CURRENT PROBLEMS OF SEMIOTIC - ANALYTICAL DIAGNOSIS IN THE EXTRAPYRAMIDAL DYSKINESIAS

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SUMMARY:
We present three cases that indicate different neostriat syndromes in the context of the current discussions related to the clinical interrelationships between the extrapyramidal syndromes on one hand and the recent physiology data of the basic ganglions on the other hand. The existence of some types of transition between the main striate syndromes imposes an analytical reporting of the diagnosis, such as the athetosis – dystonic one, or the ballistic – choreic one, avoiding synthetic reporting such as neostriat syndrome or panstriat one. It is important for a more correct evaluation of the extrapyramidal syndromes’ evolution.

Key - Words: striate syndrome, extrapiramidal dyskinesia, basal ganglions.

Involuntary movements are not only semiotic attribute of extrapyramidal syndromes, dyskinesias also occur in other motor disorders (for example fasciculation occur in lesions with chronic evolution of the peripheral motor neurons, convulsions located in the irritation of the central motor neurons in the cortex. In addition, we know consecutive involuntary movements of the distress of the sensitive – sensory systems (for example pseudoathetosis that occurs consecutively to deep sensitivity troubles, superior facial paraspasm – blepharospasm in then irritation of the trigeminal nerve – especially in the irritation of the painful terminations of the ophthalmic branch in keratoconjunctival diseases), then the nystagmus of the amblyopic people is described.

The classical analytical neurological semiology stated a clear classification of the involuntary movements that occur at the level of the innervated muscle by the spinal nerves according to amplitude, rapidity, stereotypes or variability, coordination level, continuity or discontinuity in spurs, rhythmicity, kinetically conditions (kinematics – dynamic) of occurrence – rest, posture or movement.

So, we discern: different types of tremors, chorea movements, pseudoathetosis, balism, twitches, myoclonus, myorhythmias, fasciculation and myokymia, convulsions, dystonia.

The involuntary movements of the irritated muscles by the cranial nerves are not coded so well. First of all we must note the peculiar specificity of the involuntary movements of the eyeballs, always called under the generic term of “paranystagmic movements”. They
occur, the most often, in the brainstem disorders associated or not with cerebral disorders (ocular bobbing, opsoclonia, chaotic movements of the eyeballs, floating movements of the eyeballs, etc). At the level of the irritated muscles by the other cranial nerves, the involuntary movements are framed in the classification of the above mentioned spinal nerves but anyway, there are some mentions related, especially, to the small size of the motor units at the level of the cranial nerves (faced muscle miochemistry, face hemi-spasm, myorhythmia of the palatine veil).

This paper aims to discuss especially the analytical diagnosis problems (clinical sign diagnostic) of the extrapyramidal dyskinesias. This discussion is actual due to two independent reasons.

The first reason is closely related to the occurrence of the imaging paradigm and can be summarized as it follows: the more modest results of the MRI in the extrapyramidal troubles compared to other brain trouble categories (cerebral – vascular pathologies, cerebral tumors, dismetabolic diseases, etc.). Consequently, the clinical semiology has a huge importance.

The second reason is intrinsic semiotic . In other sections of semiology it was possible to make a quite rigorous selection of the indispensable neurological signs or, at least, useful, after a hierarchy according to some definite semiotic criteria: semantic, syntactic and pragmatic criteria. For example, out of about 15-20 tendon reflex described we analyze today only 5-6 reflexes, and out of the Babinski sign equivalents, at least 12, we examine at most 4. In a global perspective, the analyze of the myoarthrokinetic and painful sensitivity is essential, while the analyze tactile and vibratory sensitivity is secondary, and that of the thermal sensitivity is analyzed, in detail, in certain situations (for example in multiple sclerosis).

The situation is different when it comes to involuntary movements, as any of them must be considered a basic symptom, or at least, a main symptom. The pragmatic doctrine of the basic symptoms that has at least the theoretic defect of the confusion symptom – syndrome has the greatest value (partially theoretical too) in extrapyramidal dyskinesias.

There are certain types of involuntary movements that are specific to the damage of certain structures: chorea for the neostriate lesions, resting tremor for paleostriate lesions, athetosis for strio-palido-talamic lesions, torsion dystonia for the putamen lesions. There are obvious extrapyramidal involvements in the occurrence of certain involuntary movements classified in the twitch category of myoclonia torticollis, facial paraspm, professional cramp.

In the current dyskinesias semiology there is the problem of the phenomenological superposition of some extrapyramidal dyskinesias:

1) superposition athetosis – dystonia;
2) superposition athetosis – chorea;
3) superposition athetosis – chorea – dystonia;
4) chorea – dystonia;
5) chorea – balism.

Despite some restrictive definitions of the types of dyskinesias there are also some transit situations. We aim to analyze the meaning of these situations, both from a bibliographic point of view and from the point of view of the Neurology Clinic Experience.

1). Superposition athetosis – dystonia of limbs (in the literature there are detailed descriptions exclusively at the level of the upper limbs. There is the problem if we have to deal with a transition, or, if it is, in fact, an association. The torsion (contraction of the upper limb followed by relaxation) has obvious similarities with pronation- supination in the athetosis. The existence of the clonic movements (fingers’ winces in athetosis) is due to the distal muscle softness. Pseudo-athetotic fast clonic movements (disordered winces) are excluded quite easy from the myoarthrokinetic sensitivity analyze.

It is interesting to mention in the literature as maximum frequency cause of the superior limb dystonia of the post-encephalitic states, states incriminated also in the relatively little elaborated tics of the cephalic extremities (pursing up lips, tongue lapping, “getting free” movements from the neck tie).

The patient H.B., 62 years old, is transferred from neuro-surgery after the evacuation of a left chronic sub-dural hematoma with a very mild right hemiparesis in the Neuro-surgery Clinic, in order to be motoric recovered. During the recovery period (practically spontaneous), 8 days long, in the third day of hospitalization, he developed a torsion distonia of the superior right limb, with approximately 38 hours duration. The phenomena is interpreted as circulatory disturbance in the ensemble of the lenticular nucleus (as a “steal” from the profound silvian territory to the superficial one, on an incipient lacunar basis of extra-pyramidal type (confirmed by MR), in the conditions of the brain’s no-expansionism. For this is also pleading the absence of some important focal lesions.

2). Superposition athetosis - chorea is the most known and started the expression “correa – athetosis movements”. Although they have a completely different degenerative evolution (Huntington), the chronic correa
as well as the acute Sydenham correae are bringing arguments for the association of some dis-kinesia with different morphologic-functional substrate. The debut of both affections with correic movements, involving only the neo-striate, the later apparition of the athetosis movements in both situations, the later being real pan-striate movements, is pleading for a qualitative difference of the two dyskinesia. The subsequent evolution, although totally different, is also pleading for the totally different essence of the two movements. In the regressive evolution of the acute correa, the athetosis movements are disappearing and distal choreic movements of the pseudo-atetosis differentiable fingers are persisting, by examining the mio-artro-kinetic sensitivity. On the other side, the aggravation of the chronic correa is leading to two distinct phenomena: the apparition of the lenticular-type distonia and of the a-kinesia of stria-nigric-type.

3). The superposition of atetosis – chorea-distonia is, with no doubt, the most complex kinetic phenomena. Before theoretically speaking about the issue, it is worth mentioning the second case, the patient S.K., 31 years old, hospitalized for distonia of congenital torsion. This coexists with correic phenomena and athetosis. The interesting anamnestic aspect is that, while distonia and athesosis are congenital, correic movements are relatively recently installed, 4 years ago, after a “flu” episode. This episode has to be interpreted as encephalitis, very plausible interpretation in the context of the apparition of new extra-pyramidal phenomena, on an aftereffect basis. This case is pleading for the distinction between correae — athetosis and for the global suffering of the lenticular nucleus in the torsion distonia, confirming the first two situations, previously mentioned.

4). The superposition correa — distonia appears in the kinetics — phenomenological description of some cases of torticolis, evidently of central extra-pyramidal nature.

5). The phenomenological superposition ballism — correa has a different nature. Under an etio-pathogenic aspect, the hemiballism is due to a small infarct, to a small haemorrhage, sometimes to a metastasis. The hemiballism of vascular pathology is constantly remitted, passing through a correa phase which is initially very strong and is diminishing later. We are exemplifying this situation with a case hospitalized in the Clinic of Neurology, J.S., 72 years old, Male, with long history of high blood pressure and cerebrally lacunar, is hospitalized for right hemibalism. This is transformed in typical hemi-correa after 48 hours. Case 3

The topographic lesion substrate, totally different, of the hemiballism (sub-thalamic body of Luys) is imposing a revision of the two circuits cortical — stria — pallial — thalamic — cortical. While the exciting circuit is directly connecting the pallium with the thalamus, the indirect circuit is accomplishing it using the sub-thalamus. As a consequence, in the lesion of sub-thalamus, the inhibitor circuit becomes not functional. In chorea, the lesion of the striate is determining the lesion of both circuits, the lesion of the inhibiting circuit being those which are determining the hyperkinetic image, and the lesion of the exciting circuit is diminishing the hyperkinetic clinical image of correa, in comparison with the image of hemiballism. The regression of hemiballism at hemi-chorea can’t be explained only by a functional readaptation of the exciting circuit. (11)

CONCLUSIONS

1. The kinetic similitude of athetosis and dystonia represents a reflection of the lesion substrate polytopic (stria- pallium — thalamic). – Case 1
2. The phenomenological superposition correae — athetosis is mostly a kinetic average of two different dis-kinesia, cinematic — dynamic, with mono, respective poly-topic lesion substrate.
3. The correa — distonia superposing is not sufficiently documented, insufficiently knowing the lesion substrate of torticolis.
4. The phenomenological superposing athetosis — distonia — correa is logically reductive to the first two conclusions, being also sustained by Case 2
5. The correa — ballism superposing is done constantly diachronic in a sole evolutive direction, from ballism to correa and is illustrated by case 3, being a typical example for this.
6. From a practical point of view, it is better to avoid the most general label of neo-striate syndrome, respectively pan-striate, the diagnosis of correic, athetosis, (hemi) ballic, distonia, hemiballic — choreic transition or association athetosis - dystonia syndrome being preferred. This is important for a better evaluation of the extra-pyramidal illnesses and syndromes evolution.
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