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Video Journal of Movement Disorders

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Video Journal of MD　総説 Review 1

自発運動の増加と低下を規定する神経伝達物質の作用 ～全身投与時の作用から～

野元正弘 [1)]

要旨

パーキンソン病モデル動物の自発運動に対する作用からドパミン、セロトニン、アセチルコリンの作用を概説した。治療では全身投与となるため、処方された薬は複数部位に分布する受容体に作用する。目的とする治療部位での作用は効果として認識され、他の部位での作用は一般に副作用として表現される。Levodopaはパーキンソン病の治療に用いられており、ドパミン神経の運動増加作用はD2受容体への作用とされ、D1受容体作用薬は全身投与すると自発運動を抑制する。同時にドパミン神経は睡眠、情動、嘔吐等にも関与しており、levodopa治療に伴う眠気、衝動、嘔気の誘発にも関わっている。非選択的セロトニン、アセチルコリン作用薬を脳内へ投与すると自発運動を抑制する。複数の薬理作用を考慮に入れておくことは、より適切な治療の手がかりになると考えられる。

Effects of neurotransmitters including dopamine, serotonin and acetylcholine on locomotor activity in animals - From the perspective of antiparkinsonian medications -

Masahiro Nomoto [1)]

Abstract

Many drugs acting on receptors of dopaminergic, serotonergic or cholinergic neurons have been used for the treatment of neurological disorders. This paper reviews the effects of these agents on the locomotor activity in animals.

Dopamine receptor agonists increase locomotor activities, and this effect has been applied to the screening of antiparkinsonian agents. D2 receptor agonists were developed for the treatment of Parkinson's disease. D1 receptor agonists have been developed, tried in model animals, and then tested in clinical trials for Parkinson's disease. They had, however, no beneficial effects in clinical treatments. Although D1 dopaminergic agents enhance locomotion by acting on the direct pathway at the striatum, they also inhibit locomotor activity by affecting the prefrontal cortex.

Even though serotonergic neurons increase or decrease locomotor activity depending on the receptor subtype, serotonergic activation eventually decreases movements as a whole. Cholinergic neurons stimulate muscarinic and nicotinic receptors. Anticholinergic agents have been used for the treatment of Parkinson's disease for a long time through the action on muscarinic receptors in the brain. Anti-choline esterase agents which increase the activity of cholinergic neurons in the brain decrease locomotor activity in animals.

A neurotransmitter agent usually has multiple actions through acting on several types of receptors. Basic knowledge on the mechanisms of action of the agents used in therapy should help physicians treat patients with movement disorders.

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Movement Disorders Review

Video Journal of MD　総説 Review 2

抗IgLON5抗体関連疾患

大野陽哉 [1)]、伏屋公晴 [1)]、木村暁夫 [1)]、下畑享良 [1)]

要旨

抗IgLON5抗体関連疾患は、2014年に閉塞性睡眠時無呼吸症候群とパラソムニアを呈する疾患として初めて報告された。その後多数の病型が報告され、2021年には①睡眠障害型、②球麻痺型、③運動異常症型、④認知機能障害型、⑤神経筋障害型に分類された。これらの症候は進行性核上性麻痺や大脳皮質基底核症候群、筋萎縮性側索硬化症などに類似することもある。ビデオ終夜ポリソムノグラフィーは本症の睡眠障害の特定に有用である。病理学的には、異常リン酸化された3リピート・4リピートタウの神経細胞への沈着を認める。早期の免疫療法開始が予後を改善しうるため、不随意運動に加え睡眠障害を伴うなど非典型例では本症を疑う必要がある。

Anti-IgLON5 disease

Ono Y [1)] , Fuseya K [1)] , Kimura A [1)] , Shimohata T [1)]

Abstract

Anti-IgLON5 disease was first reported in 2014 by Sabater et al. as a sleep disorder manifesting obstructive sleep apnea and parasomnia. Since then, many clinical features have been reported. Gaig et al. classified the clinical phenotypes into 5 subtypes: sleep disorders, bulbar syndrome, movement disorders, cognitive impairment, and neuromuscular manifestations. Characteristic sleep disorders are parasomnias and respiratory symptoms（stridor and sleep apnea）. Bulbar syndrome includes dysarthria, dysphagia, sialorrhea, central hypoventilation, vocal cord paralysis, and stridor. As neuromuscular manifestations, fasciculations in the tongue and limb muscles, muscle weakness, muscle atrophy and cramps can be observed. These bulbar and neuromuscular features may mimic amyotrophic lateral sclerosis and stiff-person syndrome. Most patients with anti-IgLON5 disease present with many motor symptoms, with an average of three per patient. The symptoms include facial/abdominal dyskinesias, cerebellar ataxia, chorea, and parkinsonism. The disease may mimic progressive supranuclear palsy or corticobasal syndrome in some cases. The patients can have various cognitive dysfunctions such as verbal or visual memory disturbance, difficulty in verbal fluency, executive dysfunction, and visuoconstructive performance disturbances. Video polysomnography is useful for detecting undifferentiated NREM sleep and poorly structured N2 sleep, which are diagnostic of anti-IgLON5 diseases. Previous studies showed a strong association between HLA-DRB1*10:01/HLA-DQB1*05:01 alleles and anti-IgLON5 disease. Neuropathological studies revealed hyperphosphorylated tau deposits（3-repeat and 4-repeat）along with neuronal cell loss and gliosis. Anti-IgLON5 disease should be considered in patients with atypical features such as movement disorders combined with sleep disturbance, because this disorder is partially treatable.

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Video Journal of MD 原著 Original Article 1

著しいdystonic tremorにBotulinum toxinが奏効したspinocerebellar ataxia type 2（SCA2）の1例

武井麻子 [1)]

要旨

SCA2の発症17年目に著しい痙性斜頸とdystonic tremorを認め、botullinum toxin type A（BoNT-A）が著効を奏した1例を経験したので報告する。BoNT-A80単位施行前後に、痙性斜頸と振戦をModified Tsui scale for cervical dystonia（Tsui scale）、とabnormal involuntary movement scale（AIMS）および表面筋電図により評価した。治療1週後にTsui scoreは14点から5点に、AIMSは24点から10点に減少し、表面筋電図上の律動波が消失した。小脳―視床―皮質経路や基底核はdystonic tremorの責任病巣として報告されているが、これらの部位はSCA2の障害部位としても報告されており、本症例でもこれらの領域に障害が拡大したと考えた。

The effect of botulinum toxin type A on spasmodic torticollis and dystonic tremor in a spinocerebellar ataxia type 2（SCA2）patient -A case report –

Asako Takei [1)]

Abstract

We report a patient with marked cervical dystonia（CD）and dystonic tremor that appeared 17 years after the onset of spinocerebellar ataxia type 2（SCA2）. We evaluated the CD and tremor before and after injection of 80 units of BoNT-A, using the Modified Tsui Scale for cervical dystonia（Tsui scale）, the abnormal involuntary movement scale（AIMS）and a surface electromyogram. One week after treatment, the Tsui score decreased from 14 points to 5 points, and AIMS showed a reduction from 24 points to 10 points. In addition, rhythmic discharges on the surface EMG disappeared. It has been reported that the cerebello - thalamo - cortical circuit and the basal ganglia are involved in the production of dystonic tremor, and that these structures are also involved in SCA2. BoNT-A injection may have beneficial effects on these disorders.

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Movement Disorders Original Article

Video Journal of MD 原著 Original Article 2

視床下核の小梗塞により発症したhemiballismの1例

越智智佳子 [1)]*、西川典子 [2)]**、安藤利奈 [2)]、野元正弘 [1)]

要旨

74歳男性におけるhemiballismの1症例を、動画を用いて報告した．右上下肢の違和感に引き続いて、激しい右上下肢のhemiballismを発症し日常生活を障害していたが、ドパミン受容体拮抗薬により軽減し消失した。Hemiballismは病変が小さいため、画像で特定するのは困難な例も多い。本例では、頭部MRI前額断像により左視床下核の病変を描出できた。

A case of hemiballism induced by a hypothalamic infarction detected by contrast-enhanced MRI

Chikako Ochi [1)]*, Noriko Nishikawa [2)]**, Rina Ando [2)] , Masahiro Nomoto [1)]

Abstract

A case of hemiballism caused by a contralateral hypothalamic infarction was presented. A 69-year-old man was referred to our neurology clinic. He sensed something unusual in his right arm and leg, and developed abnormal movements a few days later. He manifested ballism in his right arm and leg throughout the day, but not during sleep. One mg of clonazepam or 75 mg of tiapride a day had no beneficial effects. The hemiballism was relieved by treatment with 1.5 mg of haloperidol and 50 mg of sulpiride a day. Brain MRI with contrast enhancement showed a small infarction at the left hypothalamus on the frontal plane. Haloperidol was stopped 6 months later and sulpiride was discontinued one and a half year later without recurrence of involuntary movements.

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Movement Disorders Original Article

Video Journal of MD 原著 Original Article 3

リチウム中毒により舞踏運動をきたした1例

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要旨

症例は76歳男性。体のゆれ、歩行時のふらつきを主訴に受診した。以前より炭酸リチウムを内服しており、副作用なく経過していた。心不全に対してサイアザイド系利尿薬の内服を開始したが、その1カ月後より舞踏運動を生じ徐々に悪化した。四肢、体幹に左右差のない舞踏運動を認め、歩行は失調性であった。リチウム血中濃度は基準値以内であったがサイアザイド系利尿薬開始後に上昇を認め、炭酸リチウムの減量およびサイアザイド系利尿薬中止によりリチウム血中濃度は低下し症状が改善したことから、リチウム中毒と診断した。リチウム中毒での舞踏運動の報告は少ないが、併用薬によりリチウム血中濃度の上昇をきたし、基準値以内であっても舞踏運動を生じる可能性のあることを念頭においておく必要がある。

A case of chorea induced by lithium intoxication

Yuki Yamanishi [1)] , Noriyuki Miyaue [1)] , Satoshi Tada [1)] , Rina Ando [1)] , Masahiro Nagai [1)]

Abstract

A 76-year-old man developed involuntary movement and gait disturbance. He had been taking lithium for 30 years without any adverse effects. One month after he started taking thiazide diuretics to treat his aortic aneurysm, he developed chorea and ataxic gait. Lithium blood level was elevated to 1.76 mEq/L（therapeutic range 0.6-1.2 mEq/L）. Chorea and ataxic gait improved following reduction of lithium dose and cessation of thiazide diuretics. Chorea is a relatively rare involuntary movement caused by lithium intoxication. Clinicians should, however, keep in mind that lithium blood level may increase by interaction with thiazide diuretics and that chorea could develop.

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