Writer's cramp as a clinically isolated syndrome

K Agan, N Afsar, T Tanridag, D Ince-Gunal

Citation

K Agan, N Afsar, T Tanridag, D Ince-Gunal. *Writer's cramp as a clinically isolated syndrome*. The Internet Journal of Neurology. 2007 Volume 9 Number 1.

Abstract

We present a case of writer's cramp in a patient whom cranial T2 weighted MRI showed lesions in the lentiform nucleus, putamen and the internal capsule. Following the extensive work-up for the differential diagnosis of multiple sclerosis, the patient was defined as clinically isolated syndrome. The patient's symptoms resolved after intravenous methylprednisolone treatment. To our knowledge this is the first case of writer's cramp with clinically isolated syndrome in the literature.

INTRODUCTION

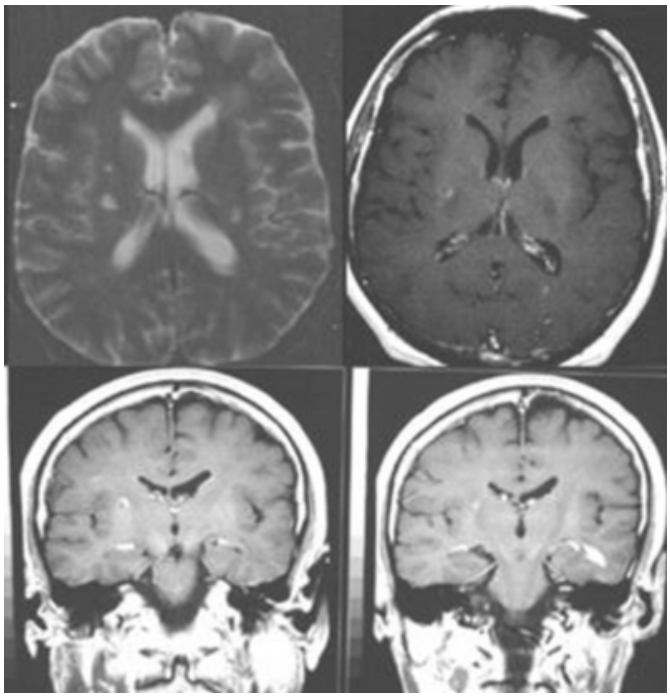
Clinically isolated syndrome (CIS) was defined as an acute or subacute episode affecting one region of the CNS especially the optic nerves, brain stem or spinal cord that is presumed to be demyelinating, with no previous history of possible demyelinating event [1, 2]. Approximately 90% of patients with multiple sclerosis (MS) initially present with CIS [2]. In most instances symptoms and signs indicate a lesion in the optic nerve, spinal cord or brain stem in an order of decreasing frequency [1,3]. Tremor, tonic spasm, ballism, palatal and other types of myocloni, dystonia, and parkinsonism due to demyelinating lesions in critical regions are known to result in such movement disorders. Movement disorders other than tremor, associated with MS occur infrequently [4]. The association of most cases of dystonia including writer's cramp (WC) with MS was thought as coincidental. Two patients described in the literature with WC have the diagnosis of laboratory supported definite MS according to the Poser's criteria [4]. The present case with WC was discussed as CIS.

CASE REPORT

A 45-year-old man presented with writer's cramp that appeared one year ago. Initial neurological examination revealed bilateral slight postural and kinetic tremors in addition to writer's cramp which has been associated with dystonic posturing of elbow, thumb and index finger. The cranial MRI showed multiple hyperintense lesions on T2-weighted sequences (figure 1) in periventricular and subcortical white matter (lentiform nucleus, putamen and the internal capsule), subsequently enhancing on T1-weighted sequences (figure 1).

Figure 1

Figure 1



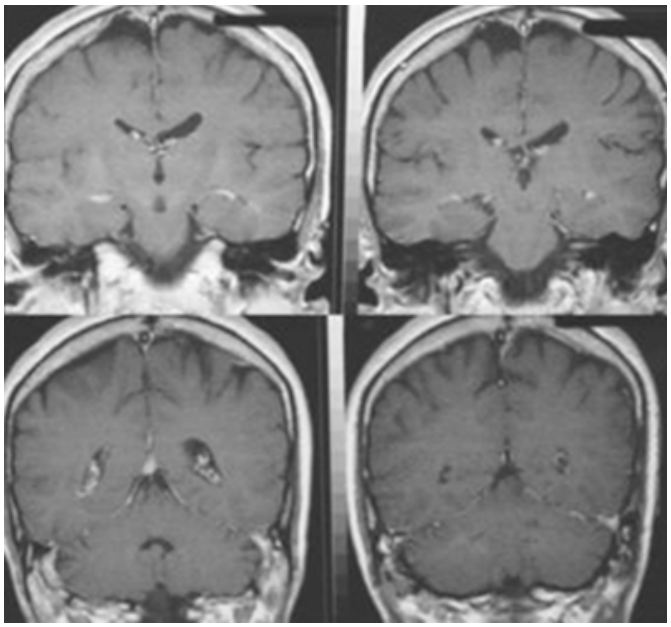
In the differential diagnosis vasculitis (Behçet's disease, antiphospholipid antibody syndrome), vitamin B12 deficiency, neuroacanthocytosis, thyroid diseases, syphilis and acquired immune deficiency syndrome were excluded. Analysis of the cerebrospinal fluid revealed normal immunoglobulin-G synthesis and negative oligoclonal bands. The writer's cramp of the patient resolved gradually during treatment with intravenous methylprednisolone (1000mg/day for 5 days).

The present case was discussed as CIS since the symptom

and MRI findings (figure 2) of the patient resolved completely following steroid treatment and no other etiology including vasculitis, infectious disease and metabolic disorders could be determined.

Figure 2

Figure 2



DISCUSSION

Approximately 90% of patients with MS initially present with an acute CIS [2]. Movement disorders are not the most common types of (CIS) like optic neuritis, brain stem and spinal cord syndromes. However in clinically definite MS, a variety of movement disorders, namely tremor, dystonias, chorea and parkinsonism have been described and were thought to be secondary to demyelinating lesions in critical regions [4, 5, 6, 7]. Since the subcortical gray matter also contains myelinated nerve fibers, striatum, pallidum and thalamus can be affected by demyelinating inflammatory process [4]. Sixteen cases of dystonia associated with MS have been found in the literature, among which two WC cases were present in laboratory supported definite MS patients [4]. Only one of these cases has MRI data presenting with thalamic and basal ganglia plaques [4]. The literature may support the hypothesis that writer's cramp could be a presenting symptom in MS patients and perhaps could be considered as a form of CIS.

Our case with a history of WC for 2 years had MRI findings supporting MS according to Fazekas criteria [8]. However, there are two defined paraclinical indicators associated with an increased risk of progression to MS. One of them is MRI abnormalities and the other is the presence of CSF oligoclonal bands [9]. In the present case, there were no oligoclonal band in the CSF. Resolution of WC and disappearance of contrast enhancing lesions on MRI at the third month following pulse methylprednisolone treatment support the diagnosis of clinically isolated demyelinating syndrome. Moreover, other conditions causing MS-like MRI changes have excluded with an extensive differential diagnosis.

Writer's cramp as a segmental dystonia has an unknown etiology and pathologic substrate. However, few studies have pointed out the cerebellum as a target neuroanatomic structure [10]. This is why for the present case, it is impossible to point out a direct relationship between demyelinating lesions and WC, thus a coincidental association can not be ruled out. In other way, such as unusual clinical course, WC might be the presenting and isolated manifestation of MS. Only the follow up of the patient and the occurrence of a second attack will lead to the diagnosis of MS.

References

1. O'Riordan J. I., Thompson A. J., Kingsley D. P. E, MacManus D. G., Kendall B. E., Rudge P., McDonald W. I., Miller D. H. The prognostic value of brain MRI in clinically isolated syndromes of the CNS: A 10-year follow-up. Brain 1998; 121: 495- 503.
2. Dalton CM, Brex PA, Miszkiel KA, Hickman SJ, MacManus DG, Plant GT, Thompson AJ, Miller DH. Application of the new McDonald criteria to patients with clinically isolated syndromes suggestive of multiple sclerosis. Annals of Neurology 2002; 52: 47-53.
3. Sailer M., O'Riordan J. I., Thompson A. J., Kingsley D. P., MacManus D. G., McDonald W. I., Miller D. H. Quantitative MRI in patients with clinically isolated syndromes suggestive of demyelination. Neurology 1999; 52 : 599-606.
4. Tranchant C, Bhatia KP, Marsden CD. Movement disorders in Multiple Sclerosis. Movement disorders 1995; 10: 418- 423.
5. Minegar AR, Sheremata WA, Weiner WJ. Transient movement disorders and multiple sclerosis. Parkinsonism and Related Disorders 2002; 9: 111-113.
6. Azzimondi G, Rinaldi R, Liguori R, Tonon C, Martini E, D'Alessandro R. Dystonia as an isolated symptom of multiple sclerosis ? Italian Journal of Neurological Sciences 1997; 18: 301- 302.
7. Waubant E, Alize P, Tourbah A, Agid Y. Paroxsymal dystonia (tonic spasm) in Multiple Sclerosis. Neurology 2001; 57: 2320-2321.
8. Fazekas F, Offenbacher H, Fuchs S, Schmidt R, Niederkorn K, Horner S, Lechner H. Criteria for an increased specificity of MRI interpretation in elderly subjects with suspected multiple sclerosis. Neurology 1998; 38: 1822-1825.
9. Tintore M, Rovira MJ, Martinez MJ, Rio J, Diaz-Villoslada P, Brieva L, Borras C, Grive E, Capellades J, Montalban X. Isolated demyelinating syndromes: Comparison of different MR Imaging criteria to predict

conversion to clinically definite multiple sclerosis. American Journal of Neuroradiology 2000; 21: 702-706.
10. Comella C, Ludlow C, Hallett M. Focal dystonias; blepharospasm, cervical dystonia, spasmodic dysphonia, writer's cramp. In: Noseworthy JH (ed) Neurological therapeutics. Principles and practice. London: Martin Dunitz, 2003: 2527-2534.

Author Information

Kadriye Agan, M.D.
Department of Neurology, Marmara University School of Medicine

Nazire Afsar, M.D.
Assoc. Prof., Department of Neurology, Marmara University School of Medicine

Tulin Tanridag, M.D.
Prof., Department of Neurology, Marmara University School of Medicine

Dilek Ince-Gunal, M.D.
Assoc. Prof.,Clinic deputy-chief, Department of Neurology, Marmara University School of Medicine