Post-Orgasmic New Daily Persistent Headache In a Patient With Hemochromatosis and Idiopathic Intracranial Hypertension Without Papilledema

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Abstract

A 56 year old female with hemochromatosis-HFE C282Y homozygous gene mutation and remote history of infrequent isolated visual auras developed new daily persistent headache (NDPH) following a single orgasm. Headache was eventually ameliorated by amitriptyline and topiramate after 18 months following the precipitating event. Her neurological examination and ancillary imaging procedures including head CT, brain MRI-MRA and MRV were normal except for a very small parietal meningioma representing an incidental finding. There was no evidence of reversible vasoconstriction syndrome (RCVS). Her cerebrospinal (CSF) pressure was elevated compatible with idiopathic intracranial hypertension (IIH) without papilledema. There was no sign of bleeding in the CSF but her protein content was mildly increased in the absence of oligoclonal bands. Hemochromatosis (HC) predisposes to migraine in women. The origin of this patient’s headache remains unclear, but the association of primary post-orgasmic NDPH, hemochromatosis and IIH represents a unique clinical presentation.

Introduction

Orgasmic headache (OH) is the classic form of sexual headache typically benign, often recurrent but self-limited, not periodic or predictable, normally lasting only for a few hours. The headache is usually severe (“thunderclap” or “explosive”) and of rapid development during the climax of sexual activity. It is precipitated by either intercourse or masturbation. Pre and post-orgasmic headaches are also identified (1, 2). Sexual headache may be classified as a Valsalva-induced or exertion type of headache; however, cases of more ominous significance has been reported in the literature. They include examples of subarachnoid hemorrhage, embolic stroke, intracranial hematoma, basilar artery dissection and basilar artery stenosis (3, 4).

New daily persistent headache (NDPH) is one of the presentations of chronic daily headache (CDH). The other three categories are chronic migraine, chronic tension-type headache and hemicrania continua (5). According to Li and Rozen, NDPH must be bilateral or global, with pain features that may be pulsating, pressure-like, or both, and accompanied by sound and light supersensitivity in 60 to 70% of the cases. Headache must last a minimum of one month and should occur at least 15 days a month, for more than one month. Headaches should last at least for four hours every day in the symptomatic days. By definition the headache will be of new onset; stated differently, NDPH should not be diagnosed in someone already suffering from other types of CDH (5).

Nick and Bakouche reported that primary post-coital headache rarely persists for up to a few days (6). To my knowledge, no examples of longer duration (weeks or months) persistent primary orgasmic-induced headache, as a variant of NDPH, has been described. Of parallel interest, a 41 year old patient with hemochromatosis (HC) due to a homozygous C282Y mutation was reported with hemicrania continua lasting for several months, slowly evolving from episodic hemicrania, with no apparent precipitant. His headache was substantially ameliorated by venesection (7).

Case Report

A 56 year old nurse was seen in consultation after she developed a paroxysmal excruciating (“thunderclap”) headache while achieving sexual orgasm. The headache was bilateral, occipital in location, radiated to the temples, was severe and throbbing in nature. It was not associated with photophobia, or vomiting. The day after the initial episode she was drowsy and her blood pressure (BP) was 160/104 mm Hg. She had no history of headache but only of rare isolated visual auras. The headache persisted with a lesser intensity but continued daily for 3 months. After 3 month she had headaches only 2 to 3 times a week, relieved by common analgesics. She experienced no more OH after the first episode. Initially she was given Gabapentin but she developed a rash. She chose then not to take additional prophylactic medications. After
18 month the original occipital daily headache returned on a daily basis, although not with the initial severity, and was accompanied by non-pulsating tinnitus. She was given verapamil following her neurological re-evaluation but she developed a rash again. Amitryptiline 10 mg h.s. reduced the intensity of her headache. She could not tolerate a larger dose. Additional improvement was achieved with the addition of topiramate at a dose of 50 mg b.i.d.

Her past history included atypical chest pain with a negative cardiac evaluation including a coronary angiogram and echocardiogram. She had been diagnosed 2 years earlier with phenotypic HC due to homozygous mutation HFE C 282-Y. Liver biopsy demonstrated iron overload with an iron index of 2.2 but no cirrhosis. Her initial ferritin and iron saturation levels were elevated. She was treated by venesection ever 3 weeks initially and subsequently every 3 to 6 months according to her blood work. Venesections did not suppress or ameliorated her headaches. Additional history included mild hypertension, sciatica, anxiety disorder, vitamin D deficiency and familial essential tremors. She had no family history of HC and genetic testing on her two children was negative. She took atenolol 50 mg a day and vitamin D 2.000 IU a day. On physical examination her BP was 110/74 and her pulse was 64. Her BMI was 23.6. Her neurological examinations on several occasions were normal except for mild postural tremor of the fingers. There was no meningismus. Her initial head CT and brain MRI showed an ovoid 9 mm, partially calcified, mildly enhancing, extra-axial left parietal lesion representing a small meningioma; otherwise imaging was normal without signs of generalized or peri-tumoral edema or hemorrhaghe, parenchymal bleed or hydrocephalus. Brain MRA and MRV with contrast were normal. Repeat brain MRI, MRA and MRV 18 months after the initial imaging studies were completed, showed no changes, including on the size and appearance the suspected small meningioma. There was no sign of RCVS and no vascular anomalies were identified in the initial and follow-up studies. Her EEG showed a normal background with no epileptic discharges, periodic complexes or sustained focal slowing. Her CSF opening pressure, once she returned 18 month later from the original visit complaining of a recurring occipital headache and tinnitus, was elevated at 28 cm of water. She had no cells and only mild elevation of her CSF protein at 57 mg/DL. There were zero oligoclonal bands in the CSF.

Discussion

Hemochromatosis is an autosomal recessive blood disorder due to various genetic mutations, resulting in elevated absorption of iron and secondary progressive accumulation of iron in the liver, pancreas and pituitary gland, among several organs. In the brain, excessive iron overload causes neurodegeneration by way of cumulative oxidative neuronal injury (8). High prevalence of headache seems to exist in women with HC, in particular on those harboring the H63D polymorphism of the HFE gene (9). Iron accumulation in the peri-aqueductal gray (PAG), an area sub-serving pain modulation given its concentration of substance P, orexin, oxytocin, endogenous opiates and endocannabinoids, is believed to result in progressive neurodegeneration (8,9,10, 11). The latter mechanism has been hypothesized as the source for recurrent headache in HC, and in another venue, for the transformation of episodic headache into CDH, by way of creating central sensitization (i.e., induction of chronic central pain) (7,9,10).

The etiology of orgasmic headache has not been ultimately defined. It has been explained on basis of several potential mechanisms that are not mutually exclusive: 1. Reversible cerebral vasoconstriction syndrome (RCVS) (12) 2. Paroxysmal hypertension with loss of cerebral vascular auto-regulation during climax. 3. Underlying undiagnosed intracranial venous stenosis resulting in transient intracranial hypertension during orgasm (13) 4. Migraine diathesis. 5. Valsalva maneuver during coitus inducing maximal pulsing distention of the intracranial vasculature (‘microdissections’?). 6. Unsuspected Arnold Chiari malformation (ACM) or cerebellar tonsil ectopia with transient impaction on the spinal canal interrupting CSF outflow (14) 7. Ill-defined hormonal release or release of pro-inflammatory cytokines during intercourse, inducing acute “post-orgasm illness syndrome”, potentially relieved by non-steroidal anti-inflammatory agents. (15). No one of these potential mechanisms however, will explain persistent post-orgasmic headache; therefore the justification to diagnose this patient with primary post-orgasmic NDPH.

The etiology of NDPH is also of multifactorial origin (5). Secondary forms of NDPH that could also apply to secondary orgasmic persistent headache include intracranial bleed, acute cerebral venous thrombosis and acute CSF leaks from unsuspected arachnoid traumatic tears during sexual activity causing low CSF pressure. NDPH could equally result from unsuspected ACM complicated by irreversible impaction of the tonsils of the cerebellum into the
cranio-cervical junction (16). Although one patient with a giant subarachnoid cyst suffered from OH, meningiomas have not been associated to OH (17). This patient's menigioma was very small, causing no mass effect, bleeding or hydrocephalus. It was believed to represent an incidental finding totally unrelated to her intracranial hypertension.

In is uncertain as to the exact nature of this patient primary post-orgasmic NDPH. It is conceivable that her IIH was significant only as a chronic subclinical accompaniment of her HC that was present all along, or constituted a delayed complication from her OH. Certainly, she exhibited none of the conditions listed above that precipitate NDPH. Conversely, it can be speculated in the absence of micro-bleeds, vasoconstriction, cerebral venous sinus stenosis or thrombosis, that HC and IIH predisposed her to develop an orgasmic headache that ultimately acted upon immediate response genes, up-regulating her PAG nocioceptive neurotransmitters receptors. It could be inferred that the latter mechanism, lead into a condition of chronic central pain sensitization, albeit of fluctuating nature, perpetuated by her previously asymptomatic intracranial hypertension (10,11,18). If subclinical neurodegeneration of the PAG was present in the background prior to the appearance of her headache, it remains to be demonstrated.

References

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