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Psychiatric disorders and their medical-social consequences in patients with amyotrophic lateral sclerosis and members of their families

Abstract

118 ALS patients and 97 members of their families were examined. All patients and relatives were observed by psychiatrists. The assessment was carried out on the Hamilton Depression Scale (for patients and members of their families) and the scale of frontotemporal dementia (for patients). Psychiatric disorders were revealed in 101 ALS patients (85%) and in 51 members of their families (52.5%) (Significantly more frequently in ALS patients, $p<0.05$). The spectrum of situational disorders in ALS patients was represented relatively evenly with insignificant predominance of situational depression. Rare cases of combination of ALS with endogenous psychiatric disorders were described. In 33 patients (28%), frontotemporal cognitive impairments reaching the level of dementia were observed in 4.2% of cases. Among members of families of ALS patients, only situational psychiatric disorders with obvious predominance of anxiety (28%, $p<0.05$) were revealed. 49 ALS patients (41.5%) and 80 members of their families (82.4%) suffered from habitual intoxications (significantly more frequently in family members, $p<0.05$). High share of anxiety disorders leads to the refusal from cooperation with professional physicians (73-76%), rejection of aggressive treatment methods (74.5%), addressing to cheaters and burden connected with unjustified material expenses (29.6% of cases). Constant nursing of families of ALS patients by neurological and psychotherapeutical services with involvement of psychiatrists and narcologists in individual cases is recommended, which could provide increase of the number of life-saving procedures at ALS and improvement of the quality of life of patients and members of their families.

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Abbreviations

ALS, amyotrophic lateral sclerosis; HDS, Hamilton depression scale; FTDS, frontotemporal dementis; ALV, artificial lung ventilation; ET, early tracheostomy

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease accompanied by gradual development of complete akinesia, impairments of speech, swallowing, respiration, and, in a number of cases, frontotemporal cognitive impairments or dementia.[1,2] Based on the data of foreign authors, depression requiring constant monitoring by the psychiatrist and/or psychotherapist is observed approximately in a half of ALS patients.[3] ALS becomes a heavy load for caregivers (family members, friends, nurses), which cannot remain non-reflected in their psychiatric state, at that, to the greater extent, the psychiatric state depends on impairments of behavior of ALS patients, rather than the locomotor defect.[1,4] The objective of this study was investigation of the spectrum of psychiatric disorders in ALS patients and members of their families.

Materials and methods

In the period from 2007 to 2013, 118 ALS patients and 97 members of their families were examined. The psychiatric examination was performed at the basis of the Department of Psychiatry and Suicidology, Municipal Hospital № 20 (V.M.G.), and at home (A.S.L.) and included interviewing and assessment on the Hamilton Depression Scale (HDS) at personal meeting with the patient or by e-mail and the scale of frontotemporal dementia (FTDS) at personal meeting with the patient's relative or by phone.[5,6] The diagnosis ALS was set based on the Revised El-Escorial Criteria of 1998 taking in consideration the results of examination by the neurologist or revision of medical document with registration of results of neurological examination, needle and stimulation electromyography and neurovisualization of the central nervous system.[7] The dynamic observation 1 time in 2-3 months in the forms of examination by the neurologist and psychiatrist in the out-patient regimen or at home was offered to all patients. All patients and their relatives were examined by psychiatrists (V.M.G. or A.S.L.) The diagnosis of different psychiatric disorders was determined based on the ICD-10. Single episodes were taken in account. At that, the urge to be examined more than twice for confirmation of the diagnosis, multiple attempts to obtain the information about ways of ALS treatment from the internet, including the night time and aspiration to spend more money to disorder and treatment were referred to manifestations of the spectrum of anxiety. Tobacco smoking more than 10 cigarettes a day, consumption of more than 200ml of alcohol a week, drug dependence with constant intake (non-steroid anti-inflammatory, antihistamine drugs) for more than 1.5months with withdrawal syndrome, the use of cholinolytics as remedies for sialorrhea treatment and psychotropic drugs (sedatives, tranquilizers, antidepressants) for more than 1.5months with withdrawal syndrome as well as administration of narcotic medicines prohibited by the RF legislation, were regarded as habitual intoxications (abuse).[8] The statistical processing was carried out using Student's criteria and chi-square, Spearman correlation analysis. The statistical software package Statistic 7.0 was applied.

Results

Psychiatric disorders were revealed in 101 ALS patients (85%) and 51 members of their families (52.5%). ALS patients suffered from psychiatric disorders significantly more frequently than members of their families ($p<0.05$). The spectrum of revealed psychiatric disorders is represented in and medical-social consequences of these disorders – in.

From it can be seen that the majority of ALS patients had situational disorders (n=63, 53.3%), while 5 patients (4.2%) had endogenous diseases or syndromes (1 case of acute psychosis was registered). At that, in 33 patients (28%) frontotemporal cognitive impairments were observed (mean log rank 1.47 [1.92; 1.26]), reaching the level of dementia in 4.2% of cases (mean log rank -0.4 [1.07; -3.09]). The mean score on the HDS in ALS patients with situational depression was 17±4, with situational anxiety disorder - 15±5, and with situational anxiety-depressive disorder - 21±6 ($p>0.05$ between the groups). The spectrum of situational disorders in ALS patients was represented relatively evenly with insignificant predominance of situational depression. In the process of specification of family psychiatric anamnesis it was revealed that in ALS patients with endogenous psychiatric disorders family cases of psychiatric disorders were observed, while in patients with situational disorders indirect documentary unconfirmed data on psychiatric impairments in relatives were received in 15% of cases.

Among members of families of ALS patients, only situational psychiatric disorders were detected with obvious predominance of anxiety ($p<0.05$). The mean score on the HDS in family members of ALS patients with situational depression was 12±4, with situational anxiety disorder -22±5 ($p<0.05$ in comparison with the previous parameter), and with situational anxiety-depressive disorder -16±6. Single cases of ALS phobia and other types of phobia were noted, as well as cases of paranoiac syndrome in two families of ALS patients, which manifested with delirious ideas of causing material damage for medical personnel. It was not possible to collect family anamneses of psychiatric disorders in these families, however, the information about presence of discirculatory or post-traumatic encephalopathy and experience of work in extreme situations and/or on the leading position in members of these families was documented.

From, it can be seen that the majority of families at different stage of the disease of the ALS patient refused dynamic monitoring in the form of personal or electronic communication (61.8%) and addressed to methods of ALS treatment not stipulated by the International Standard (64.4%). The refusal from primary examination by the doctor from our group was seen in patients with probably severe psychiatric disorders and syndrome of social isolation only in 7.6% of cases.

Methods stipulated by the International Standard of ALS treatment included the therapy with Riluzole, non-invasive (NLV) and artificial lung ventilation (ALV), drugs for sialorrhea treatment, psychotropic drugs, adaptive-home devices.[9] It should be noted that the single treatment method currently not included in the International Standard of ALS treatment, but offered to patients by the co-author (G.N.L.), was early tracheostomy (ET), which was justified for the treatment of impairment of expectoration in patients with bulbar debut of ALS not able to adapt to the NLV. The NLV, ET and ALV were referred by the authors to the methods of aggressive palliative therapy.

Methods not included in the International Standard of ALS treatment comprised any experimental pharmaceutical treatment methods, high doses of neuroprotective drugs in the form of infusions based on the physiological solution with the risk of development of central pontile myelinolysis in non-swallowing patients with long-term hyponatremia, including injections in collapsed veins with the risk of thrombophlebitis development; intramuscular injections in atrophied muscles with the risk of development of post-injection complications and non-absorption of (expensive) drugs; treatment with stem cells, as well as hyperbaric oxygenation, immunotherapy, chemotherapy and physiotherapeutic treatment methods in case if these methods were presented by people offered them as means directed to slowing of progression of the disease. In case if experimental treatment methods were realized on the commercial basis and presented as means able to cure the ALS patient or improve their condition, the authors regarded these activity as fraudulent.

The refusal from any aggressive ALS treatment methods indicated at the moment of examination was registered in 74.5% of families of ALS patients. It should be noted that the ALV (that is artificial prolongation of life at ALS) is not indicated to elderly ALS patients, as well as in case of frontotemporal cognitive impairments, psychiatric disorders, expressed toxic abuses, since it is not possible to determine clearly a patient's determination for this action and document a will specifying conditions for disconnection of these patients from the apparatus in the future.[10]

It is necessary to mention that the refusal from the use of aggressive ALS treatment methods and, sometimes, repeated visits of doctors and administration of drugs, was always seen in families of ALS patients with endogenous psychiatric disorders, frontotemporal cognitive impairments or dementia and psychiatric disorders, however, members of these families were either adapted (in case of presence of premorbid psychiatric peculiarities in ALS patients) or readapted (at development of psychiatric peculiarities in patients within the frame of this disease) to this difficult situation.

Addressing to cheaters that resulted in significant material expenses was seen approximately in one third of families of ALS patients (29.6%). This being the case, only 15.2% of the addressed people were treated abroad. From them, about one third of families were treated in organizations regarded by us as fraudulent.

At analysis of habitual intoxications in families of ALS patients, it was found out that 49 patients (41.5%) and 80 members of their families (82.4%) suffered from them. In family members of ALS patients, habitual intoxications were observed significantly more frequently ($p<0.05$). Among ALS patients, tobacco smoking and drug abuse were distributed almost evenly (16.9% and 18.6%), while alcoholic and narcotic dependence were represented by single cases (4.2% and 3.4%) ($p<0.05$). Among members of families of ALS patients, toxic abuse (with the exclusion of one case of narcomania) were represented approximately equally (17.5% and 19.5%) with slight predominance of tobacco smoking (28.8%) ($p>0.05$). Drug dependence both in ALS patients and members of their families, in general, was observed in connection with administration of hypnotic and non-steroid anti-inflammatory drugs (painkillers).

The analysis of intake of cholinolytic drugs and abuse connected with their administration revealed that ALS patients used them, mainly, for sialorrhea treatment often ignoring their intake as psychotropic

drugs (16,1%), while members of their families, in general, used them in the form of antidepressants (15.4%) ($p>0.05$).

The comparative analysis of distribution of frontotemporal cognitive disorders and the use of cholinolytic drugs for ALS treatment demonstrated that drug abuse from the latter developed in 15 from 33 patients (15.3%) with frontotemporal cognitive impairments. In general, it was characteristic for patients with bulbar debut of ALS required several cholinolytic drugs for correction of sialorrhea, as well as emotionally-volitional disorders. Data reliably confirming strong correlation connection between the presence of frontotemporal cognitive, bulbar disorders and requirement of two cholinolytics were received ($r=0.7$; $p >0.001$). At that, in one patient, withdrawal of cholinolytics penetrating the central nervous system with change to botulinum toxin for sialorrhea correction led to improvement of assessment on the scale of frontotemporal dementia and ability to critical analysis based on the Hamilton scale in 2 months.

Discussion

Previously, neither in the international nor domestic literature, the spectrum of psychiatric disorders in families of patients with one of the most severe disorders (ALS) was considered as a whole. Besides, the analysis of medical-social consequences of these disorders was not performed. Such factors as some types of toxic abuse (intake of alcohol and smoking) were previously regarded in the international literature only as risk factors of ALS development.[11,12] However, taking in consideration insufficient knowledge of genetics of this disease and the risk of its development in the family, in this work the authors made an attempt, in particular, to assess the risk of medical-social complications in families with the persisting huge medical-social problem, in cases with ill and not yet ill people. Investigations of cooperation of ALS patients are scarce and devoted only to some treatment methods, at that, studies of harm for families of ALS patients connected with fraudulent activity of cheating organizations have been never performed (1). Currently, the only organization dealing with the informational struggle in this area is the site www.alsuntangled.com. The actuality of investigation of psychiatric condition of ALS patients and their relatives is connected with the circumstance that the lifespan of ALS patients directly depends on their psychiatric state.[13]

It has been shown that severe depression in ALS patients can be diagnosed in 9% of cases, moderate and mild in 37% of cases.[3] The value of severe depression in this work approximately corresponds to the percentage of ALS patients refused primary examination by the physician in our study (7.6%). If the latter is added to the total percentage of situational depression in our work (moderate and mild based on the mean score on the HDS 17±4) the sum equals to 33.6% meaning that it is slightly lower than in the study of J.G.Rabkin.[3] At insignificant predominance of situational depression in the spectrum of situational disorders in ALS patients, the severity degree of all of these disorders on the HDS is approximately the same, being closer to moderate.

In the current study, in the spectrum of psychiatric disorders among members of families of ALS patients, anxiety disorders dominate significantly both on the number (29.8%) and severity degree (22±5). The second place is taken by situational anxiety-depressive disorders, with the role of situational depression being insignificant. The high share of anxiety cases appears to lead to impairment of healthy lifestyle, habitual intoxications (82.4% among all family members), the refusal from cooperation with professional physicians (73-76% of cases) and addressing to cheaters (29.6% of cases). Dissociation observed in the conscious of ALS patients can be the reason, based on the data of foreign studies: 94% of patients do not want to speed up approaching of the lethal outcome, but, at that, they refuse gastrostomy and NLV significantly more frequently, in comparison with their relatives, which causes feelings of "suffering" and "loneliness" in members of their families (according to the Hospital Scale of Anxiety and Depression) that are higher than in patients themselves.[14] It is quite obvious that both ALS patients and members of their families often need active psychological nursing and sometimes consultation of the psychiatrist or narcologist.[15]

If a patient with ALS takes the drug Riluzole metabolized by cytochrome CYP2A1 and causing drug-induced hepatitis in 3-12% of cases, it is necessary that the patient have reduced the number of cigarettes a day, as smoking intensifies the work of this cytochrome, and abstained from strong alcoholic beverages for achievement of the maximal activity of the drug and minimization of adverse effects.[1,9]

The analysis of correlation of frontotemporal cognitive, cognitive, bulbar disorders and administration of cholinolytics, which have not been earlier performed in the international and domestic literature, deserves special attention. The fact of partial regress of frontotemporal cognitive and psychiatric disorders at withdrawal of cholinolytics at ALS, which requires further investigation, has been performed for the first time.

Conclusion

The following general recommendations on the life-style for ALS patients and members of their families are obligatory: they should maintain the proper regimen of sleeping and wakefulness, avoid any types of toxic abuses, and address for help to specialists in the area of psychiatry, psychotherapy and narcology in difficult situations.

Taking in consideration high prevalence of psychiatric disorders among ALS patients and members of their families in the RF, it is recommended to perform joint consultations for patients and members of their families by the neurologist and psychiatrist (psychotherapist), because other consultation conditions can be connected with high professional harmfulness for both the physician-neurologist and specialist in the area of ALS. Multidisciplinary monitoring of ALS patients should include, by all means, regular observation by the psychiatrist and/or psychotherapist, and, along with the disease progression, mostly at home, consultations of members of their families. Cholinolytic drugs should be used with caution in ALS patients taking in consideration the presence of frontotemporal cognitive disorders in some of them. In cases, when patients with ALS have frontotemporal cognitive and moderate or expressed psychiatric impairments, as well as endogenous psychiatric diseases, the psychiatric consultation is required. It should be actively recommended by other physicians having contacts with the ALS patient.

Psychiatric examination is regulated by articles 23, 24, 25 of Law of the Russian Federation "On psychiatric help and guarantee of rights of citizens at rendering psychiatric help". Obtaining consent for the interview with the psychiatrist in cases not included in the term "acute psychosis and confusion" is a duty of the physician in charge of the Department of General Somatic Diseases. Voluntary transfer to the psychiatric department is performed based on article 28-35 of the same law and confirmed by the consent of the patient and their representative in the case report form and the record of the

specialist. In case of severe psychiatric disorders, the decision about involuntary hospitalization is made. The decision and its justification are referred only to the competence of the psychiatrist based on the points "A" and "B" of article 29 of the RF law "About psychiatric help". The decision about involuntary examination is made by the psychiatrist and documented in the case report form by the record of the physician-in charge, Head of the Department, Head Physician or its Vice-Head. In case verbal or written message informs about direct danger of the patient for themselves and other people, the decision about involuntary examination and transfer of the patient to the psychiatric department is made by the psychiatrist immediately without the patient's agreement and their juridical representatives' permission.

The decision about patient's competence is made by the court. The court designates a psychiatrically healthy person responsible for making decisions connected with the ALS patient's health. At the first stage, an ALS patient refusing the treatment, based on the decision taken by the court and agreement of members of their family, is subjected to involuntary hospitalization in the psychiatric hospital in case of their immediate danger for themselves and other people or their helplessness, that is incapability to satisfy their life-related requirements on their own, or in case of the risk of causing significant damage to their health due to worsening of the psychiatric condition, in case the person having been left without psychiatric help. In this case, life-saving palliative operations (gastrostomy, tracheostomy) can be performed to the ALS patient for improvement of the quality of life of the patient and members of their family, prolongation of the patient's life with the improved quality.

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Conflicts of interest

Author declares there is no conflict of interest.

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