DIFFERENTIAL-DIAGNOSTIC ASPECTS OF LATERAL AMYOTROPHIC SCLEROSIS

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The lateral amyotrophic sclerosis (LAS) is a systemic lesion of the corticomuscular pathway which has an unspecific relation to metabolic diseases (17). In certain cases the vegetative nervous formations (4), the brain stem structures and the reticular formation (5) can be affected.

According to T. L. Bunina (1), M. S. Rossina (4), L. T. Kurland et al. (15) and others about 20 000 persons in all the world are taken ill with LAS every year. In the USA, Canada and Europe the LAS incidence rate is 4—6 per 100 000 inhabitants but on the Marian's islets it reaches 420 per 100 000 (11). The clinical, etiopathogenical and therapeutical aspects of LAS have been studied by numerous authors (2, 3, 7, 13, 19, 25). Besides the classic picture described by Charcot there exists also a syndrome of LAS. L. T. Kurland et al. (16) report this syndrome in cases of neurolues and epidemic encephalitis and I. Simek (24) — in a case of high spinal compression. T. L. Bunina (1), B. Schott et al. (21) point out that the vertebro-basilar insufficiency can sometimes imitate the LAS syndrome. T. P. Stortebecker (22) observed the LAS syndrome in cases of degenerative lesions of discs $C_{4\ 6}$. I. Boudouresques et al. (8) described the LAS picture in a case of cervical myelopathy caused by spondylarthrosis, platybasis, discal hernia and rheumatic deformation of cervical vertebrae. E. Klimkova-Deutscheva (4) reports the LAS syndrome in a case of vibrational disease. M. Sercl et al. (23) associate the development of LAS in single instances with severe infectious diseases, gastrointestinal lesions, hard physical work and the presence of cranio cerebral injury. N. A. Shapoval (6) describes the LAS syndrome with a chronic form of Ixodes encephalitis. Recently the etiology of this disease is associated with the so-called slow viruses.

Material and methods

In the period from 1979 till 3rd quarter of 1982 in the Clinic of Neurology at the Department of Neurology and Neurosurgery, Higher Institute of Medicine — Varna city, 20 patients (7 with Charcot's disease and 13 — with LAS syndrome) were hospitalized. The patients' distribution according to sex and age is presented on table 1.

Results and discussion

One can see that LAS occur most frequently in the 4th and 5th decade. The initial signs of disease begin at the age between 45 and 72 years. In the Ist group the initial symptoms are located in the upper limbs in 4 patients and in the lower ones — in other 2. The bulbar symptoms dominate clinically in 3 patients.

Table 1.

Distribution of the patients according to sex and age

Sex	Age in years	40—50	51—60	61—70	71—80
Females	6	2	3	1	—
Males	14	4	5	3	2
Total	20	6	8	4	2

The neurologic symptomatics of LAS is clearly outlined in 5 cases, while the clinical picture shows certain discretion and atypical nature in other 2 patients. 5 of these patients are physical workers and 2 are intellectual ones. In one case the clinical symptoms begin after an acute viral illness, in two cases — after sustained physical labour while in other 4 cases the presumed provoking factors are still unknown.

In the second group the initial signs affect the upper extremities in 7 patients and the lower ones — in one patient only. The bulbar phenomena occur in 4 patients. In 11 cases the diagnosis is made during the first hospitalization but in 2 ones — in the repeated hospital stay. 9 of these patients are physical workers and 4 are intellectual ones. A deformans cervical spondylosis is established in 6 patients, a vertebro-basilar insufficiency is found in 3 ones and a polyarthritis — in another one patient only. The presumed etiology remains unclear in the other 3 cases.

The progress of the clinical picture and the initial symptoms of the disease coincide with the literature data available. The LAS syndrome is diagnosed by us in more than a half of our patients with this disease studied. The bulbar phenomena could be due to the disturbance of infratentorial haemodynamics and to the cervical spondylosis alone or to their combination. In one patient the LAS manifestations are preceded by an acute cerebrovascular insufficiency. In accordance with L. T. Bunina (1), E. Eiben et al. (11) we accept that the diagnostics of LAS requires longer observations and a wider laboratory information. A number of authors report diagnostical difficulties in atypical cases of this illness (9, 10, 12, 20). The interpretation of the neurological phenomenology from the etiological point of view is difficult in a few of our cases only. The clinical characterization shows that not all cases of combined lesion of the cortico-muscular pathway could be added to the LAS. A multidirectional information revealing the etiological moment for the clinical picture is essential to differentiation between the symptomatic LAS and the classic form of the disease, indeed.

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ДИФФЕРЕНЦИАЛЬНО-ДИАГНОСТИЧЕСКИЕ АСПЕКТЫ ЛАТЕРАЛЬНОГО АМИОТРОФИЧЕСКОГО СКЛЕРОЗА

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РЕЗЮМЕ

Авторами проведена клиническая оценка 20 случаев, из которых 7 относятся к классической форме болезни, а 13 можно отнести к синдрому латерального амиотрофического склероза. Первые признаки заболевания наступают в среднем и пожилом возрасте. Подчеркивается необходимость в комплексных динамических исследованиях заболеваний с инициальными проявлениями болезни Шарко. Это имеет значение для более четкого разграничения классической формы болезни от синдрома латерального амиотрофического склероза.