Transient alternating hemichorea as presenting sign of progressive supranuclear palsy

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We report the case of a man who developed unilateral alternating chorea four years before presenting a typical clinical form of Steele-Richardson-Olszewski syndrome. It is known that choreic dyskinesias may occur during the course of this syndrome but this is the first account of choreas presenting sign of the syndrome.

Key-Words: Chorea—Steele-Richardson-Olszewski syndrome

Introduction

Progressive supranuclear palsy is a clinical sign that is found in a restricted number of degenerative diseases of the nervous system, the most common of which is Steele-Richardson-Olszewski syndrome (SROS) [6]. Supranuclear ophthalmoplegia, though not always the presenting sign of these conditions, must be observed at some stage during the course of the illness for the diagnosis of SROS to be established [1,3]. We have recently observed the case of a man who developed alternating hemichorea four years before the onset of a typical clinical picture of SROS.

Case report

This male railway worker first developed neurological symptoms in April 1982 at the age of 46. His past medical history revealed only a primary syphilis, which had been treated on its occurrence, when he was 20. Family history was unremarkable. Neurological signs appeared abruptly; they were observed by a university neurologist, who described choreoathetoid movements of the upper and lower limbs on the left side. Involuntary movements were also present during gait, which was otherwise normal. No postural abnormalities were observed; there were no pyramidal or cerebellar signs; sensory examination was also normal; cranial nerves were unaffected. Muscle tone was reduced in both left limbs. Routine laboratory findings were in the normal range, except for a mild hyperlipemia. EEG recording was normal; CT scan revealed mild cortical and subcortical cerebral atrophy. A descriptive diagnosis of left hemichorea was made. The patient was first treated with tiapride (200 mg t.i.d.) for one month, but, as there was no clinical improvement, valproic acid (400 mg t.i.d.) was started.

Dyskinetic movements gradually ceased within a few months. Therapy was withdrawn. In the summer of 1984 choreic movements appeared on the right side. The clinical picture was quite similar to the one described above, but affected the other side. Laboratory examinations (including serology for syphilis) were normal. Neuroleptic treatment (haloperidol 2.5 mg q.i.d.) was then started; it reduced the intensity of choreic movements, although they displayed a fluctuating course throughout the following years. Blepharospasm was first noted during a routine neurological examination in autumn in 1985. In the same period the patient experienced unexplained falls. A CT

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scan, performed in December 1985, showed no focal lesions, but revealed a mild atrophy of the cerebral cortex and a slight enlargement of both lateral ventricles. In January 1986 dyskinetic movements were almost undetectable; therefore haloperidol was withdrawn.

In November 1986, when he was admitted to our department, the patient complained of sudden falls and of dyskinetic movements on the right side. Neurological examination revealed the presence of right hemichorea with reduced muscle tone in the right limbs. No cerebellar, pyramidal or somatosensory signs were detected. Muscle jerks were normal on both sides. Posture was normal on standing and walking, but impairment of the righting reflex was revealed by the pull test. Blepharospasm and eye opening apraxia were observed. Extraocular motility was impaired both in the horizontal and vertical planes. Neuropsychological examination revealed a diffuse attentional deficit with abnormal scores on tests for frontal lobe functions; a mild impairment on visual and verbal recall; performance on constructive praxia was normal. Routine analyses were within the normal range. EEG recording showed theta activity in the frontal-temporal regions of both hemispheres. A CT scan confirmed the presence of mild cortical and subcortical atrophy (Fig. 1). A diagnosis of progressive supranuclear palsy with hemichorea was made; choreic movements were successfully treated with haloperidol (2.5 mg t.i.d.) and with tiapride (100 mg b.i.d.).

In July 1987 the patient was readmitted to our department due to the onset of facial dyskinesias, which were thought to be side-effects of the neuroleptic treatment. Choreic movements were not seen. Facial dyskinesias improved with the administration of trihexyphenidyl (1 mg b.i.d.). The patient was dismissed in August 1987. He was readmitted in December 1987 due to the occurrence of sudden falls. Ocular signs were then prominent, including supranuclear opthalmoplegia in all directions (including convergence) and severe eye opening apraxia with secondary conjunctivitis. To open his eyes, the patient had to rub them before pulling the upper eyelid with his finger. Other signs included: axial dystonia, postural instability with retropulsion, impaired gait, reduced blinking, bradykinesia, severe dysarthria with palilalia, hypotonia on the right side. After discharge in January 1988 the patient was seen several times in the outpatient clinic. No dyskinesias were observed and so neuroleptic therapy was discontinued in April 1989. Since then, the patient has been taking no drugs; his clinical condition has progressively worsened but choreic movements have not reappeared.

Discussion

Although histological examination is the only reliable criterion for the diagnosis of SROS, clinical observation may provide data which are useful for an *in vivo* provisional diagnosis. Recently, Golbe et al. [3] set strict diagnostic criteria for a clinical diagnosis of SROS. The clinical picture of our patient meets all of them and so it may be assumed that he has SROS. Abnormal involuntary movements are not a common feature of this syndrome; they were not mentioned in the original nine cases reported by Steele, Richardson and Olszewski [11], who only observed "some athetoid posturing of the hands in case 3". However, subsequent case reports have documented the occurrence of rest tremor, myoclonus, chorea and dystonia in patients with SROS [1,2,4,5,6,7,8,10].

The present case is unique because chorea was

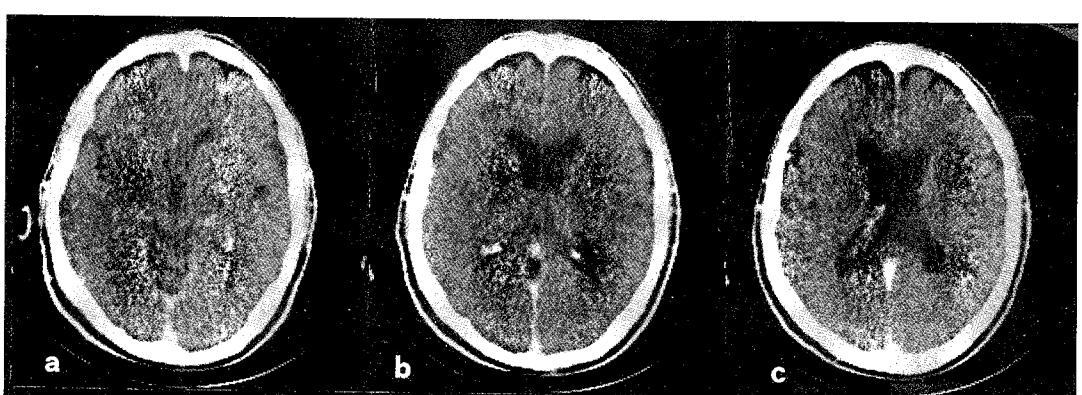

Fig. 1. *CT-scan showing diffuse, mild, cortical and subcortical atrophy. a, b and c are three tomographic sections of a dorsoventral series, which are spaced approximately 15 mm from each other.*

the presenting symptom of the disease, which preceded the onset of supranuclear ophthalmoplegia, by about four years. Further, chorea was always unilateral: at the beginning it appeared on the left side, afterwards on the right. Thus, the clinical appearance of chorea was peculiar both in terms of time and of topography.

A critical analysis of our case prompts the following remarks. First, since progressive supranuclear palsy may be associated with a number of neurological conditions, the existence of diseases other than SROS must be ruled out. In particular, corticobasal degeneration may be associated with abnormal involuntary movements on one side of the body [6,9]. We think it unlikely that this patient has corticobasal degeneration because of (1) the symmetric distribution of cerebral atrophy, as shown by CT-scan (Fig. 1), and (2) the lack of any significant degree of cortical sensory loss or apraxia.

Second, it might be argued that hemichorea on the right side may have been brought about by the neuroleptic therapy started soon after chorea was detected on the left side.

We believe, however, that this is unlikely, because, when hemichorea occurred in the right side, neuroleptic therapy had been discontinued for more than one year.

Sommario

È descritto il caso di un paziente, che ha presentato una sintomatologia coreica unilaterale ed alternante, a cui è seguita dopo quattro anni la comparsa di una tipica sindrome di Steele-Richardson-Olszewski. La presenza di corea è stata già osservata in alcuni pazienti con sindrome di Steele-Richardson-Olszewski. Questo articolo costituisce il primo resoconto di un caso clinico in cui le ipercinesie coreiche hanno costituito il sintomo di esordio della malattia.

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