present the ALS through the concept approach, epidemiology and clinical disease, thus presenting their signs and symptoms. Materials and Methods: Literature review by theoretical frame of reference, published in English and Portuguese in the period 2005–2015, in humans. The databases were “Medline”, “PubMed”, “Bireme” and “Scielo”. Results and Discussion: There were used the keywords “Amyotrophic Lateral Sclerosis” and “epidemiology” in the article search. There are variations in the incidence depending on the region. The prevalence is about the same among the countries of Europe, America, Asia and Brazil3,4. The median survival is 2-5 years3. It manifests by painless and progressive paresis, a complex clinical syndrome focal1,2,3,4,5, followed by muscular atrophy and cramps. The diagnostic criteria are divided into: bulbar, cervical, thoracic and lumbosacral1,2. The treatment is made by riluzole, that blocks the release of glutamic acid, delaying the disease progression by breaking glutamate neurotoxicity. A multidisciplinary approach is essential for the disease treatment. Frequently, is necessary a symptomatic treatment for dooling, pseudobulbar symptoms, cramps and spasticity1,2,5. When there are present signs of lower and upper motor neurons in the bulbar region and two other spinal regions or three other spinal regions, is defined the diagnosis of ALS5. When these signs are present in only two spinal regions is given the diagnosis of probable ALS1,2. Having a hint of diagnosis of ALS by clinical findings, nerve conduction studies with repetitive stimulation and electromyography (EMG) confirm the lower motor neuron degeneration and exclude the neuromuscular junction disorders. The ALS differential diagnosis is performed by nuclear magnetic resonance, the column to differentiate from Spondylotic Myelopathy (image studies in nervous specific segments), and cerebral, to differentiate from vascular disorders. The incidence in Europe is 2.08, in North America 1.80, China 0.46 and Japan 1.47/100,000, the prevalence, that increases with age3,7,6, in Porto Alegre is 5.07, Europe 5.40, US 3.40, China 1.2 and Japan 11.3/100,000, with wide variation by location5. In Guam Island there is an incidence of 3.9/100,000 people3. It occurs more in men than in women and white people are more affected than non-white people3,4,6,7. The early symptom is the asymmetric muscle weakness2,3. In Brazil, the average age is lower, around 52 years (in Europe is between 59 to 65 years)3. Conclusion: There are differences in incidence of disease, according to sex and race. There are similarities of symptoms in patients with ALS and other diseases, requiring tests for the differential diagnosis.
References