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Remarks Concerning Root Clinical Manifestation of Amyotrophic Lateral Sclerosis (Coincidence or Evolution of the Disease)

Uwagi dotyczące korzeniowych przejawów klinicznych stwardnienia bocznego zanikowego (współistnienie czy ewolucja schorzenia)

Amyotrophic lateral sclerosis is a degenerative disease of the nervous system, whose etiology, despite extended investigations, still remains unestablished (7, 9, 13).

The role of genetic factors is discussed: the so-called family cases (4, 11), metabolic disturbances (7, 9, 13), toxic factors (7), slow viruses infections (79), anomalies within DNA (7), autoimmunological phenomena (79).

The lack of certain information concerning etiology of the disease is the reason why its efficient treatment still cannot be undertaken. Great hopes were awoken by the attempts at applying TRH and bromokryptine (5, 8). However, they turned out to be inefficient.

The essence of the disease is degeneration of pyramidal tract, of pyramidal cells and of the anterior horn of the spinal cord, as well as of nuclei of certain cranial nerves (4, 9, 10, 11).

The dynamics of the disease largely depends on the clinical form as well as on the degree of the disturbance of axonal trnsport (1).

On the basis of the prevailing symptoms one can distinguish the following clinical forms of the disease: classic form, bulbar form, pseudopolyneuropathic form defined as lumbar or peroneal (2, 3, 6, 9, 14).

In the paper there have also been presented the reversible forms of motoneuron disease (12).

In pseudopolyneuropathic form of amyotrophic lateral sclerosis the first characteristic symptom of the disease is the presence of the paresis of the foot dorsal flexor which may be the only clinical manifestation of the disease for many years. Only later traits characteristic of amyotrophic lateral sclerosis do occur.

Pseudopolyneuropathic form of amytrophic lateral sclerosis in the initial stage of the disease may cause considerable diagnostic difficulties, leading to the wrong diagnosis and even to making attempts at operative treatment in the belief that ailments and symptoms are the consequence of the process causing the compression of nerve roots or of cauda equina.

In Neurology Clinic of the Medical Academy in Lublin we made observations of three patients suffering from with amyotrophic lateral sclerosis, in whom the first ailments and symptoms approximated those being concomitants of the paralytic form of the sciatic neuralgia in the course of pulpaceous nucleus hernia of the intervertebral disc.

CASE DESCRIPTION

A patient J. W., aged 38, a manual worker, was admitted to Neurology Clinic due to prolonged, lasting one year, weakening of the muscular force of the left inferior limb. Reticulographic examination, carried out 5 months earlier, revealed the presence of pulpaceous nucleus hernia of the intervertebral disc on the level L4-L5 with the symptoms of light impression of the fifth lumbar root on the left side. The applied pharmacological and sanatorium treatment did not improve the state of his health. The patient did not give his consent to operation. Internistic examination did not show any deviation from the norm. Neurological examination revealed the presence of fibrillation within the tongue, limited extension weakening of the right hand, probably of post-traumatic origin, single fasciculation in pelvic and shoulder girdles, muscular atrophy in the further part of the left inferior limb and the atrophy of pelvic girdle muscles. Fasciculation was also present in the right inferior limb, although it was weaker. Within the whole left inferior limb a considerable weakening of deep reflexes and the weakening of muscular strength was noted. Pathological reflexes and dysaesthesia were not found.

Cerebrospinal fluid showed an increased level of protein (0.99 g/l). The obtained myelographic picture was normal. Electrographic examination comprised the following muscles: left quadriceps muscle of the thigh, left tibial anterior muscle, left short extensor muscle of the toes, left gastrocnemius muscle and left abductor muscle of the great toe. A high-level neurogenic record was demonstrated, probably on the level of the anterior horns of the spinal cord. The examination of nervous conduction also showed a neurogenic process.

On the basis of a detailed analysis of the ailments, symptoms, evolutions of the disease as well as of results of auxiliary specialistic examinations amyotrophic lateral sclerosis was finally diagnosed.

A patient L. P., aged 53 years, a farmer, was admitted to Neurology Clinic due to his complaints about resting dyspnoea, slimming of the muscles of the superior and inferior limbs, decrease of his hands' efficiency, specially that of the right hand. The patient had pains in lumbar-sacral area for ten years. For three years now the pain has been radiating along the right inferior limb. One and a half year ago the falling of the right foot was a concomitant pheomenon. Just before his admission to Neurology Clinic the patient had stayed in Neurosurgery Clinic, where an operation was performed on the structures of intervertebral discs' pulpaceous nucleus on the level L4-L5 and L5-S1, present in the canal. During internistic examination no deviations from the norm were found. Despite that, the patient complained of the resting dyspnoea. The atrophy of hypodermis was generalized. There was found the atrophy of shoulder girdle muscles, as well as that of the muscles of arms, forearms, hands and inferior limbs, specially of the right shank muscles. The muscular force was distinctly

reduced. Deep reflexes in the superior and inferior limbs were weakened, but no dysaesthesia was revealed. Lack of extension of the right foot and toes was noted. No pathological reflexes were found. The Lasegue's symptom was present on the right side at the angle of 60°.

In the radiogram of cervical spine porosis and sharpening at the shaft's edges were found. Electromyographic examination comprised the following muscles: left deltoid, right interosseous dorsal I, right anterior tibial, right short extensor muscle of the toes. Nervous conduction was examined in the right ulnar nerve and right peroneal nerve. The obtained results of the electromyographic examination and of the conduction examination testified to the neurogenic process of a small (bioelectrically) degree in the superior limb, and long-term, markedly advanced process in the left limb. Anomalies in electromyographic record were related with great probability, to the damage of the spinal cord. Based on the whole of clinical observations, motoneuron disease was diagnosed in the form of an untypicaly proceeding amyotrophic lateral sclerosis.

A patient Z. S., aged 45 years, a postwoman, was admitted to Neurology Clinic due to the weakening of the right foot. This symptom had occurred ten months earlier for the first time. For the last three years muscular contractions within shanks and thighs occurred periodically. She was ill many times and fell down while walking. In the past the patient had twice attacks of pain of the radicular syndrome character in the lumbar-sacral region. On admission she did not report any ailments of that type. During internistic examination no deviations from the norm were found. There was a post-traumatic cicatrix within the skin of the left wrist. Neurological examination revealed the following symptoms: marked hyperreflexion in the superior limbs at the normal muscular force and absence of pathological reflexes, periodically occurring fasciculation in the inferior limbs, falling down of the right foot due to the weakening of power of its extensor muscle, intensification of the patellar reflex and Achilles tendon reflex, tendency to the Babiński's sign (extensor plantar response) on the left side, the weakening of superficial sensibility within the right inferior limb.

Basic examinations did not show any deviations from the norm. The composition of cerebrospinal fluid was also normal. The review radiogram of the lumbosacral section of the spinal cord revealed a curvature of the lumbar part with a marked asymmetry of the intervertebral fissure L3-L4, with its narrowing on the left side. Reticulographic examination showed the normal picture. Electromyographic examination comprised the following muscles: right anterior tibial, left short extensor muscle of the toes. The conduction in the peroneal nerves was examined bilaterally. The obtained results of electromyographic examination as well as conduction examinations showed neurogenic changes, probably on the level of anterior horns of the spinal cord. The patient was discharged from the Clinic with the diagnosed motoneuron disease in the form of amyotrophic lateral sclerosis.

DISCUSSION

While presenting the cases, it is difficult to determine unequivocally if we have to do with the coincidence of occurrence of amyotrophic lateral sclerosis and of simple radicular-spinal syndrome of the lumbosacral region, or whether the presence of radicular symptoms is only one of the consequences of the development of the so-called pseudopolyneuropathic, that is peroneal, form of the motoneuron disease. In all the patients there occurred the weakening of the muscles supplied by the peroneal nerve. Each of the patients had a dyscopathy of the lumbosacral section of the spinal cord, which at first made us treat the weakening of the muscles as a certain or doubtful consequence of radiculopathy. One of the patiens had been subjected to an operation of the prolapsed pulpaceous nerve of the intervertebral disc, after which the function of the foot did not improve. Only in one patient there were found changes in the tongue muscles in the form of fibrillation. In two of the patients deep reflexes were weakened, or simply stopped. In amyotrophic lateral sclerosis deep reflexes are usually markedly raised as a result of the damage of the pyramidal tracts. Electromyographic examination in all the patients displayed, in a certain or probable way, the changes of neurogenic origin on the level of anterior horns of the spinal cord, which was helpful with determination of the final diagnoses.

It seems that on the basis of a detailed diagnostic analysis of the situation of the presented patients, one can regard a part of the symptoms of medullar radices dysfunction found in them as an integral element of amyotrophic lateral sclerosis, whereas another part — as a consequence of vertebral-radicular conflict existing earlier.

On the basis of observations made in our Clinic it should be stressed that in patients with the symptoms of the extensor muscles paresis of the foot or of feet and with concomitant dyscopathies one should always consider the possibility of the initial form of the motoneuron disesase, and in diagnostic procedures, according to the degree of difficulties with specifying the diagnosis one should evaluate the whole of neurological state of the patient and take into consideration electrophysiological examinations, contrast examination of the spinal canal (its lower part or the whole), examination of cerebrospinal fluid, and in some situations — also tomographic examination of the spinal canal or examination of the magnetic resonanse. It is worth remembering that the clinical picture of other diseases of the nervous system (the syndrome of the superior aperture of the thorax, cervical myelopathy, neuropathy, myasthenic syndromes and the so-called mild fasciculation, can approximate the one found in the course of amyotrophic lateral sclerosis.

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STRESZCZENIE

Przedmiotem obserwacji było troje chorych z rzekomopolineuropatyczną, strzałkową postacią stwardnienia zanikowego bocznego. U każdego z przedstawionych chorych obserwowano uprzednio zmiany o charakterze dyskopatycznym w lędźwiowym odcinku kręgosłupa oraz dolegliwości bólowe o typie korzeniowym z pojawieniem się osłabienia mięśni zaopatrywanych przez nerw strzałkowy. Na podstawie przeprowadzonych badań trudno jednoznacznie ustalić, czy mamy do czynienia ze współistnieniem stwardnienia zanikowego bocznego i zespołów korzeniowych lędźwiowo-krzyżowych w przebiegu zmian kręgosłupowych, czy obecność tych objawów jest elementem tego schorzenia. Należy podkreślić, że u pacjentów z objawami niedowładu prostowników stopy należy rozważać możliwość obecności choroby motoneuronu.