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Progressive Myalgic Encephalomyelitis (ME) or A New Disease? A Case Report

Howes S[1] and Goudsmit EM[2]*

[1]Retired Science Writer, Croydon, UK

[2]Retired Health Psychologist, London, UK

***Corresponding author:** Goudsmit EM, 23 Melbourne Road, Teddington, Middx, TW11 9QX, UK

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Abstract

This is a report on a patient with a history of increasing dizziness and muscle weakness after minimal exertion. In her twenties, the symptoms became more pronounced following glandular fever. After excluding other diseases, her physician diagnosed myalgic encephalomyelitis (ME), now commonly referred to as chronic fatigue syndrome (CFS). The condition followed a relapsing-remitting course until about five years ago, when she experienced a sudden deterioration and developed new symptoms such as blurred vision in one eye and urinary incontinence. Whether this is a case of progressive ME or a new disease remains uncertain. Research is required both to increase diagnostic clarity and to establish whether the recommended behavioural interventions designed for CFS are appropriate for the subset of patients with neurological symptoms and abnormalities on MRI.

Keywords: Progressive myalgic encephalomyelitis; Multiple sclerosis; Vestibular dysfunction

Abbreviations

ASOT: Antistreptococcal antibody titres; CADASIL: Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy; ECG: Electrocardiogram; FODMAP: Fermentable Oligo- Di- Mono-Saccharides and Polyols; CFS: Chronic Fatigue Syndrome; ME: Myalgic Encephalomyelitis; MRI: Magnetic Resonance Imaging; MS: Multiple Sclerosis; MCS: Multiple Chemical Sensitivity.

Case Presentation

The patient is a severely disabled 59 year old female. She was diagnosed with myalgic encephalomyelitis aged 29 as her symptoms were consistent with the descriptions of this illness at the time [1,2]. Four years ago, her GP requested a neurological consult after she had developed additional symptoms, notably weakness in the left leg and transient blurring in the left eye. Moreover, her condition had worsened and she was no longer able to live an independent life.

Her medical history is not typical of ME as she recalled experiencing periods of weakness in her legs, nausea and dizziness intermittently during childhood. When she was 8, her GP considered acute rheumatic fever as her ASO titre was raised (>1200 U/ml) but the patient does not remember red or tender joints, tremors, arthalgia or erythema marginatum. Moreover, her ECG was normal and there were no other signs of cardiac involvement. It is noteworthy, however, that during and after the 6 years of prophylactic penicillin G, she felt slightly better and was almost symptom free for a year. The dizziness returned when she was 15 and her muscle fatiguability became more pronounced at age 17. At 18, she was diagnosed with glandular fever but at the time, few physicians were aware of post-viral fatigue syndrome and as her symptoms persisted, her GP prescribed a course of 2 mg diazepam. This had a beneficial effect on her dizziness and the patient requested repeat prescriptions until she developed a tolerance to the drug. Once aware of the dangers associated with chronic use of benzodiazepines, she reduced the dose over a number of months and was able to stop the medication. She completed her University course and even managed to study for a postgraduate degree.

A magazine article on ME led her to request a consult with a specialist in the field. He determined that she probably suffered from the illness, as did a professor of Immunology at a London medical school. Blood tests revealed raised titres to the Coxsackie B virus.

Around the age of 40, a cold triggered a sudden increase in her dizziness and she became housebound. Betahistine, prochlorperazine, antihistamines and rehabilitation exercises had no appreciable effect. A year later, she was treated for a ductal carcinoma in situ (DCIS) with a lumpectomy followed by a wide excision. This is of note because having recovered from the surgery; she began to notice symptoms when exposed to perfumes and after eating certain foods. This syndrome is now recognised as multiple chemical sensitivity (MCS) [3].

The weakness in her legs and the dizziness were not investigated further but a change to a low gluten, low sugar diet helped her improve to the extent that she was able to go out on short trips and complete a doctoral degree from home. A course of cognitive behaviour therapy helped with nightmares but did not alleviate her somatic symptoms while several attempts to gradually increase activity levels just made her feel more unwell.

The sudden deterioration in her mid-fifties was preceded by an otherwise unremarkable gastro-intestinal upset. This resulted in post-prandial pain in the peri-umbilical area, especially at night, and she lost weight (from 47 kilos to 41 kilos). Other new symptoms included faecal and urinary incontinence, low blood pressure (89/55 mmHg), frequency of micturition, loss of fine motor co-ordination, tremor, paresthesiae in hands and feet, clumsiness, episodes of sweating as well as dry eyes and mouth. Blood tests including a panel for Sjogren's syndrome were normal. Since that time, she has also been diagnosed

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with osteoporosis and cervical spondylosis. Difficulty breathing at rest was evaluated and attributed to muscle weakness.

In the neurology clinic, she was found to have nystagmus indicative of both peripheral and central vestibular dysfunction. Further examination included various scans but none were helpful in establishing a diagnosis. The report on the most recent MRI noted large patches of confluent signal change in cerebral white matter bilaterally. These were most prominent in the posterior frontal regions but also involved the parietal and temporal lobes. The lesions had slightly blurred margins and showed increased diffusion without any haemorrhagic element. There was involvement of deep and periventricular white matter. Cerebral cortex and juxtacortical white matter were spared. The corpus callosum was not involved. The basal ganglia, brainstem and cerebellum were also spared. There was no mass effect or atrophy. The lesions were larger and more prominent than the year before but similar in overall distribution. Conclusion: progressive white matter changes. According to the neuroradiologist, "this is not the appearance of ordinary small vessel disease, MS or CADASIL". A second opinion at another centre of excellence was also unable to associate the abnormalities with a known disease.

The initial diagnosis of ME is supported by the presence of:

1. Muscle fatigue following minimal exertion and prolonged recovery lasting up to five days.

2. Evidence of CNS involvement, e.g. dizziness and vertigo, not explained by other factors e.g. menopause, inner ear infection etc.

3. Problems with circulation e.g. heat intolerance, feeling cold when it's hot outside; symptoms worsen after hot bath, cold extremities.

As descriptions of this illness note, the symptoms typically fluctuate and all tend to worsen after exertion. The course can be stable, relapsing/remitting or progressive, although there is a paucity of literature about the latter.

Most cases of ME tend to start as an unremarkable viral infection, which may be accompanied by myalgia, lymphadenopathy, or a gastro-intestinal upset [1,4]. However, instead of recovering, patients begin to experience profound fatigue following activities which were previously completed without difficulty. Also typical is a prolonged delay in the restoration of muscle power [5]. This is sometimes described as post-exertional fatigue and while this is a minor criterion for the diagnosis of chronic fatigue syndrome (CFS), it is considered a cardinal feature of ME [1,6,7]. The assumption of equivalence, i.e. that ME is an older term for the condition now referred to as CFS, has been challenged [2] but the current consensus is that most of the differences between the diagnostic categories of ME, CFS and ME/CFS reflect the variance between the case definitions as well as the inclusion and exclusion criteria used in research [8].

Based primarily on the history and findings, we propose that the patient's current illness represents a progressive form of ME. Knowledge of this and other cases suggests that its main characteristics are:

1. A worsening of existing neurological symptoms or new symptoms, e.g. blurred vision in one eye, weakness in one leg, incontinence.

2. A sudden increase in sensitivities and gastro-intestinal symptoms.

3. Any improvements are limited and the disability tends to show a downwards trend.

4. The patient has to spend more time at home or in bed.

5. The development of new auto-immune diseases or symptoms suggestive of the latter, particularly Sjögren's syndrome.

Supportive evidence of pathology:

Abnormal white matter lesions on MRI which are not considered incidental or age-related, or indicative of MS.

Given the lack of a definite diagnosis and her doctors' limited experience treating individuals with progressive ME, the patient is currently managing her illness on her own with advice from the MS Society and other individuals with similar symptoms. She is pacing her activities [9] and has regular physiotherapy for pain in her neck and back. She uses daily preservative-free eye gel for her dry eyes. The post-prandial pain resolved on the FODMAP diet [10] although the weight loss continued, despite an increased intake of carbohydrates and meat. She continues to suffer from an irritable bladder and urinary incontinence which are being investigated. The faecal incontinence improved following the complete removal of gluten from her diet. At the moment, her condition has stabilised but as she is no longer able to tolerate oral antibiotics or the supplements prescribed for osteoporosis, the patient is aware that she is at increased risk of additional symptomatology in the future. She has asked for supportive counselling to help her cope but her local health authority does not offer this service to the housebound.

Discussion

The patient's symptoms and particularly, their close association with exertion, support a diagnosis of ME [1,2]. While the presence of muscle fatiguability and dizziness at an early age could have various other causes, her family history revealed that her mother had been forced to stop work as a result of a condition resembling ME. There may therefore be a hereditary factor which predisposed the patient to the illness. The relationship between infections such as glandular fever and the exacerbation of her symptoms is also consistent with ME.

The sudden deterioration after many years and the presence of symptoms resembling MS have been documented by support groups and online but as far as we aware, this is the first report focusing on progressive ME in the scientific literature. Further study is essential to determine the extent of the differences between this subset and the wider population with CFS and to examine whether the recommended interventions for the latter are as effective for the former.

Of particular interest are the MS-like symptoms. This aspect of ME has been largely overlooked despite the work of the renowned MS specialist Charles Poser [11]. In one article, he compared and contrasted the two disorders and noted that individuals with MS are more likely to demonstrate signs of disease such as nystagmus, cerebellar incoordination, ataxia, spasticity, hyperreflexia, abnormal plantar responses and sensory disturbances, principally of vibration and position senses. He emphasized that like MS; 'chronic post viral

fatigue syndrome' is associated with post-exertional worsening, paresthesiae and dizziness. However, in his view, the parasthesiae tend to be of a different type: in the latter, they often have a burning, painful component and a migratory characteristic. "Such sensory complaints are unusual in MS." Moreover, any lesions found on MRI are periventricular in location, rather than the cortex. Also of significance is that individuals with postviral syndrome rarely have specific complaints such as diplopia, incoordination and true gait ataxia, nor do they exhibit abnormalities such as nystagmus, sustained clonus or Babinski signs.

According to Poser, the similarities between the two disorders have undoubtedly resulted in misdiagnosis as well as inappropriate treatment. Indeed, he estimated that in his practice, 22% of 366 referrals with presumed MS actually fulfilled the criteria for, and probably had CFS [12].

Although there is growing interest in the structure and function of the brain in patients with CFS, the findings on MRI have been equivocal and difficult to interpret. One reason is the changing concept of the illness, which began as a post-infectious syndrome and is now hard to differentiate from neurasthenia [13]. In one of the older studies, Schwartz et al. evaluated 16 individuals with strictly-defined CFS and found abnormalities in 8 [14]. They reported that the most frequent abnormal findings were small foci of increased T2 signal in white matter involving the centrum semiovale, the corona radiata, the internal capsule, the periventricular region or the sub cortical white matter (U fibres). The cortex was of normal signal intensity in all cases. However, other studies have reported different results [15-18]. As a review concluded, perhaps the most consistent findings in CFS are a decline in grey matter volume and enlargement of the ventricular volume [18].

The lack of knowledge relating to progressive ME means that our understanding of this condition is extremely limited. Without studying this subgroup, it is possible that the findings from the research on CFS or ME/CFS may not applicable to these patients and that certain interventions could have significant adverse effects. It is also important to consider whether the history described above represents a progression of ME or a novel disease, resembling both ME and MS.

Conclusion

The patient has symptoms both of a progressive form of ME as defined above, and of MS. However, the absence of diplopia, true ataxia, spasticity and Babinski signs and the findings on MRI are not consistent with MS. At the moment, the lack of a recognised 'label' leaves individuals with increasingly severe ME in a medical no-man's land. It is hoped that this case history encourages physicians and researchers to study this group, not only to increase diagnostic precision but also to improve the management of this most challenging disease.

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