Multiple Sclerosis and Intracranial Hypertension Presenting as Paroxysmal Kinesigenic Dyskinesia

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Abstract

Background: Paroxysmal movements in the form of tonic spasms, dystonia, choreoathetosis and hemidyskinesia have been reported in patient with multiple sclerosis (MS). Intracranial hypertension with headache and papilledema resembling idiopathic intracranial hypertension (IIH) exceptionally occurs in patient with MS. Conversely, IIH may rarely present as movement disorder in children.

Aims of the study: Report an unusual patient with MS and intracranial hypertension anteceded by movement disorder.

Methods: Clinical examinations and ancillary diagnostic testing.

Results: A 54 year old patient presented one year earlier to her diagnosis of MS with symptoms of paroxysmal kinesigenic diskinesia (PKD). Her initial neurologic examination and brain MRI were normal. A year later her examination became abnormal and her MRI showed demyelination. Her cerebrospinal fluid contained oligoclonal bands and the opening pressure was markedly elevated, in the absence of headache and papilledema.

Conclusion: PKD may antecede MS and acephalgic intracranial hypertension without papilledema in exceptional patients.

Introduction

Paroxysmal symptoms in multiple sclerosis (MS) include paresthesia, dysarthria, ataxia, limb pain, diplopia, itching, akinesia, and abnormal movements, as they were detailed in the classic publication of Osterman and Westerberg in 1975 [1]. Tonic seizures of spinal origin were present in five of their twenty two patients, and were characterized by tonic painless contraction of a limb. Subsequently, Libenson, et al, described a patient with left arm and leg tonic spasms secondary to demyelination of the contralateral cerebral peduncle. Hyperventilation induced the spasms but the electroencephalogram (EEG) remained normal during the attacks [2].

Case Report(s)

A 54 year old female was seen initially in neurological consultation because of paroxysmal, painless, repetitive brief involuntary torsion movements of the left hand and left foot, consistently triggered by initiation of volitional movements of the affected limbs. They attacks took place several times a day. Symptoms have begun four month earlier. She had no warnings and no after effects. By the time she was seen in consultation her involuntary movements have become less frequent. She had no history of similar episodes in the past, no family history of neurological disorders and her review of systems was otherwise negative. She had been overweight since adolescence. Her BMR was 49.4 kg/m2. She had mild hypertension treated with hydrochlorothiazide 25 mg a day and varicose veins. Her general physical and neurological examinations were normal. Her CBC and chemistries including potassium levels were normal. Her sedimentation rate, CK levels and ANA titer were normal. She had low vitamin D blood levels. Her brain MRI showed only a congenital small pineal cyst. Her EEG was normal. Symptoms cleared within a few weeks completely. She returned a year later complaining of poor balance, frequent accidental falls, and waddling gait. This time her neurological examination revealed ataxia of gait, dysmetria of her left arm movements, distal weakness of her left leg, increased leg reflexes and a left Babinski sign. She had no papilledema. Her follow up brain MRI showed multiple irregular periventricular and subcortical hemispheric demyelinating lesions highly suggestive of MS, not present a year earlier. There was no hydrocephalus. Her spinal fluid opening pressure was 40 cm of water. Her CSF IgG level was elevated at 80 mg/L and she had 10 IgG oligoclonal bands. Cultures were negative for bacteria and fungi. Subsequent brain MR venography (MRV) showed no thrombi or stenotic lesions to otherwise explain her intracranial hypertension.

Treatment was initiated with glatiramer acetate, vitamin D supplementation and topiramate 50 mg b.i.d., the latter to assist in weight control, in addition to a low calorie diet. No relapses have occurred in follow up visits.
Discussion

Idiopathic intracranial hypertension is typically manifested as headache and visual disturbance due to bilateral papilledema, more commonly encountered in young women undergoing rapid weight gain [3]. Intracranial lesions are absent. Atypical patients with unilateral papilledema and pseudopapilledema have been reported [4]. Conversely, IIH without papilledema is increasingly recognized in individuals with chronic daily headache on whom the elevated intracranial pressure seems to serve as a perpetuating mechanism for their headache [5]. Additional signs and symptoms of IIH are intracranial noises and cranial nerve paresis, none present on this patient. Of relevance to our subject of interest is that not all patients with IIH have headache (“acephalgic IIH”). De Simone, et al, described two patients with IIH associated with papilledema manifested as transient visual obscuration that complained of no headache [6]. Gonzales-Hernandez, et al, on their retrospective case review of fifty-five patients with IIH, found that approximately 15% of their patients had no headache [7]. Elevated intracranial pressure is encountered rarely on individuals with MS. All of the three patients reported by Newman, et al, had headache and papilledema, in addition to signs and symptoms of MS [8]. In David and colleagues, patient, intracranial hypertension was the presenting sign of MS [9]. The latter authors suggested that intracranial hypertension in MS is based on CSF pathway blockage caused by small plaques of demyelination or by associated inflammation [9]. In support to their argument, their patient exhibited some degree of ventricular enlargement on his head computerized tomography scan (head CT). Involuntary movements may exceptionally appear in children with IIH in the form of torticollis, responding to CSF drainage [10]. It is unclear if this patient had a subclinical elevation of CSF pressure when first evaluated for her symptoms of PKD, since a lumbar puncture was not done at the time.

Movement disorders other than tremors are infrequent in MS. Tranchant, et al, described fourteen patients with MS of whom nine had dystonia, three parkinsonism and two had myoclonus [11]. From their extensive review of the pertinent literature they concluded that tremors, ballistic movements, tonic spasms and palatal myoclonus were probably symptoms secondary to the disease process, while parkinsonism, dystonia and myoclonus in patients with MS are probably coincidental [11]. Berger and associates reported eight patients with MS on whom paroxysmal dystonia was the presenting symptom, preceding additional signs of demyelination [12]. The interval between the appearance of paroxysmal movements and other signs of MS ranged from three days to ten years, although in most cases it was two years or less. Paroxysmal kinesigenic choreoathetosis as early symptom of MS was reported by Roos, et al [13]. Subsequently, two patients with paroxysmal dyskinesia as the initial manifestation of MS were published [14]. Demyelinating lesions were identified over the posterior limb of the internal capsule contralateral to the movements on these patients. Dyskinetic movements anteceded MS by five months in their first patient and by one month, in their second patient [14]. The patient herein discussed had PKD as a presenting and reversible symptom of MS one year earlier to the appearance of other symptoms. Her case is also unusual because her first neurological examination was normal and her first brain MRI failed to show demyelinating lesions. Making her case even more unusual, she was found to have intracranial hypertension in the absence of headache, papilledema or hydrocephalus. The intimate pathogenesis of paroxysmal dyskinesias is yet to be defined. There are primary and familial presentations and secondary forms to brain lesions of different locations [15]. Specific demyelinating CNS lesions of predictable location accounting for PKD in MS have not been established. While Roos patients had contralateral internal capsule demyelination, Fragoso, et al, first patient had two spinal cord lesions and the second had a contralateral subcortical parietal lesion and a thalamic lesion [13, 14]. This patient had multiple areas of bilateral subcortical and periventricular hemispheric demyelination that were not present by the time she experienced her movement disorder. The aforementioned anatomic inconsistencies in the above cases led Nociti, et al, to question if movement disorders in MS, other than tremors, simply represent two coinciding diseases, each of frequent appearance, given the abundance in many patients of paraganglionic MS lesions detected by MRI, that result in no apparent clinical manifestations [16]. Of Nociti’s, et al, seven hundred thirty three patients with MS, only twelve, or 1.6%, had movement disorders including, blepharospasm, tourettism, and hemifacial spasms. Only one of their patients had hemidystonia and none had PKD [16].

The significance of obesity (BMI > or =30 kg/m2) in the pathogenesis of MS is unknown. Yet, a large body size at age 20 is associated with 96% increased risk of developing the disease [17].
Conclusion

PKD may antecede MS and acephalgic intracranial hypertension without papilledema in exceptional patients.

References

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