New Daily Persistent Headache As A Presenting Symptom Of Hashimoto's Encephalopathy

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Abstract

BACKGROUND: Hashimoto's encephalopathy (HE) is a rare autoimmune neurological disorder often unrecognized. Although headache is experienced by a significant number of patients with this condition, it is overshadowed by more striking neuropsychiatric symptoms and normally is not a presenting complaint. A patient with Hashimoto's encephalopathy and symptoms simulating migraine with aura was previously reported.

METHODS: Repeated clinical neurological examinations and follow-up, ancillary tests including serology, imaging procedures, electroencephalogram (EEG), spinal fluid examinations (CSF) and neuropsychological evaluation.

CASE REPORT: A patient is described with HE manifested as new daily persistent headache, taking the form of hemicrania continua without cranial autonomic signs. Headache was associated with non-fluent aphasia. Indomethacin was ineffective but he responded to steroids and to monthly intravenous immunoglobulin infusions.

CONCLUSION: Hashimoto's encephalopathy may present with headache with a mixed phenotype of both new daily persistent headache and atypical hemicrania continua, amenable to treatment by immunosuppressant agents.

Introduction

Hashimoto's encephalopathy (HE) is a rare chronic neurological disorder with protean and fluctuating clinical manifestations [1,2]. Patients have very elevated serum titer of thyroid microsomal and antithyroglobulin antibodies. In HE, headache may occur in up to 80% of the patients, but is normally mild or periodic, is not a presenting sign, or is usually attributed to previous migraine and hypothyroidism. Furthermore, if headache is present, it is overshadowed by other more striking concurrent symptoms of behavioral or psychiatric nature [3].

Jimenez Huete, et al, reported a female patient with history of periodic global headache associated with sound and light super-sensitivity, who developed an acute left sided headache and aphasia. Her thyroglobulin and thyroid antimicrosomal antibodies titers were found markedly elevated. The authors concluded that HE may be imitate migraine with aura [4].

Case Report

A 50 year old male heavy equipment operator was referred in neurological consultation because of persistent, daily, left sided headache of abrupt onset with fluctuations in intensity. He remembered the exact day that his headache had begun, six months earlier. Pain was pressure-like and a constant ache when milder, and pulsating, 10/10 in the VAS, when intense, always left sided. He experienced superimposed, very intense, brief jolts of sharp pain on the left side of the head, mostly located over the forehead. The patient and his family, also noted that his speech had become laborious and the words were poorly enunciated, to the point that he could not be understood. He reported no other symptoms and had no anteceding illness.

His previous medical history included pan-sinusitis, hypothyroidism and vitamin B12 deficiency. He had no history of psychiatric illness. His father had Parkinson’s disease and one great aunt had Alzheimer’s dementia. He was receiving thyroid and B12 replacement therapy. General physical and neurological examinations were normal except for non-fluent aphasia. Neuropsychological testing confirmed erosions in the areas of verbal abilities and working memory, but preservation of awareness of his level of impairment.

The following tests were normal: brain MRI and MRA, EEG, sedimentation rate, ANA, CK, serum protein electrophoresis, Lyme’s titer, HIV test, angiotensin converting enzyme, SSA-SSB antibodies and a complete paraneoplastic antibody profile.

CSF studies showed a mild elevation of IGG (33 mg/L) but a normal IGG/albumin ratio. There were no CSF oligoclonal bands. His serum thyroid microsomal antibody titer was more than 1.000 IU/ML and his thyroglobulin antibodies were 227 IU/ML.

A two week trial of indomethacin at a dose of 50 g t.i.d. was ineffective. Intravenous methyl prednisolone at a dose of 1 gram daily follow by a three weeks taper of prednisone provided substantial relief from his headache. Eventually he was began on intravenous...
immunoglobulin (IVlg) five days-monthly infusions at a dose of 0.4 grams per kilo, resulting in significant improvement of his aphasia. He takes ibuprofen 600 mg t.i.d. for headache, which returned after prednisone was stopped, but with lesser intensity.

Discussion

New daily persistent headache (NDPH) is refractory to treatment, although few examples of post-infectious NDPH, may respond to a course of intravenous methyl prednisolone [5]. According to Rozen, the headache in NDPH must be bilateral or global, developing in someone without previous history of tension-type headache [6]. The pain may be pulsating, pressure-like, or both, with photo and phonosensitivity. Along with chronic-tension type headache, chronic migraine and HC, NDPH constitutes a subtype of chronic daily headache [6] Given than this patient’s had no history of antecedent headache, that it began abruptly, to the point that he remembered distinctly the date of onset, and because it became daily from the offset, accompanied by light and sound sensitivity, his headache can be diagnosed as NDPH, even than his pain was hemi-cranial without shifting sides. Conversely, because of its continuity with periodic exacerbations, its strict lateralization and nature of the pain, including superimposed “jabs and jolts”, it could be also classified as HC, albeit, he had no history of episodic paroxysmal hemicrania and cluster headache. The lack of cranial dysautonomia and therapeutic response to indomethacin, do not preclude the diagnosis of HC. Up to 1/3 of patients with HC do not report autonomic symptoms, and many patients do not respond to indomethacin. [7, 8] Originally, an absolute therapeutic response to indomethacin was considered a pre-requisite for the diagnosis of HC [8]. I suggest that this patient presented an overlapping phenotype of NDPH and HC that defies strict classification.

The diagnosis of HE can proof very difficult. Many patients are erroneously diagnosed with viral encephalitis, neurodegenerative dementia, and Creutzfeld-Jakob disease [1,2,3] Castillo, et al, on their series of twenty patients from the Mayo Clinic, identified abnormalities on brain magnetic resonance and cerebrospinal fluid analysis in only around 25% of the patients with HE [1] Most of their patients were euthyroid. The same group of investigators encountered that 80% of patients with HE, exhibited aphasia, while headache was reported in 50 %, of the patients, although not as the primary symptom of concern. Of interest, regarding the patient herein discussed, other than headache, the other presenting major symptom he exhibited was an expressive language disorder, suggestive of primary progressive aphasia, a localized brain neurodegenerative disorder. The pathophysiology of HE has not been elucidated. Because of its specific association with thyroid antibodies typical of Hashimoto thyroiditis, it was believed at first that the neuropsychiatric symptoms of this disorder were based on hypothyroidism. Overtime it became apparent that many patients with HE were euthyroid. Subsequently, it was proposed that antibodies primary directed against the thyroid gland, “leaked” across the blood-brain barrier into the brain parenchyma, inducing an autoimmune lymphocytic response based on shared antigens between brain and thyroid. Additional theories advanced to explain the development of HE are, among others, isolated CNS vasculitis due to precipitation of immune complexes, pandysautonomia, demyelination, intratechal synthesis of thyroid antibodies, global hypoperfusion, CNS demyelination, low CSF hypocretin-1 level (in cases of hyper somnolence or coma) and changes in the estrogen/progesterone ratio during the luteal phase, in women with menstrual exacerbations [1,2,3,9,10] The current prevailing hypothesis still calls for an autoimmune explanation because HE is amenable to treatment with immune modulators. The presence of serum thyroid antibodies is interpreted as a non-specific marker of an autoimmune process, ultimately based on cell apoptosis triggered by interleukin-1 beta and the cellular membrane induction of Fas ligands [11]. It is possible to speculate than in HE, an associated inflammatory response (asymmetric in this example) affecting the trigemino-vascular brain innervations, may result in peri-vascular nocioceptor activation and hemicrania continua.

Patients with HE normally respond to steroids, hence, this condition is also named “steroid responsive encephalopathy” [1]. Current knowledge supports treatment of HE with intravenous gamma globulin (IVlg) because it is effective, well tolerated, and lacks the well known undesirable adverse side effects caused by the long usage of steroids [2]. No particular studies have been directed to the treatment of headache in patients with HE, given the relative rarity of the disorder and because of the reasons listed in the introduction. Regardless, steroids are indicated for most HE patients, even if they do not have headache. Of specific relevance to this case, steroids were also indicated given that some patients with HC may respond to intravenous steroids, as reported by Prakash, et al [12]. IVlg infusions ameliorated this patient’s aphasia and cognitive impairment, but were only partially effective in relieving his headache, aside
the expected limited exacerbations during the actual infusions. Eventually, he needed to take ibuprofen on a daily basis since indomethacin was ineffective, as he was wary of taking steroids on a long term basis.

References

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