204. Chorea-acanthocytosis (degenerative disease of the basal ganglia with acanthocytosis)

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Several cases have been reported of an hereditary neurological disease with acanthocytosis since Levine et al.'s description in 1968. This is a report of 3 patients with the same disorder, with pathological study in 1 autopsy case. Two patients were brothers of second cousin parents and the other was a sporadic female patient. Onset of symptoms was in the late teens to early twenties with choreatic movements involving the oro-linguo-facial region and limbs. Other clinical features included muscular hypotonia, loss of deep tendon reflexes and amyotrophy, but no intellectual impairment. Laboratory data revealed 10-20% acanthocytes in peripheral blood, elevated CPK, slight hemolysis and slightly impaired liver function. Hypoxanthine guanine phosphoribosyl transferase (HGPRT) and β-lipoprotein in serum were normal. Biopsy studies of muscle and sural nerve showed peripheral nerve lesion. Computerized tomograms of the brain revealed caudate atrophy. Enzyme, protein and lipid analyses of erythrocytes membrane were within normal limits but a ratio of long fatty acid (24:1/24:0) was low in 2 brothers. Neuropathological examination in the autopsy case revealed neuronal loss of the caudate nuclei and chemical analyses of basal ganglia and cerebral cortex showed normal glutamic acid decarboxylase, choline acetyltransferase and HGPRT.

205. Clinical experience with modified Met-Enkephalin in choreatic movement disorders

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Twenty patients with Huntington's chorea and other choreatic movement disorders including drug-induced dyskinesia received oral treatment with modified Met-Enkephalin. Dosage was between 50 and 150 mg/day. Clinical evaluation was based on video recordings of the patients as well as various motoric and psychological tests. Positive results could be observed in patients with Huntington's disease while dyskinesias were not influenced significantly.

206. Gilles de la Tourette syndrome (TS), its clinical and pathophysiological consideration

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Eighty-eight (77 male, 11 female) patients with multiple tics lasting over 1 yr were subjected to the study to discuss the pathophysiology of TS from the viewpoint of developmental neuropsychology. Neurological and psychological abnormalities were analyzed in contrast to the stage of the illness and age of occurrence. The effectiveness of dopamine antagonists and agonists were evaluated. Mean age of onset was 6 yr 5 mos. Initial symptoms consisted of facial and neck tics, followed by extremities and truncal involuntary movements. Vocal tics including coprolalia tended to occur several years after the onset. Psychological problems were observed in 1/3 of the cases; behaving 'spoiled' in preschool and 'lack of attention' at school age. In 1/4, posture abnormalities with muscle hypotonia and in 2/3 soft neurological signs were observed. Haloperidol and pimozide were effective for motor tics (70–80%); however, haloperidol was more effective for vocal tics. Low dose of L-dopa was effective for a female TS with dystonic posture. These clinical characteristics suggest that the central catecholamine balances have somatotopical and age-dependent specificity. Different effects between 2 DA antagonists and effect of L-dopa on a case suggested that the multiplicity of catecholamine dysbalance and postsynaptic supersensitivity play roles as pathophysiology of TS.

207. Progressive supranuclear palsy. Classification and identification of stage of progress

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The disease known as progressive supranuclear palsy was established in 1964 by Steele, Olszewski and Richardson. Many studies have been conducted since. The authors have examined 4 such cases. Two of them had heredity factors of some extrapyramidal symptoms. From observing these cases it appears necessary to follow the stage of progress of all symptoms and it is useful to use the method outlining the arrangement of symptoms. We divided these into 3 stages: eye symptoms, retrocollis and dementia were commonly observed in every case. From our observations and previous literature, the extrapyramidal symptoms showed a high degree of variability. We classified this disease into: (1) parkinsonian type, (2) ataxic type, and (3) common type. CT, EEG, and eye symptoms were investigated in each of the 3 subtypes and stages. Lastly, we thought that many patients showed many eye symptoms,

ABSTRACTS
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