Genital sensory aura in migraine

Javier Casas-Limón [a], M. Luz Cuadrado-Pérez [a], Jordi González-Menacho [b], Josep M. Olivé-Plana [b], Carlos Manuel Ordás-Bandera [a], Gloria Vives-Masdeu [b], Jordi A. Matías-Guiu [a], Jesús Porta-Etessam [a]

[a] Department of Neurology. Hospital Clínico San Carlos. Universidad Complutense. Madrid. [b] Department of Neurology. Hospital Universitari Sant Joan de Reus. Universitat Rovira i Virgili. Reus, Tarragona, Spain.

Corresponding author: Dr. Javier Casas Limón. Servicio de Neurología. Hospital Clínico San Carlos. Profesor Martín Lagos, s/n. E-28040 Madrid.

E-mail: jcasaslimon@gmail.com

The first case report was partially presented as a poster at the LXIII Annual Meeting of the Spanish Neurological Society (Barcelona, November 2011).

Accepted: 06.07.12.

How to cite this article: Casas-Limón J, Cuadrado-Pérez ML, González-Menacho J, Olivé-Plana JM, Ordás-Bandera CM, Vives-Masdeu G, et al. Genital sensory aura in migraine. Rev Neurol 2012; 55: 445-7.

Versión española disponible en www.neurologia.com

Migraine aura consists of reversible focal neurological symptoms that usually develop gradually over 5 to 60 minutes and last less than 60 minutes. Aura symptoms normally occur just before or at the onset of migraine headache. Less commonly, the subsequent headache lacks migrainous features, or there is no headache at all. Typical aura symptoms are visual, sensory or language disturbances, and may occur alone or in succession [1].

Sensory aura is the second in frequency after the visual aura (31% versus 99% in Russell and Olesen's series). It may appear in form of paraesthesia, hypoaesthesia, or both. Sensory symptoms classically begin in one hand (96%), spread to the arm (78%), and reach the perioral region (67%), sometimes including the tongue. Then they can extend to the ipsilateral foot and leg in 24% of cases [2]. We have not found in the available literature any description of sensory aura involving the genital region. Here we report two patients who experienced prominent sensory symptoms in the genital region during their migraine auras.

Patient 1. A 39-year-old woman had been suffering migraine attacks for the last 7 years. She had no other diseases. Headache frequency was variable, from 0 to 3 attacks per month, and headache duration ranged from 1 to 3 days. She could identify some triggers, like eating chocolate, sleeping more than usual, or weather changes. Head pain was mainly unilateral, with side-shift. She described it as a pulsating, severe pain that increased with physical activity. There were associated nausea, photophobia and phonophobia. In the most severe episodes, the headache was preceded by right hemianopia, which was consistently followed by intense paraesthesia in both the right half of her mouth and the ipsilateral vulva. The paraesthetic sensations appeared gradually in the right perioral region and then extended to the right genital area sparing other parts of the body. Genital sensations were most disturbing for the patient. The whole sensory aura usually lasted 15 to 20 minutes, but could persist up to 40 minutes. The neurological exam was normal between attacks. Periodic gynaecological exams were also normal. The patient did not take any preventives. She did not find any remedy for her auras, but acute treatment with rizatriptan 10 mg was always effective in terminating her headaches.

Patient 2. A 42-year-old man, with no relevant medical history, began suffering migraine episodes 8 years before consulting. The episodes usually started with photopsias dispersed through the entire visual field, followed by paraesthesia in the left arm, face and tongue during 20-40 minutes. Then he felt a severe pressing bilateral parietal headache, with associated photophobia, but no nausea or vomiting, which could last from several hours to 3 days and interfered with his daily activities. This happened approximately twice a month. In the last 2 years, those episodes had persisted, but he also had a new type of attack about once per month. It started with distressing paraesthesia clearly localized in his testicles, more evident on the left. After 8-10 minutes the sensory symptoms gradually progressed upwards to the left hand, arm and face, where they were not as intense. The whole sensory aura lasted 20 minutes and was followed by a headache similar to his previous migrainous headaches. Between attacks, both the neurological and the urological exams were normal. Brain and lumbar magnetic resonance imaging (MRI) were performed, and did not show any abnormalities. After initiating preventive treatment with lamotrigine 100 mg per day, headache frequency did not change, but both the duration and the severity of his sensory auras, including those with genital paraesthesia, were reduced to the half.

Nowadays there is growing evidence that migraine aura has not a vascular cause. Despite some implications in cerebral perfusion, aura symptoms are originated in the brain [3]. Cortical spreading depression (CSD) is now accepted as the mechanism underlying migraine aura. CSD was first described by Leão in 1944 [4], who induced a slowly spreading depression of electroencephalography (EEG) activity in rabbits through local electrical stimuli. During migraine aura, a pronounced depolarization of neurons and glia initiates at the occipital pole, and then propagates steadily across the cortex followed by a period of depressed electrophysiological activity [5]. As a result, there is an initial wave of cortical hyperemia followed by prolonged oligoemia lasting more than 60 minutes [6].

CSD has been the target of important research. Some studies in humans support the hypothesis that CSD is the cause of migraine aura. For instance, Hadjikhani et al used high field functional MRI (fMRI) to prove that a wave of transient hyperperfusion followed by sustained hypoperfusion did spread through the occipital cortex at a rate of 3.5 mm/min while patients were experiencing a visual aura. Moreover, a correlation between these cortical phenomena and the progression of aura symptoms could be demonstrated [7]. On the other hand, the studies performed by Bolay et al in experimental models have revealed that CSD may activate the trigeminal nucleus caudalis via trigeminovascular afferents. Therefore it is possible that CSD could also lead to the development of headache [8].

As stated above, sensory aura consisting of tingling or numbness is the second in frequency after the visual aura, and normally involves the hand, the arm, the face and/or the leg [2,9]. We have not found in the literature any cases of genital symptoms during the migraine aura. Other non-aura sensory symptoms have been linked to migraine. For example, cephalic and extracephalic allodynia may occur as a sign of sensory sensitization during migraine episodes [10,11]. In addition, spontaneous body pain has been described in a few cases as a consequence of central sensitization [12,13]. However, the genital sensations did not have a painful component in our two patients. Besides, they were

invariably connected to typical aura symptoms in a marching pattern [1]. Accordingly, they should be taken as true aura manifestations.

Vulvodynia and scrotodynia are well-known syndromes producing persistent pain, soreness or burning sensations within the genital region, not attributable to local disease or to a specific neurological disorder. These localized topographic pain syndromes have been occasionally associated with glossodynia or stomatodynia [14-16], suggesting a common pathologic substrate, most probably central sensitization although a coexisting small fibre neuropathy cannot be excluded. During her migraine auras, our first patient had transient paresthesia in the same separate areas, i.e. the mouth and the genital area. In this case the symptoms would also be a consequence of central dysfunction, but they should be attributed to CSD. Interestingly, fMRI studies have shown that genital touch may activate the insular and frontal cortical regions just close to the cortical sensory representation of the perioral region [17].

The reasons for a low incidence of sensory symptoms in the genital region during migraine aura are difficult to explain. Perhaps, the cortical neurons representing the genital area have a higher threshold to be activated during the CSD. Anyhow, the progression of sensory aura symptoms does not necessarily follow the somatosensory homunculus, as classically described by Penfield and Rasmussen [18]. Otherwise, certain feelings of embarrassment can preclude the patients from confessing this sort of symptoms, so they may have been under-recognized.

Genital disturbances have not been described in migraine before, but other neurological disorders are known to produce this kind of symptoms, with or without an emotional content. Seizures are probably the neurological events in which genital symptoms are best characterized. Not only orgasms may trigger reflex epileptic seizures [19], but genital symptoms may represent true epileptic manifestations. Genital automatisms or sensations may be part of complex partial seizures [20]. Sexual auras presenting as erotic pleasant feelings or thoughts with or without sexual arousal and orgasm are associated with temporal lobe epilepsy, occurring predominantly in women [21]. Genital sensations like numbness, tingling, vaginal feeling with compulsion to masturbate, and pain or unpleasant sensation in the genitals may indicate the existence of an epileptic focus in the post-central gyrus, interhemispheric fissure, and perisylvian region [22]. Neuronal hyperexcitability is a common pathophysiological mechanism shared by seizures and migraine aura. Lamotrigine is an antiepileptic drug with proven efficacy in migraine auras [23,24]. In fact, our second patient had a significant reduction of his aura symptoms once lamotrigine was initiated.

In conclusion, genital symptoms during the migraine aura have not been communicated in the available scientific literature up to now. Here we present two patients whose main complaint was the occurrence of genital sensory symptoms during their migraine auras. These symptoms resemble the genital sensations that have been related to seizure activity in some epileptic patients. Like other migraine auras, this type of auras should be taken as a consequence of CSD.

References

1. Headache Classification Subcommittee of the International Headache Society. The international classification of headache disorders, 2nd edition. Cephalalgia 2004; 24 (Suppl. 1): S9-160.
2. Russell MB, Olesen J. A nosographic analysis of the migraine aura in a general population. Brain 1996; 119: 355-61.
3. Cohen AS, Goadsby PJ. Functional neuroimaging of primary headache disorders. Expert Rev Neurother 2006; 6: 1159-71.
4. Leão AAP. Spreading depression of activity in cerebral cortex. J Neurophysiol 1944; 7: 359-90.
5. Guiou M, Sheth S, Nemoto M, Walker M, Pouratian N, Ba A, et al. Cortical spreading depression produces long-term disruption of activity-related changes in cerebral blood volume and neurovascular coupling. J Biomed Opt 2005; 10: 11004.
6. Tfelt-Hansen PC. History of migraine with aura and cortical spreading depression from 1941 and onwards. Cephalalgia 2010; 30: 780-92.
7. Hadjikhani N, Sánchez del Río M, Wu O, Schwartz D, Bakker D, Fischl B, et al. Mechanisms of migraine aura revealed by functional MRI in human visual cortex. Proc Natl Acad Sci USA 2001; 98: 4687-92.
8. Bolay H, Reuter U, Dunn AK, Huang Z, Boas DA, Moskowitz MA. Intrinsic brain activity triggers trigeminal meningeal afferents in a migraine model. Nat Med 2002; 8: 136-42.
9. Cologno D, Torelli P, Cademartiri C, Manzoni GC. A prospective study of migraine with aura attacks in a headache clinic population. Cephalalgia 2000; 20: 925-30.
10. Burstein R, Yarnitsky D, Goor-Aryeh I, Ransil BJ, Bajwa ZH. An association between migraine and cutaneous allodynia. Ann Neurol 2000; 47: 614-24.
11. Burstein R, Cutrer MF, Yarnitsky D. The development of cutaneous allodynia during a migraine attack. Clinical evidence for the sequential recruitment of spinal and supraspinal nociceptive neurons in migraine. Brain 2000; 123: 1703-9.
12. Cuadrado ML, Young WB, Fernández-de-las-Peñas C, Arias JA, Pareja JA. Migrainous corpalgia: body pain and allodynia associated with migraine attacks. Cephalalgia 2008; 28: 87-91.
13. Prakash S, Shah ND, Dholakia SY. Recurrent limb pain and migraine: case reports and a clinical review. Cephalalgia 2009; 29: 898-905.
14. Gaitonde P, Rostron J, Longman L, Field EA. Burning mouth syndrome and vulvodynia coexisting in the same patient: a case report. Dent Update 2002; 29: 75-6.
15. Petruzzi M, De Benedittis M, Pastore L, Serpico R. Vulvostomatodynia. Maturitas 2007; 58: 102-6.
16. Mancuso G, Berdondini RM. Simultaneous occurrence of dysaesthetic peno/scroto-dynia and stomatodynia. Int J STD AIDS 2005; 16: 830-1.
17. Pukall CF, Strigo IA, Binik YM, Amsel R, Khalifé S, Bushnell MC. Neural correlates of painful genital touch in women with vulvar vestibulitis syndrome. Pain 2005; 115: 118-27.
18. Vanopdenbosch L, Herroelen L. Leão's cortical spreading depression and the somatosensory homunculus: a contradiction? Headache 1998; 38: 322-3.
19. Ozkara C, Ozdemir S, Yilmaz A, Uzan M, Yeni N, Ozmen M. Orgasm-induced seizures: a study of six patients. Epilepsia 2006; 47: 2193-7.
20. Bancaud J, Favel P, Bonis A, Bordas-Ferrer M, Miravet J, Talairach J. Paroxysmal sexual manifestations and temporal epilepsy. Electroencephalogr Clin Neurophysiol 1971; 30: 371.
21. Currier RD, Little SC, Suess JF, Andy OJ. Sexual seizures. Arch Neurol 1971; 25: 260-4.
22. Ruff RL. Orgasmic epilepsy. Neurology 1980; 30: 1252.
23. Steiner TJ, Findley LJ, Yuen AWC. Lamotrigine versus placebo in the prophylaxis of migraine with and without aura. Cephalalgia 1997; 17: 109-12.
24. Pascual J, Caminero AB, Mateos V, Roig C, Leira R, García-Moncó C, et al. Preventing disturbing migraine aura with lamotrigine: an open study. Headache 2004; 44: 1024-8.