Arq Neuropsiquiatr 1993, 51(2):267-269

CHOREA AS A SIGN OF SYSTEMIC LUPUS ERYTHEMATOSUS ACTIVITY

CASE REPORT

P. CARAMELLI*, S.M. TOLEDO*, P.E. MARCHIORI**, E.R. BARBOSA***, M. SCAFF****

SUMMARY — Chorea may occur as a neurological manifestation of systemic lupus erythematosus and is often associated with detection of antiphospholipid antibodies. No evidence of chorea as a sign of lupus activity has been established. We describe a patient with systemic lupus erythematosus associated with antiphospholipid antibodies who developed chorea, which has been considered a sign of lupus activity.

KEY WORDS: chorea, systemic lupus erythematosus, lupus activity.

Coréia como sinal de atividade do lupus eritematoso sistêmico: relato de caso.

RESUMO — Coréia pode ocorrer como manifestação neurológica do lupus eritematoso sistêmico, estando frequentemente associada à presença de anticorpos antifosfolípides. Evidência de coréia como sinal de atividade lúpica não foi descrita. Descrevemos o caso de uma paciente, com lupus eritematoso sistêmico e anticorpos antifosfolípides, que desenvolveu coréia, sendo esta manifestação considerada sinal de atividade da doença.

PALAVRAS-CHAVE: coréia, lupus eritematoso sistêmico, lupus em atividade.

Chorea is a rare but well recognized neurological manifestation of systemic lupus erythematosus (SLE). It occurs in less than 2% of patients with SLE and can be the first evidence of the disease [1,9]. Previous reports emphasized the relationship between chorea in SLE and antiphospholipid antibodies, but the pathogenesis of this association is still poorly understood [1,7]. These antibodies might be related to direct damage to the basal ganglia [1]. Evidence lacks that could establish a close relationship between chorea and SLE activity.

We describe the case of a patient with SLE in whom chorea was an early manifestation of the disease and has been considered a probable sign of SLE activity.

CASE REPORT

LAL, a 15 year old white female was admitted to our Hospital with a history of personality changes and memory impairment in the last four years. Episodes of polyarthralgia were also mentioned. Six months after the onset of symptoms involuntary (choreic) movements appeared first affecting her right hand, and after a few months involving the four limbs, mouth and tongue. This clinical picture progressed slowly, with deterioration of cognitive

Departamento de Neurologia, Faculdade de Medicina da Universidade de São Paulo (FMUSP), São Paulo: * Médico Pós-Graduando; ** Professor Associado; *** Médico Assistente; **** Professor Titular. Aceite: 11-setembro-1992.

Dr. Paulo Caramelli — Divisão de Neurologia, Hospital das Clínicas, FMUSP — Caixa Postal 8091 — 01065-970 São Paulo SP — Brasil.

functions. Haloperidol (3mg daily) has been prescribed for control of the choreic movements with partial response. One year later the patient developed an epileptic partial crisis and phenobarbital was started. No more seizures were observed.

Physical examination revealed a mild hypertension (180x100mmHg). Neurological evaluation disclosed: moderate dementia (Mini-Mental Status 21/30); choreic movements involving the four limbs, and also mouth and tongue. There were no motor deficits and the deep tendon reflexes were globally hyperactive. Laboratory findings included mild anemia (Hb 10,9g/dl) with normal WBC count and thrombocytopenia (48x109/L), normal erythrocyte sedimentation rate and positive serum autoantibodies: antinuclear antibody (>1/200), lupus anticoagulant (APTT 78,4 seconds; control 50,0 seconds) and anticardiolipin (IgG 106 U). Computerized tomography (CT) of the head showed a moderate cortical atrophy which was more prominent in the left peri-sylvian area.

Immunosuppresive therapy with prednisone (60mg daily) and azathioprine (150mg daily) was started. Propranolol (80mg a day) was also prescribed for hypertension. A good clinical response was obtained with complete control of the chorea even after discontinuation of haloperidol. At discharge she was receiving prednisone, azathioprine, propranolol and phenobarbital (100mg a day). After a ten-month follow-up period at the Outpatient Unit the patient remained clinically stable. An attempt to reduce the immunosuppressive dosages resulted in relapse of symptoms (choreic movements). Laboratory investigation yielded no sign of systemic activity. However, restoration of previous doses of the same immunosuppressive agents permitted again control of chorea.

COMMENTS

The association between chorea and SLE is well established as has been pointed out by several reports [1,4,6,7,9]. Unfortunately, the pathogenesis of this occurrence is still unknown. Neuropathological investigations failed to demonstrate lesions in brain areas which may be related to the clinical picture of chorea [9]. Nevertheless, owing both to the thrombotic tendency of these patients [2] and to observations that chorea can disappear spontaneously and reappear in different localization in the same patient, it has been suggested that reversible ischemia could be occurring [3]. Absence of any abnormality in the basal ganglia region on CT scans and MR imagings carried out during episodes of chorea supports this hypothesis [1,7].

Antiphospholipid antibodies include lupus anticoagulant and anticardiolipin antibodies as well as those accounting for false-positive tests for syphilis. Several clinical manifestations have been associated with the presence of these antibodies, including thrombosis, thrombocytopenia, recurrent abortion and some neurological disordes, specially chorea [2,3,8]. Asherson et al. described twelve patients with chorea and SLE, nine of which with positive serological tests for anticardiolipin, antibodies and lupus anticoagulant [1]. This association suggests that these antibodies could cause direct damage to the basal ganglia by binding phospholipid structures in this region [1]. A possible mechanism that has been postulated is that these antibodies might bind phospholipids in the endothelial cell membrane resulting in blockade of arachidonic acid release and reduction in prostacyclin production, thus increasing platelet aggregation and thrombosis [5].

However, a recent report suggests that the occurrence of antiphospholipid antibodies in immune diseases of the nervous system could be just an immune epiphenomenon. Marchiori et al. studied 81 patients with multiple sclerosis, Guillain-Barré syndrome and SLE detecting significant levels of antiphospholipid antibodies (specially anticardiolipin) in many of them [10].

Our patient has major neurological manifestations associated with SLE, namely chorea, dementia and seizures. It could be assumed that it is a predominantly cerebral form of the disease. The presence of antiphospholipid antibodies confirms the tendency of their detection in SLE cases with chorea.

No close relationship between SLE activity and chorea has been established. The laboratory data of our case do not show evidence of systemic activity of the disease, but based on the criteria of lupus activity described by Urowitz et al. the disease should be considered as active [11]. Furthermore, both the good response obtained with immunosuppressive treatment and the recurrence of

symptoms with dose reduction point to a possible liaison between chorea and SLE activity. In our opinion, although observed in an isolated case, chorea may be considered to occur not only as a manifestation of SLE but also as a sign of SLE activity.

REFERENCES

1. Asherson RA, Derksen RHWM, Harris EN, Bouma BN, Gharavi AE, Kater L, Hughes GRV. Chorea in systemic lupus erythematosus and "lupus-like" disease: association with antiphospholipid antibodies. Semin Arthritis Rheum 1987, 16:253-259.
2. Asherson RA, Harris EN. Anticardiolipin antibodies: clinical associations. Postgrad Med J 1986, 62:1081-1087.
3. Bingley PJ, Hoffbrand BI. Antiphospholipid antibody syndrome: a review. J R Soc Med 1987, 80:445-448.
4. Bouchez B, Arnott G, Hatron PY, Wattel A, Devulder B. Chorée et lupus érythémateux disséminé avec anticoagulant circulant: trois cas. Rev Neurol 1985, 141:571-577.
6. Carreras LO, Vermylen JG. Lupus anticoagulant and thrombosis: possible role of inhibition of prostacyclin formation. Thromb Haemost 1982, 48:28-40.
6. Greenhouse AH. On chorea, lupus erythematosus, and cerebral arteristis. Arch Intern Med 1966, 117:389-393.
7. Kamashta MA, Gil A, Anciones B, Lavilla P, Valencia ME, Pintado V, Vázquez JJ. Chorea in systemic lupus erythematosus: association with antiphospholipid antibodies. Ann Rheum Dis 1988, 47:681-683.
8. Levine SR, Welch KMA. The spectrum of neurologic disease associated with antiphospholipid antibodies. Arch Neurol 1987, 44:876-883.
9. Lusins JO, Szilagyi PA. Clinical features of chorea associated with systemic lupus erythematosus. Am J Med 1975, 58:857-861.
10. Marchiori PE, Reis M, Quevedo MEZ, Callegaro D, Hirata MTA, Scaff M, Oliveira RM. Cerebrospinal fluid and serum antiphospholipid antibodies in multiple sclerosis, Guillain-Barre syndrome and systemic lupus erythematosus. Arq Neur quiat (São Paulo) 1990, 48:465-468.
11. Urowitz, MG, Gladman DD, Tozman ECS, Goldsmith CH. The lupus activity criteria count (LACC). J Rheumatol 1984, 11:783-787.