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# Cervical dystonia as first manifestation of multiple sclerosis

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Sirs: Cervical dystonia is very rare in patients with multiple sclerosis. A case is reported in which cervical dystonia was the initial manifestation of multiple sclerosis. Although a simple coincidence of multiple sclerosis and cervical dystonia cannot be excluded, epidemiological data and pathophysiological considerations make a causal correlation more likely. Multiple sclerosis as the cause of secondary cervical dystonia may probably be underrecognized.

Typical signs and symptoms of the first manifestation of multiple sclerosis (MS) include sensory disturbances, motor paresis, optic neuritis, and eye motility disorders [5]. Sustained dystonia, unlike paroxysmal movement disorders [1], is considered an extremely rare manifestation of MS [6, 9, 10, 13, 15]. We describe a patient in whom cervical dystonia was the *presenting* feature of MS.

A 38-year old woman of Italian origin presented with a ten month history of cervical dystonia with combined leftward rotation of 70 degrees and rightward tilt of 40 degrees, partially relieved by touching the left cheek, but persisting during sleep. Transitory improvement had been achieved with tiapride and biperiden. Three days before admission she noted loss of right retroauricular sensation. On admission, she was taking biperiden 12 mg/d, tizanidine 6 mg/d, and clonazepam 1.5 mg/d for the dystonia, as well as citalopram 20 mg/d for panic attacks. She had no history of exposure to neuroleptic drugs prior to onset of dystonia, drug or alcohol addiction, perinatal asphyxia, trauma, arterial hypertension or diabetes. Her family history did not reveal any patients with movement disorders or psychiatric diseases.

Neurological examination showed 12 points on the Tsui rating scale [14] for cervical dystonia, and hypesthesia behind and below the right ear, most likely corresponding to segments C2 and C3, but no other pathological signs. Slit-lamp examination excluded Kayser-Fleischer rings. Blood tests showed normal results for cell counts, electrolytes, liver and renal function, thyroid stimulating hormone, vitamin B 12, and ceruloplasmin. Enzyme-immuno-assays for herpes simplex virus, tick borne encephalitis, treponema, and borrelia were negative. Cell count ($3.0 \times 10^9$ (reference $< 4.8 \times 10^9$)) and total protein content (354 mg/l (reference < 480 mg/l)) of the cerebrospinal fluid were normal, but intrathecal IgG production was shown by the presence of four oligoclonal bands and an elevated IgG Index (13.6, reference < 7.0). MRI of the cervical spine showed two non-enhancing lesions right dorso-laterally at levels C3/4 and C2/3 (Fig. 1A). Cranial MRI revealed two right-hemispheric ovoid lesions in periventricular (Fig. 1B and C) and juxtacortical areas (Fig. 1D). Visual evoked potentials of both eyes were within normal limits, but the side difference was abnormal (Fig. 2A). Somato-sensory evoked potentials recorded after stimulation of the left arm showed a markedly reduced amplitude of the potentials recorded from the scalp (Figs. 2B and C).

A diagnosis of laboratory supported definite MS (according to Poser's criteria) or of MS (according to the new diagnostic criteria by McDonald et al. [8], i. e., two attacks, clinical evidence of one lesion, three MRI-detected lesions consistent with MS plus positive CSF) was made. The patient improved after injection of botulinum toxin A into the neck muscles combined with pulsed high-dose intravenous methylprednisolone (500 mg daily for five days). She received the last injection of botulinum toxin 12 months after the first presentation. Fifteen and 27 months after the first presentation, she showed only minimal signs of cervical dystonia (both times 3 points on the Tsui scale). Her medication at the last consultation consisted of tizanidine 6 mg/d, alprazolam 1 mg/d, paroxetine 40 mg/d, and mianserine 30 mg/d. Except for tizanidine, the other medications were for psychiatric symptoms.

In most cases of focal dystonia, the cause remains unknown, leading to a diagnosis of primary dystonia. In contrast, some clinical "red flags" were present in our case and pointed to secondary focal dystonia. Sensory deficits in the C2 and C3 dermatomes and persistence of dystonia during sleep were suggestive of secondary focal dystonia. Paraclinical examinations such as imaging, electrophysiological and laboratory testing supported the diagnosis of MS.

We cannot exclude a coincidence of MS and focal dystonia in our case. However, the probability of such a coincidence is extremely low (1: 3.500.000) according to reported prevalences of MS in Italy of 50/100.000 [12], and of cervical dystonia of 57/100.000 [3]. Furthermore, the persistence of cervical dystonia, also during sleep, is a strong argument for secondary,

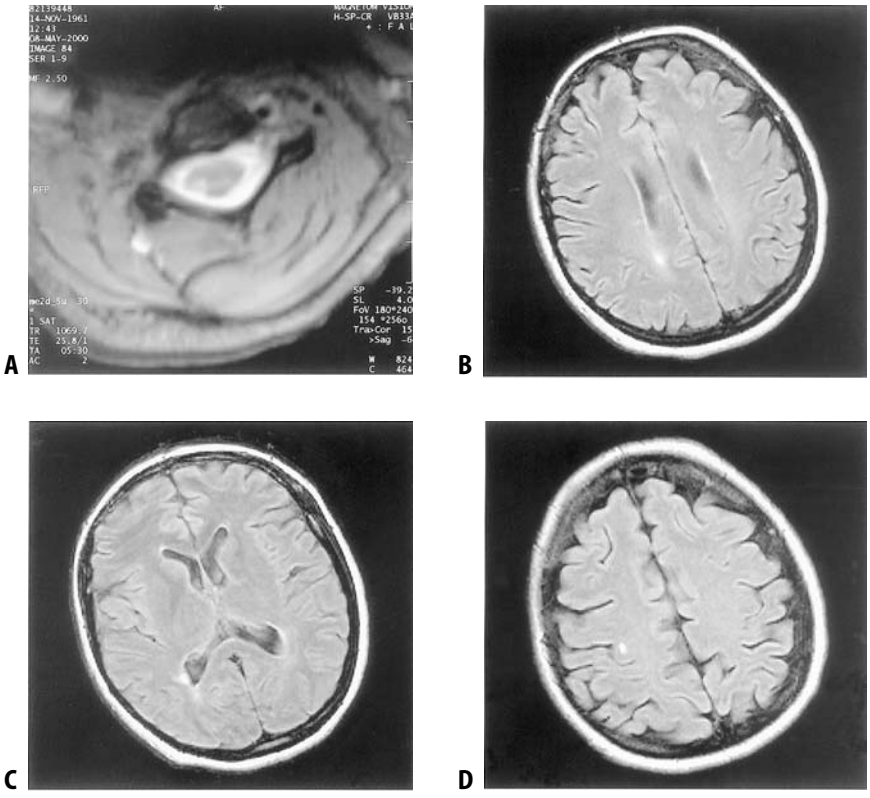

**Fig. 1** **A:** MRI of the cervical spine: T2-weighted axial section at level C3/C4. Hyperintense lesion is present right dorso-laterally. **B–D:** MRI of the head. FLAIR sequences. Right-hemispheric ovoid lesions in periventricular (**B, C**) and juxtacortical location (**D**). Note rightward head tilt due to dystonia. The lesions did not enhance with gadolinium in T1-weighted images

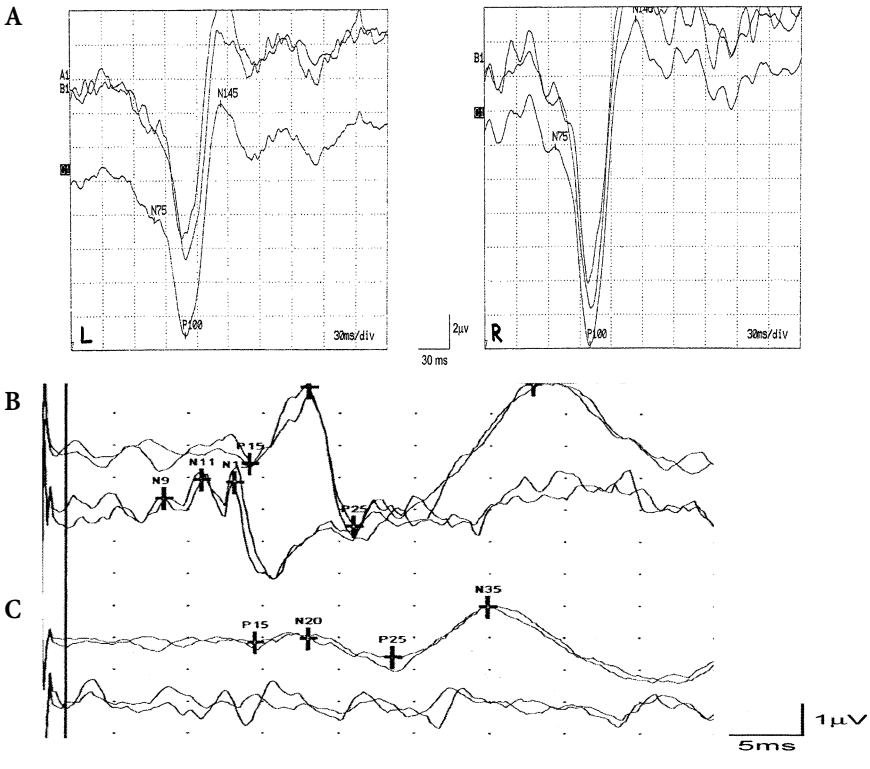


**Fig. 2** **A:** Visual evoked potentials. The latency of component P100 on both sides (right: 99.0 ms; left: 108.0 ms) is within the range of normal values, but the side difference (9.0 ms, normal < 6.0 ms) is pathological, with prolongation of recording after stimulation of the left eye. The upper two traces: averaged curves of 200 recordings each. Lower traces: average of the two curves. L denotes recording after stimulation of the left eye; R, after stimulation of the right eye. **B and C:** Somato-sensory evoked potentials: Recordings after stimulation of the right median nerve at the wrist (**B**); of the left median nerve at the wrist (**C**). The top traces represent the recordings from the scalp, the bottom traces, from the processus spinosus C6. The reference was at $F_z$. Latencies of P15 and N20 are within normal limits, without pathological side differences. Components N9, N11, and N13 after stimulation of the left arm can not be identified (probably for technical reasons in the presence of dystonia); the potentials recorded from the scalp show a markedly reduced amplitude (diminution of amplitude N20-P25 by 86 %; normal < 45 %)

i. e., in this case MS-related, dystonia [11]. Thus, we propose the alteration of central processing produced by an MS lesion as the more likely explanation for the cervical dystonia in the present case [7]. Anecdotal reports describe torticollis resulting from different cervical lesions [2, 4] and the association of spinal MS plaques with torticollis [6] and hand dystonia [15]. However, dystonia was not the first manifestation of MS in any of these reports.

To conclude, MS may probably be an underdiagnosed cause of secondary focal dystonia, and conversely, secondary focal dystonia may herald multiple sclerosis and should be included in the differential diagnosis of patients presenting with this clinical feature.

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