

peripheral motor neurons, progressive course and onset of terminal respiratory insufficiency. The amount of patients including ones in Uzbekistan that are suffering and dying from ALS is increasing. The development of a familial ALS (FALS) shows the role of mutations of the gene producing the superoxide dismutase 1.

Data on morbidity of ALS on base of register for the period of 2013–2014 was studied. Special attention was paid to geographic epidemiology of ALS in different regions of Uzbekistan and to features of clinical manifestations of this disease in Uzbek nationality.

During the analyzed period there were 3.4 cases of ALS per 100,000 per year registered. 70% were patients from Andijan valley, 30% from other regions. Gender analysis determined that 60% of patients were women (mean age 43 ± 4.2 years), 40% men (mean age 48 ± 3.4 years). Most patients associated the debut of ALS with virus disease. Disease of 80% of patients debuted by bulbar form, of 20% by spinal form. The FALS is absent in our country. The average life expectancy was 2.5 ± 0.45 years during the bulbar form and 3 ± 0.86 years during the spinal form. The onset of decompensation period on the average 6 month later after debut.

The analysis of the register of ALS in Uzbekistan demonstrated that ALS was the most common disease in Andijan valley, primary occurred in women, is not of a personal nature and mainly declares itself by bulbar form.

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Motor Neuron Disease 1

Rehabilitation of blood-spinal cord-barrier toward amyotrophic lateral sclerosis therapy

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Background: ALS patients and transgenic mice expressing ALS-associated superoxide dismutase 1 (SOD1) mutations show alterations in the blood-spinal cord barrier (B-SC-B) as suggested from the reduction of levels of various tight junction proteins (TJPs) including zonula occludens-1 (ZO-1), occludin and claudin-5 between endothelial cells and early protection of the B-SC-B integrity was found to delay onset of motor-neuron impairment and degeneration.

Objective: The aim of this research was to investigate if inhibition of the axis CXCL12/CXCR4 receptor widely expressed in neurons and glial cells and modulates neuronal apoptosis, may improve motor neurons survival by increasing the expression of tight junction proteins and rehabilitation of the barrier.

Materials and methods: Transgenic mice model of ALS were treated with AMD3100, antagonist of CXCR4. Motor function, weight changes and survival were evaluated. In a separate experiment, mice were sacrificed after one month of treatment and levels of proteins essential for the formation of the barrier in comparison with proteins that do not participate in the barrier were measured.

Results: We found that chronic administration of AMD3100 to ALS mouse model was effective in restoring the expression of tight

junction proteins and considerably increase the survival, confirming the importance of early treatment for rehabilitation of the barriers to prevent infiltration of neurotoxic products and microhemorrhages.

Conclusions: These data reveal that multi-faceted action of AMD3100 may provide a novel option for ALS therapy leading to rehabilitation of B-SC-B proteins and thus preventing additional damage to motor neurons.

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Motor Neuron Disease 1

Far beyond our typical dengue fever on three cases reported: weakness, visual loss and aphasia as initial clinical presentations?

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Background. The mosquito-borne dengue virus human infection is endemic in Brazil and the most populated São Paulo State concentrates a great number of cases. There is a wide range of possible clinical manifestations. Secondary dengue-related diseases and complications can show up with nonspecific symptoms to more severe hemorrhagic shock. Possible neurological manifestations are also a part of initial clinical assessment of an infected patient and can define severity of affection. Objective. This cases report aims to study complications dengue fever can present with and alert health care professionals dealing with this disease.

Patients. Here we present three cases of adult patients admitted in March 2015 at our hospital during a current epidemic of dengue viral infection in the State who experienced neurological complications in the convalescent phase of infection: a man with weakness (Guillain-Barré syndrome), a woman with visual loss (unilateral maculopathy), and a woman with aphasia (post-seizure Todd's palsy). The patients had in common a previously acute febrile disease diagnosed as dengue fever.

Results. After specific investigation and treatment, all of them had their complaints improved.

Conclusion. These various disease presentations on patients referred to neurological care should be promptly diagnosed and treated. To call attention and inform health teams on these different presentations is essential. The dissemination of new clinical guidelines for health professionals on non-tertiary services including these neurological complications might be useful and can yield better outcomes.

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