ON THE CLINICAL PICTURE OF CEREBELLAR ATROPHIES

P. Hubenov, S. Geneva, R. Hubenov, L. Haverova

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The description of cerebellar atrophies goes back quite a long way. Pierre Marie suggested in 1893 the incorporation into the term «cerebral hereditary ataxiae» of certain number of diseases which differ clinically from Friedreich's disease because of intact or even increased tendon and periosseal reflexes. Spinal cord is mainly affected in Friedreich's disease while cerebellar atrophy dominates in the rest diseases. However, this concept was later on reevaluated because these diseases often were characterized by spino-cerebellar phenomenology. Together with that pathologo-anatomically, a combined lesion of myelon and cerebellum is established. K. Henner et al. (1961) designate these diseases as spino-cerebellar hereditary degenerations with a view to unified nosology. Recently, one can speak about a group of spino-cerebellar degenerations. There are also other pathological forms where cerebellar atrophy comes to the fore.

The aim of the present work is to demonstrate some of these nosological units illustrated with clinical observations.

Olivo-ponto-cerebellar atrophy

J. Dejerine and André Thomas described the main clinical manifestations in 1900. Pathologo-anatomically, there was an atrophy of basal parts of pons, degeneration of its nuclear cellular structures and transversal fibres forming pontine brachia. Atrophic process involved lower oliva with supervening olivo-cerebellar fibre degeneration. Cerebellar cortex was relatively less affected. This enabled the distinguishing between the olivo-ponto-cerebellar atrophy itself and the rest cerebellar atrophies. Some authors (3, 6, 9) outlined the importance of hereditary factors in the etiology. In single cases there was a simultaneous lesion of the cerebellum and other structural nervous formations. Welte (cited after 4) suggested the term spino-ponto-cerebellar degeneration to distinguish these forms. Some investigators (2, 3) described patients with extrapyramidal signs. An illustration in this relation we presented by the following observation:

V. N. M., age 45, c. r. No. 25 518/1982 with diagnosis: olivo-ponto-cerebellar atrophy with extrapyramidal signs.

The onset of the disease was 6—7 years ago with slightly-expressed speech disorder to which a gradually increasing staggering in an indefinite direction was added. Gait became almost impossible and speech unclear, hardly understandable. Neurologically, there was head and tongue tremor, Negro's symptom (static and locomotor ataxia), bilateral intention tremor. Retinal examinations did not show any deviations from the normal picture. CAT revealed a slight internal and external hydrocephaly. There was low-voltage bioelectrical activity, with desynchronization, normal reactivity during functional testing without hemispheric asymmetry and localized pathological focus. The treatment performed did not cause any tendency towards improvement.

This decription showed that extrapyramidal symptomatics could be included into the clinical picture of the olivo-ponto-cerebellar atrophy. H. Rosenhagen (1949) observed a similar phenomenology in half of the cases. J. Barciano (1982) described in 42 per cent of the cases extrapyramidal symptoms and in 27 per cent they were with neostriary character. Commonly, extrapyramidal signs occurred during the course of the illness as in our case. P. Castaigne et al. (1965) reported a patient with initial paleostriary signs of the disease. According to C. Popp and J. Gruner (1962) extrapyramidal symptoms could dominate in the clinical picture of the disease with certain cases. The illness had a progressive course and remissions were almost absolutely absent (10). Initial manifestations of the disease appeared with our patients at the age of 40 years.

Cerebello-olivary atrophy

It was first decribed by G. Holmes in 1907. According to J. Greenfield (1954) the illness began after the age of 40 years. Balance disturbances came to the fore followed by upper limb incoordination. Pathologo-anatomically, cerebellar cortex atrophy, as well as atrophy of the upper part of vermis and inferior oliva predominated.

The following chinical observation illustrated the picture of the disease.

R. I. D., age 58, c. r. No. 7424/1983 with clinical diagnosis cerebello-olivary atrophy with outlined posterior tract syndrome.

The disease started in 1982 with arm and leg formication. Soon a gradually increasing staggering in an indefinite direction was added. Neurologically, there was quadrihyporeflexia stronglier expressed in the legs, disorders of articular-muscular sense, dysmetry during knee-foot test and partially concerning the arms with slightly marked intention tremor. Bilateral ataxia during walking increased when visual control was excluded. Incidentally, choreoathetotic hyperkinesiae were observed. There was 54 mg % protein and 5/3 cells in the cerebrospinal fluid. A Wassermann test was negative. REG revealed data concerning decrease of the elasticity of the cerebral vessels and a slightly expressed pulse deficit in the left side. However, there was no tendency towards disappearing of the neurological symptomatics after the treatment carried out.

The late onset of the disease argued for this diagnosis together with correlation of cerebellar symptoms and signs of posterior tracts and partially of the extrapyramidal system as well as with negative Wassermann test in blood and CSF. P. Castaigne et al. (1965) described a similar case without incidental striary manifestations. Pathologo-anatomically, there were alterations mainly in the upper part of the cerebellar vermis, in the lower oliva and posterior tracts of the spinal cord — predominantly in Gall's fascicles.

Late cortical cerebellar atrophy

It was first described by P. Marie, C. Foix and T. Alajouanine in 1922. K. Henner reported in 1935 a female patient aged 42 years with lamellar cerebellar atrophy. In the next 10 years an associated cerebral and posterior-tract symptomatics advanced in this patient. O. Stary (1950) confirmed this pathological form by using of pneumoencephalography in one case. The atrophic process involved mainly the upper part of the cerebellum. Both anterior and superior parts of the vermis and neighbouring hemispheric regions were most severely affected.

In general, this localization measured up to paleocerebellum. Histologically, there were alterations in almost all layers of the cerebellar cortex with their distinct prevalence in Purkinje's cells. A retrograde atrophy of lower oliva was also established. Most probably, olivo-ponto-cerebelar fibres originated from this region.

The dissociation between upper and lower extremities concerning their damage was considered most typical within the clinical picture. Both statics and gait were severely destroyed while incoordination of the upper extremities was slightlier expressed. Some authors (1, 15) considered not only the genetic predisposition but also the chronic alcohol intoxication factors in the etiology of the disease. However, we could not establish these etiologic factors in our 8 observations. We presented the following case as illustration:

I. P. V., age 41, c. r. No. 1379/1970. His illness started with staggering in an indefinite direction that ceaselessly increased. Then his speech became unclear and hardly understandable. Epileptic fits with unconsciousness were added on this background. Neurologically, a bilateral horizontal nystagmus, slightly delayed active movements, quadrihyperreflexia, well-expressed axial-paraxial reflectory phenomena, slightly expressed arm/hand dysmetry and intention tremor could be established. Gait was severely destroyed of paleocerebellar type. Speech was dysarthric and hardly understandable. EEG demonstrated slight diffuse changes without any epileptic activity. During hospitalization in the neurological clinic no epileptic fits could be observed.

The late development of cerebellar symptomatics with prevalence of the paleocerebellar syndrome allowed us to consider this case a late cortical cerebellar atrophy. To these atrophies some other conditions belong, too, e. g. paraneoplasmatic cerebellar atrophy, congenital atrophy of the granular layer, and dentorubral atrophy.

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К КЛИНИКЕ АТРОФИЙ МОЗЖЕЧКА

П. Хубенов, С. Генева, Р. Хубенов, Л. Хавезова

РЕЗЮМЕ

В настоящей работе рассматриваются клиническая и патологоанатомическая картины атрофий мозжечка. Описываются случаи оливо-понто-церебеллярной, церебелло-оливарной и поздней кортикальной атрофий мозжечка. Проведенными наблюдениями подтверждаются малочисленные литературные данные в связи с тем, что одновременно с церебеллярной лезией могут быть затронуты и другие нервные структурные образования, как экстрапирамидальная система и задние столбики спинного мозга.