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COMMUNICATION WITH THE OUTSIDE WORLD ONLY BY FACIAL EXPRESSIONS, A CLINICAL CASE OF AMYOTROPHIC LATERAL SCLEROSIS IN A YOUNG WOMAN

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Abstract:

ALS takes away the ability to move, speak, write, print - but does not take away the need to express feelings, emotions and desires. ALS kills the motor neurons in the brain and spinal cord that send signals to different muscle groups. Amyotrophic lateral sclerosis can occur both in the elderly and at a younger age. Given the rarity of the disease at a young age, we decided to highlight the clinical case of ALS in a young woman who communicated with the outside world only through facial expressions. With careful care and timely diagnosis of ALS, despite being bedridden, patients have a survival rate of more than 5 years.

Keywords: neurodegenerative disease, amyotrophic lateral sclerosis, bulbar disorders, facial expressions

RELEVANCE OF THE PROBLEM

Amyotrophic lateral sclerosis (ALS) is usually remembered when people talk about astrophysicist Stephen Hawking in a wheelchair with a computer monitor wrapped in wires. Doctors diagnosed ALS at the age of 21, predicted two years of life for him, but the scientist lived up to 76 years. By the end of his life, Hawking had only one facial muscle of the cheek, with the help of which he communicated with the outside world. To communicate with the outside world, such patients eventually have to resort to special devices. Stephen Hawking, for example, used a special computer system that was controlled by a cheek muscle. And Jason Becker, an American rock musician in the neoclassical metal genre, a virtuoso guitarist, used a board with letters that he pointed to with his eyes.

Amyotrophic lateral sclerosis (ALS) is an idiopathic degenerative disease of the central nervous system, with selective damage to the motor neurons of the anterior horns of the spinal cord and brain stem (peripheral motor neurons) and cortical (central) motor neurons and is characterized by steady progression. ALS is a predominantly sporadic disease, but in 5-10% of cases it has a familial pattern (transmitted in an autosomal dominant manner). In some familial cases, a mutation was found on the long arm of the 21st chromosome - in the gene encoding the enzyme superoxide dismutase-1.

As is known, ALS most often begins at the age of 50-70 years, more often in men, with asymmetric weakness and weight loss of muscles in the distal parts of the arms and/or legs, less often with the development of bulbar disorders or spastic tetra- or paraparesis. In most cases, all limbs and bulbar muscles are involved within 2-3 years. ALS is very rare in young women, so we set out to highlight this clinical case.

A young woman, I.D., was brought to the TMA Clinic for inpatient treatment. born 1981 – 37 years old from Namangan region, who could not complain on her own due to bulbar disorders. According to relatives, the patient cannot speak, lack of voice, choking, lack of movement in all limbs, cannot hold her head and neck.

From the anamnesis of Morbi: She considers herself ill since the spring of 2013, at the age of 31, she worked as a teacher at school. I began to notice weakness in my right hand (I began to often drop chalk from my hands while writing on the blackboard). During the year, weakness developed in the legs. She received inpatient treatment. A year later, by 2014, speech and swallowing disorders were added, after which ALS was diagnosed. From the onset of the disease within 1-1.5 years, the patient stops walking due to severe weakness in the limbs.

Status presents: The patient's general condition is stable and severe. Consciousness is clear. Recumbent passive position, height 160 cm, weight 35 kg. The

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skin is of normal color, elasticity is reduced. Subcutaneous adipose tissue is poorly developed. Peripheral lymph nodes are not enlarged. There are no edema. Body temperature 36.7⁰C. Respiratory rate 20 per minute. Weakened vesicular breathing is heard in the lungs. Heart sounds are muffled, blood pressure is 100/70 mm Hg. Art., pulse 82 beats / min, rhythmic. The abdomen is soft and painless on palpation. The liver and spleen are not enlarged. The stool is prone to constipation. Diuresis independent.

Neurostatus: In the mind, verbal contact is impossible due to anarthria, he understands spoken speech, communicates only with the help of facial expressions, facial expression is calm, adequate, the emotional background is reduced, friendly for examination. Symptom Danzig - Kunakova negative. Study of FMN: 1-pair: no change. 2-pair: no pathology. 3,4,6 pairs: full range of eyeball movements, no nystagmus. The pupils are equal in size, the reaction to light is preserved. 5-pair: Valle points are painless. The strength of the masticatory muscles is equal to 3 points on both sides. 7-pair: asymmetric face, central paresis on the right. 8-pair: hearing is not changed. 9, 10-pair: gross bulbar paresis: aphonia, dysphagia, rhinorrhea, swallowing is disturbed, the soft palate hangs down, weakly contracts during phonation, the uvula is in the midline, not deviated to the sides, pharyngeal reflexes are not caused; 11-pair: does not hold his head on his own due to paresis, cannot raise his shoulders from both sides - a symptom of "hanging head"; 12-pair: cannot protrude tongue, anartria; the tongue is atrophied, there are fibrillations of the tongue, a symptom of "boiling tongue".

In the motor sphere: mixed tetraplegia. There are no active movements in the limbs, there are contractures in the elbow and ankle joints, muscle tone in the limbs is reduced due to severe muscle atrophy. Muscle fasciculations are noted, muscle strength is 0 points in the proximal muscle groups, 1 point in the distal arms, on the legs 0 point in the proximal muscle groups, 1 point in the distal muscle groups. Tendon reflexes are high with an extended reflexogenic zone, BR D=S, TR D=S, PR D=S, AR D=S, positive pathological reflexes: Babinsky, Rossolimo, Jacobson-Laska from 2 sides, reflexes of oral automatism Marinescu- Rodovichi, proboscis on both sides. The sensitive area is unchanged. Coordinator samples: not possible to investigate. There are no meningeal symptoms. Higher nervous activity is not disturbed. The functions of the pelvic organs are preserved.

Laboratory studies: KLA: Hb116g/l, er- 3.9, cp 0.9, leu- 5.9, ESR 14mm/h. OAM: color - s / w, transparency - pr., Rel. density - 1021, reaction - acidic, protein - 0.033%, epithelium 6-5, leukocytes 16-18, mucus +, urates +. Coagulogram: hematocrit 42%. Fibrinogen 288 mg/dl, ethanol test - negative, thrombotest - 5. Blood biochemistry: urea 7.8 mmol/l, creatinine 75.2 mmol/l, glucose 5.1 mmol/l. ECG: Sinus arrhythmia with a heart rate of 100-83 bpm, EOS deviated to the left, intraventricular conduction difficulties. Hypoxic changes in the myocardium.

Differential diagnosis was carried out with the following diseases: Kennedy's bulbospinal amyotrophy, cervical myelopathy, Kugelberg-Welander's spinal amyotrophy.

When making a diagnosis, we used the ALSFRS-R (Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised) app.

- "Amyotrophic lateral sclerosis, bulbar form with the formation of tetraplegia, dysphagia, stage 3".

The patient received symptomatic treatment: riluzole, L-carnitine and protein nutrition.

CONCLUSION: Amyotrophic lateral sclerosis can occur both in the elderly and at younger ages. ALS takes away the ability to move, speak, write, print - but does not take away the need to express feelings, emotions and desires. With careful care and timely diagnosis of ALS, despite being bedridden, patients have a survival rate of more than 5 years.

USED LITERATURE

1. Amyotrophic lateral sclerosis (ALS) and other motor neuron diseases (MND) (Lou Gehrig's disease, Charcot's syndrome) Authors: Michael Rubin, MDCM, New York Presbyterian Hospital-Cornell Medical Center. Medical Review Apr 2022
2. Amyotrophic Lateral Sclerosis: Clinical Heterogeneity and approaches to classification. I.S. Bakulin, I.V. Zakroishchikova, N.A. Suponeva, M.N. Zakharov State Budgetary Institution "Scientific Center of Neurology"
3. Handbook on the formulation of the clinical diagnosis of diseases of the nervous system. Ed. V.N. Stock, O.S. Levin. M.: Medical Information Agency, 2006. C. 149–152. [Reference book on formulation of clinical diagnosis of nervous system diseases.
4. Al-Chalabi A., Hardiman O., Kiernan M.C. et al. Amyotrophic lateral
5. sclerosis: moving towards a new classification system. Lancet Neurol
6. 2016;15(11):1182–94. DOI: 110.1016/S1474-4422(16)30199-5. PMID: 27647646.
7. Kiernan, M. C., Vucic, S., Cheah, B. C., Turner, M. R., Eisen, A., Hardiman, O., ... & Zoing, M. C. (2011). Amyotrophic lateral sclerosis. *The lancet*, *377*(9769), 942-955.

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8. Brown, R. H., & Al-Chalabi, A. (2017). Amyotrophic lateral sclerosis. *New England Journal of Medicine*, *377*(2), 162-172.
9. Gordon, P. H. (2011). Amyotrophic lateral sclerosis. *CNS drugs*, *25*(1), 1-15.
10. Zavalishin, I. A. (2009). Amyotrophic lateral sclerosis: hands. for doctors. GEOTAR-Media.
11. Hardiman, O., Van Den Berg, L. H., & Kiernan, M. C. (2011). Clinical diagnosis and management of amyotrophic lateral sclerosis. *Nature reviews neurology*, *7*(11), 639-649.
12. Ingre, C., Roos, P. M., Piehl, F., Kamel, F., & Fang, F. (2015). Risk factors for amyotrophic lateral sclerosis. *Clinical epidemiology*, *7*, 181.