

A case of cranial autonomic dysfunctions predominant migraine: Migraine sans ache

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ABSTRACT

Pain is the essential part of migraine headaches along with other features, whereas, cranial autonomic symptoms (CAS)/signs are a predominant and essential part of trigeminal autonomic cephalalgias. What if other features of migraine are present but the pain is absent? One such variation in migraine symptomatology is reported in this case, where the pain was not there but CAS/signs were predominant along with other features of migraine. The possible mechanism for such variation, in this case, maybe desensitization of afferent loop (comprised by nociceptors in extracranial and intracranial pain-sensitive structures supplied by the trigeminal nerve) and direct activation of the trigemino-cervical complex and brainstem structures causing dissociation of pain from the rest of the features of migraine. In this case, as the pain part gets completely dissociated from CAS/signs, hence this condition can be termed as “migraine sans ache.” The patient was started on anti-migraine treatment and she responded wonderfully.

Key words: Anti-migraine therapy, Cranial autonomic symptoms, Dissociated headache, Nociceptors, Trigemino-cervical complex

Migraine is a common disabling primary headache disorder with recurrent headaches manifesting in attacks lasting from 4 to 72 h. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity, and association with nausea and/or photophobia and phonophobia [1]. Migraine attacks can be associated with cranial autonomic symptoms (CAS) like conjunctival injection, lacrimation, nasal congestion, rhinorrhea, eyelid edema, forehead and facial sweating, miosis, ptosis, or aural fullness. These CAS are typical and essential features of trigeminal autonomic cephalalgias (TAC) but can be associated with migraine attacks [2,3].

Different variations in migraine symptomatology have been described in past and we are here reporting such a variation in this case, where CAS were prominent features rather than pain. Such variation is rare and has not been reported in the literature till now. Detailed symptoms and signs of the patient are discussed and also the possible explanation of this dissociation is discussed by analyzing the pathophysiology of migraine especially the role of the trigemino-cervical complex. Therefore, this case helps us to better understand the intriguing pathophysiology of migraine. Consent was obtained from the patient in question.

CASE REPORT

A 28-year-old woman presented with episodes of CAS in form of redness and tearing from eyes, bilateral facial and forehead flushing, and bilateral aural fullness without any pain in the head or neck for the last eight months. These episodes were associated with triggers like traveling, hunger, or decreased sleep. Occasional prodromal yawning was present. Photophobia and phonophobia were not present but the nausea was present during few episodes. These CAS evolve gradually, peak over 2–4 h, and subside in 12–24 h. The patient had no pain, but she felt restlessness and discomfort during these episodes. Some activity restriction was also present during these episodes. Though the pain was not there, she tried analgesics due to the discomfort but were ineffective. She visited our clinic for these CAS and discomfort during episodes. No menstrual associations with CAS were present. Stressors were absent. Psychiatric co-morbidities screened by the Patient Health Questionnaire showed negative results. No history of any eye inflammatory conditions such as uveitis or conjunctivitis in the past. No history of any connective tissue or autoimmune disorder in past. Family history for headaches was negative.

The patient was afebrile, her blood pressure was 120/70 mm of Hg, with a normal respiratory, cardiac and abdominal system on general physical examination. Neurological examination including higher mental functions, cranial nerves, motor, sensory, cerebellar, and extrapyramidal system was within normal limits.

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Access this article online	
Received - 18 July 2021 Initial Review - 03 August 2021 Accepted - 14 August 2021	Quick Response code 
DOI: 10.32677/ijcr.v7i9.3007	

Local eye examination during attacks shows conjunctival injection and watering in both eyes. Bilateral forehead flushing was also present during attacks.

On investigation, the patient's blood counts, electrolytes, renal function, and liver function tests were within normal range. Her erythrocyte sedimentation rate was 9 mm/h. Thyroid function test, Vitamin B12, and Vitamin D levels were within normal range. Autoimmune and connective tissue disorder workup (ANA, c-ANCA, p-ANCA, and APLA) was negative. Magnetic resonance (MR) imaging brain with MR angiography of the brain and neck vessels did not show any abnormal parenchymal or vascular lesions.

Although she was not fulfilling the criteria of episodic migraine without aura as she was having characteristic features of migraine other than pain, a diagnosis of "migraine without aura with predominant CAS" was considered. The patient was started on propranolol 20 mg twice daily for prophylactic treatment and naproxen sodium 500 mg with domperidone 10 mg combination for acute termination of CAS. She was advised to follow-up with a headache diary which was pointing out CAS in terms of onset, severity, laterality, and frequency. Her CAS with other features such as prodromal yawning, nausea, and restlessness gradually improved and completely subsided over 3–4 months.

DISCUSSION

One or more unilateral or bilateral CAS can be present in around 27–73% of patients with migraine [4-7]. However, CAS themselves does not define migraine, and unilateral CAS are characteristics of TAC [1]. In this case, episodic bilateral long-lasting CAS were present with nausea, restlessness, and some activity restriction. It can be inferred that, in this case of migraine, the pain part gets dissociated leaving behind a significant CAS part and few other associated features such as nausea and activity restriction. However, these CAS predominant episodes do not fulfill current the International Classification of Headache Disorders-3 classification criteria for migraine or any other headache of current classification. As these CAS episodes were episodic, bilateral, and of long duration, these cannot be justified in the TAC group also. As the pain was absent with other features of migraine, therefore "migraine sans ache" terminology can be used for this type of headache. Other disorders where CAS is found to be associated with headaches are few secondary headaches like pituitary dysfunction, intracranial aneurysm, and sinus headaches [8]. These secondary headaches were ruled out by brain imaging. Lesions in the higher autonomic center of the brain and any bilateral lesions in the sympathetic and parasympathetic nervous systems were also ruled out.

In this case, to properly understand the phenomenology, we need to understand the pathophysiology of CAS in different headache disorders. Sensory oversensitivity causes activation of nociceptors in extracranial and intracranial pain-sensitive structures supplied by the trigeminal nerve causing pain [9].

Signals from the trigeminal nerve traverse trigeminal ganglion and reach the trigeminal nucleus caudalis and the upper cervical spinal levels forming a trigemino-cervical complex. From the trigemino-cervical complex through genetically sensitized pathways signals reach to brainstem, medullary, diencephalic, hypothalamic, and thalamic areas, and finally to the parasympathetic superior salivatory nucleus (SSN) in the pons, activation of which causes CAS. Therefore, pain in the trigeminal nerve and upper cervical spinal nerves supplied area of the head and neck is a predominant feature with CAS depending on the involvement of brainstem sensitization and activation. This phenomenology is present in most migraine patients.

Rozen TD described such dissociation of pain and CAS in 2 patients with long-lasting autonomic symptoms with hemicranias (LASH) where CAS exists beyond pain and lasts longer than pain [10,11]. But in LASH, pain and CAS were hemicranial and LASH was described as indomethacin responsive TAC. Toribio-Díaz ME also described 10 cases with prolonged eyelid edema (one of CAS) accompanied with and beyond the pain of migraine [12]. He considered it as an accompaniment of migraine and it was improved with anti-migraine treatment. Our case is unique and different in many aspects. Here, the pain part gets completely dissociated from CAS, hence such a condition can be termed as "migraine sans ache." The presumed cause of this dissociation of pain from the rest part of migraine may be desensitization of afferent loop and direct involvement of the trigemino-cervical complex or brainstem structures (without the participation of nociceptors) causing reflex activation of SSN in pons producing CAS without pain.

CONCLUSION

This case helps us to better understand the pathophysiology of migraine and possible variations in phenomenology in migraine. Therefore, migraine is still a mysterious disease, and variations in phenomenology are possible. We need to identify and report these variations more frequently so that we can better understand this very common disease.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Singh AS, Singhvi JP, Mann HS. A case of cranial autonomic dysfunctions predominant migraine: Migraine sans ache. *Indian J Case Reports*. 2021;7(9):383-385.