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Chorea in Behçet's Syndrome

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Summary. Pure chorea has not yet been reported in Behçet's syndrome. The case is presented of a 58-year-old man affected by relapsing choreic dyskinesia associated with a psychotic disorder 31 years after onset of the disease. The possible beneficial effect of ACTH is discussed.

Key words: Behçet's syndrome – Chorea – Psychiatric disorders

Zusammenfassung. Es wird ein Patient mit Behçets Syndrom beschrieben. 31 Jahre nach Krankheitsbeginn trat eine choreatische Hyperkinese auf, die 3 Monate dauerte und mit psychotischen Symptomen kombiniert war. 60 Tage später kam es zu einem Rückfall. Die mögliche therapeutische Wirkung von ACTH wird diskutiert.

Introduction

Neurological involvement is a well-known complication of Behçet's syndrome. Many clinical pictures have been described [1, 2]. Although the basal ganglia is a typical site of neuropathological lesions [3], extrapyramidal signs are rarely reported and only one case with involuntary movements has been described [4]. We present a patient affected by Behçet's syndrome in whom psychotic features and choreic dyskinesia were episodic manifestations of the disease.

Case Report

A 58-year-old man had had recurrent attacks of bilateral uveitis, which had started in the left eye 31 years earlier, after the transitory appearance of aphthous stomatitis. Recurrent fever of unknown origin, myocardial ischemia and ulcerative gastroduodenitis were also reported. No cases of neurological disease were known in his family.

He had been under our care 3 years earlier for unstable gait. Neurological examination revealed ataxic gait, bilateral hypotonus and slight dysarthria. Routine serum analyses were normal, except for a slight increase in ESR (20 mm in 1 h). CT scan showed cortical atrophy.

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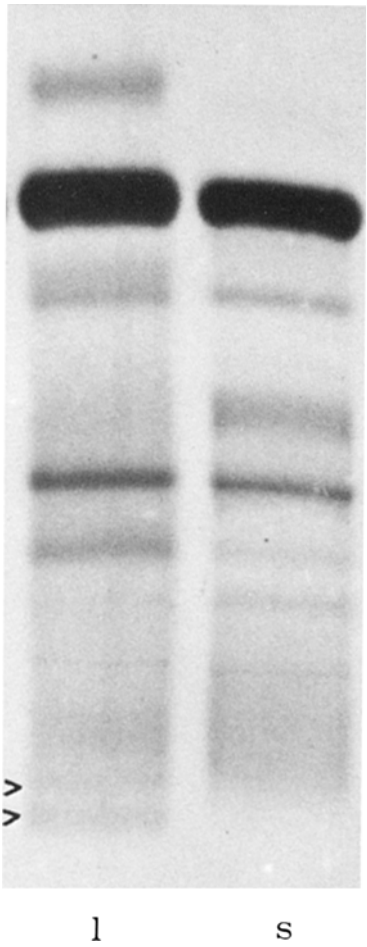

Fig. 1. Electrophoretic pattern on agarose of spinal and serum proteins. Oligoclonal bands are visible in the gamma region, in spinal CSF

At 58 years of age, he was readmitted to our hospital because his wife noticed a deterioration in his motor performances with clumsiness of movement and a tendency to drop objects. The patient appeared anxious, suspicious and restless. There was choreic dyskinesia of the head, shoulders, trunk and especially of the hands, with grimacing and pursing of the mouth and lips. He was unable to hold out his tongue, which moved continuously in his mouth. Ataxic broad-based gait and bilateral hypotonus persisted; weakness of the left arm and increased reflexes on the left were observed.

His IQ of 98 was in the normal range, as was his rating on the Wechsler intelligence scale. Visual acuity was 4/10 for both eyes. Anterior cortical cataracts and opacity of the left corpus vitreum were noticed; the fundi oculi were normal. CT scan sections of the basal ganglia did not show any abnormality.

The tuberculin skin test (5 TU) was negative. Routine serum analyses were normal, as were C-reactive protein, mucoproteins, TAS, plasma fibrinogen, IgG, IgA and IgM. Serum immune complexes were increased (23%; normal value less than 5%) using the C1q binding method.

The CSF contained normal protein and glucose levels and 28 lymphocytes/ml. IgG (6 mg%; normal 0.8–3.5 mg%), IgG to albumin ratio (33.6%; normal 22%), the Link index (0.82; normal less than 0.70), and IgG intrathecal synthesis (9.05 mg/day) were increased. Oligoclonal bands were visible in the gamma region (Fig. 1).

An almost complete regression of the dyskinesia was observed 9 days after pimozide (4 mg/day) and ACTH (25 U/day) therapy. Pimozide was continued, but the choreic movements relapsed 2 months later, associated with hostility, obsessive trends and phobias; ACTH treatment (25 U/day) was again given. One month later the patient appeared well, quiet and cooperative, without mood and thought disturbances. No involuntary movements were present.

Discussion

According to Lehner's criteria [5], this patient was affected by Behçet's syndrome. The "minor symptoms" of recurrent fever, gastroduodenitis and psychiatric disturbances in this case are well-known manifestations of the disease [3, 6]. On the other hand, extrapyramidal syndromes are rarely reported; these are essentially parkinsonian-like with bradykinesia, expressionless face, stiffness and tremor [7].

To the best of our knowledge, only one case of hyperkinesia has been reported in Behçet's syndrome [4]. In that case the patient showed facial grimacing and choreo-athetoid movements, which disappeared completely in 2 days.

In our case a similar picture of dyskinesia occurred in association with mental disturbance, initially of a neurotic type, eventually evolving into a full psychosis. No athetoid movements were present. The chorea was almost generalized with severe impairment of motor performances. It lasted 3 months and, interestingly, remitted after a few days of pimozide/ACTH therapy. After 2 months of only pimozide treatment a relapse was observed, which remitted after the reintroduction of ACTH.

The reason for this relapse could be a spontaneous evolution of the disease, insufficient therapy or the withdrawal of a possible beneficial effect of ACTH in both the relapses.

A beneficial effect of steroid therapy has been reported [1, 8] and a better prognosis of the Behçet's patients has been correlated with the introduction of this treatment [3].

The etiology of the disease is still unknown, but a hypothetically slow virus, possibly neurotropic, has been suspected [9]. The presence of a high spinal level of immunoglobulins and IgG [10] and the finding, in our case, of oligoclonal bands with intrathecal IgG production support this hypothesis. This suggests persistent activity of the pathological agent within the central nervous system, as in subacute panencephalitis and possibly in multiple sclerosis [11].

References

1. O'Duffy JD, Goldstein NP (1976) Neurologic involvement in seven patients with Behçet's disease. Am J Med 61:170–178
2. Wolf SM, Scotland DL, Phillips LL (1965) Involvement of nervous system in Behçet's syndrome. Arch Neurol 12:315–325
3. Alemà G (1970) Behçet's disease. In: Vinken PJ, Bruyn GW (eds) Handbook of clinical neurology. North-Holland Publishing Company, Amsterdam, cp 24, pp 475–512
4. Scotland DL, Wolf SM, White HH, Dubin HV (1963) Neurologic aspects of Behçet's disease. Am J Med 43:544–553
5. Lehner T, Barnes CG (1979) Criteria for diagnosis and classification of Behçet's syndrome. Clinical and immunological features. Academic Press, London, pp 1–9

6. Epstein RS, Cummings NA, Sherwood EB, Bergsma DR (1970) Psychiatric aspects of Behçet's syndrome. J Psychosomatic Res 14:161–169
7. Pallis CA, Fudge BJ (1956) The neurological complications of Behçet's syndrome. Arch Neurol Psychiat 75:1–14
8. Lessof MH, Jefferys DB, Lehner T, Mattock M, Sanders MD (1979) Corticosteroids and Azathioprine in Behçet's syndrome. In: Lehner T, Barnes CG (eds) Behçet's syndrome. Clinical and immunological features. Academic Press, London, pp 267–275
9. Evans AD, Pallis CA, Spillone JD (1957) Involvement of the nervous system in Behçet's syndrome: report of three cases and isolation of virus. Lancet 272:349–353
10. Motomura S, Tabira T, Kuroiwa Y (1980) A clinical comparative study of multiple sclerosis and neuro-Behçet's syndrome. J Neurol Neurosurg Psychiatry 43:210–213
11. Fraser KB (1977) Multiple sclerosis: A virus disease? Br Med Bull 33(1):34–39

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