Indian J Lepr 2012, 84 : 317-320

http://www.ijl.org.in

Case Report

Is there a relation between Hemicrania continua and Leprosy?

KS Ferreira[1], DJ Freitas[2], JG Speciali[3]

Received : 15.05.2012 Revised : 07.08.2012 Accepted : 30.09.2012

The association of hemicrania continua and leprosy has been described in 2008. This relation can be causal or casual. Hemicrania continua is a strictly unilateral, moderate to severe, continuous, indomethacin-responsive primary headache with autonomic cranial symptoms and leprosy is an usual cause of peripheral neuropathy. Prevalence has fallen in the past years, but transmission continues and leprosy remains a public health problem. The objective of this study is to report one case of headache fulfilling the IHS criteria for HC, presented during the course of leprosy. A 61 years old woman started hypo and hiperpigmented lesions with impaired sensation to touch on right side of face (malar). She had biopsy in facial lesion and histopathology compatible with a borderline leprosy form. At the same time, she reported new headaches, daily and continuous, without pain-free periods, unilateral (which were located in the same side of the leprosy lesion in face), throbbing and severe (VAS=8) with ipsilateral conjuntival injection and lacrimation that improved with indomethacin. We hypothesize that the local injury on the face of this patient predisposes a mechanism of central sensitization, resulting in trigeminal autonomic cephalgia. Relation between trigemino-autonomic cephalalgias and leprosy provides insights into craniofacial pain mechanisms.

Keywords: Hemicrania continua, leprosy, pain mechanisms, central sensitization

Introduction

Hemicrania continua (HC) is a strictly unilateral, moderate to severe, continuous, indomethacin-responsive primary headache with autonomic cranial symptoms. It lasts more than three months and is defined by the current International Classification of Headache Disorders (ICHD-II). Leprosy is the leading infectious cause of disability and an usual cause of peripheral neuropathy. Prevalence has fallen in the past years, but transmission continues and leprosy remains a public health problem. Neuropathic pain is a sensory manifestation of leprosy that affects approximately 60% of patients during their illness. It's a consequence of lesion or dysfunction of the nociceptive pathway at the peripheral or central nervous system. (Garbino et al 2011, Rodrigues and Lockwood 2011). The initial form of leprosy is called indeterminate and is characterized by a hypochromic macula with

[1] KS Ferreira, MD, Headache Clinic
[2] DJ Freitas, MD, Department of Internal Medicine
[3] JG Speciali, PhD, Headache Clinic,
Clinical Hospital, Medical School of Ribeirao Preto, University of São Paulo, Ribeirao Preto, Sao Paulo, Brazil
Correspondence to: KS Ferreira **Email :** karenferreira@usp.br

reduced sensitivity. Depending on the cell-mediated immune system, this initial form may heal spontaneously or leprosy patients may develop distinct clinical groups as follows: polar tuberculoid (TT), borderline tuberculoid (BT), borderline borderline (BB), borderline lepromatous (BL), and polar lepromatous (LL). (Garbino et al 2011, Rodrigues and Lockwood 2011). The reported frequency of cranial nerve paralysis in patients with leprosy varies from 10 to 22%. Facial, trigeminal and olfactory nerves are the commonest nerves to be affected. (Gourie-Devi 2006).

The association of hemicrania continua (HC) and leprosy has been described providing insights into craniofacial pain mechanisms. A single case, with a type of headache fulfilling the IHS criteria for HC and mononeuritis multiplex due to leprosy, was reported in 2008, but there is no other case in literature (Prakash and Dholakia 2008). Herein, we report one case of headache fulfilling the IHS criteria for HC, presented during the course of leprosy.

Objective

The objective of this study is to report one case of headache fulfilling the IHS criteria for HC, presented during the course of leprosy.

Design/Methods

We described a single case in our Headache Clinic - Clinical Hospital, Medical School of Ribeirao Preto, University of São Paulo, Ribeirao Preto, Sao Paulo, Brazil, who fulfills the IHS criteria for HC, during leprosy course. After signing the consent form, participant was then interviewing.

Results

Clinical Report

A 61 years old woman started hypo and hiperpigmented lesions with impaired sensation to touch, pain and temperature on chest, left leg and right side of face (malar). (Figure 1) Some

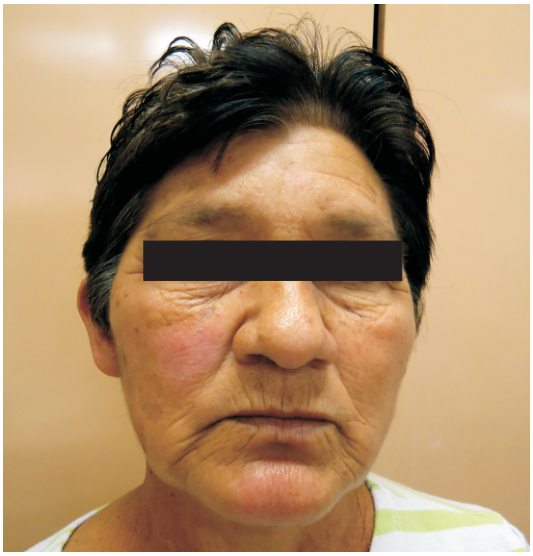

Figure 1 : Leprosy scar in the right side of face (malar).

lesions had red colour and some lesions were hypopigmented. They had absence of oedema with no autonomic disfunction in around the lesions and absence of nerve trunk or cutaneous nerve lesions. She had biopsy in facial lesion and histopathology compatible with a borderline (BB) leprosy form. Other laboratory tests, including antinuclear antibodies and anti-HIV 1 and 2 were negative. Magnetic resonance cranial was normal. At the same time, she reported new headaches, daily and continuous, without pain-free periods, unilateral (which were located in the same side of the leprosy lesion in face), throbbing and severe (VAS=8). During exacerbations, she had ipsilateral conjuntival injection and lacrimation that persisted throughout the attack. Attacks were not aggravated by routine physical and they were not accompanied by agitation, nausea, photophobia or phonophobia. Response to analgesics like paracetamol or naproxen was poor. She had used amitriptyline with no improvement. The patient received World Healthy Organization (WHO) multidrug

therapy for multibacillary leprosy, a 12 months scheme (dapsone, clofazimine and rifampicin). Then, she reported type 1 reaction with this treatment, with red colour and oedema on the facial lesion and also on the rest of the body, without nerve involvement. She had used prednisone (1mg/kg/day) during the course of multidrug therapy for control of these reactional episodes, for nine months. Headaches remained daily during leprosy treatment.

We met this patient in our Neurology Clinic 1,5 year after leprosy diagnosis, with the same headache. Because the headache fulfills the IHS criteria for HC, we decided to start indomethacin, 75 mg 12/12 h. She reported total improvement of her headache. She has been followed for six months, without worsening.

Discussion

In the currently paper we reported one case of HC, developed during the course of cutaneous leprosy. The pain was continuous and lasted 24 hours daily, with exacerbation periods and trigemino-autonomic symptoms. Indomethacin 75 mg 12/12 h induced remission. As she had no improvement after leprosy treatment, we believe this case of headache fulfill the IHS criteria for primary HC.

The relation between HC and leprosy can be causal or casual. Based on temporal association, localization of pain (autonomic signs happened in the region of the leprosy injury) and the previous case of HC and leprosy reported in 2008 by Prakash and Dholakia, we developed the hypoteses of causal relation probably triggered by peripheral neuropathy and/or related to type 2 reaction during treatment.

The WHO treatment for multibacilar leprosy envolves a 12 months scheme (MDT; dapsone, clofazimine and rifampicin). Two types of imune reactions can occur. The type 1 reversal reactions occurs in groups that have an unstable cell mediated immune reaction, such as in the borderline leprosy (or: the type 1 reversal reactions can be associated with any leprosy subtype, although they are more common in patients with borderline leprosy, and it's associated with a unstable cell mediated immune reaction). The type 2 reaction, or erythema nodosum leprosum, occurs almost exclusivelly in BL and LL patients. Deterioration of the nerve function and pain occur mainly during reactional episodes. These reactions usually respond preferentially (or: better) to oral steroids or thalidomide, respectively, but the pain remains more longer (Garbino et al 2011, Rodrigues and Lockwood 2011).

In other hand, cranial nerve paralysis generally occurs along with any types of peripheral neuropathy and rarely may also be an isolated feature. Facial, trigeminal and olfactory nerves can be affected.(Gourie-Devi 2006).

The pathophysiology of HC seems to envolve hypothalamus and the brain stem. A recent positron-emission tomography study of a cohort of patients with hemicrania continua demonstrated significant activation of the contralateral posterior hypothalamus and ipsilateral dorsal rostral pons in association with the headache. In addition, there was activation of the ipsilateral ventrolateral midbrain, which extended over the red nucleus and the substantia nigra and bilateral pontomedullary junction. (Matharu and Goadsby 2005).

We know that sensitization can contribute to the worsening or change in the pattern of a pre-existing headache, or to the development of a new headache. Accordingly, the role of nerve injury in both initiation and maintenance of pain has been recognized in other pain disorders such as complex regional pain syndrome. (Vernadakis et al 2003).

We hypothesize that the local injury on the face of this patient and neuropathy associated become peripheral triggers for headache, perhaps due to a mechanism of central sensitization (ipsilateral dorsal rostral pons). The continuous input generated by these axons would be conducted to the second and third neurons (trigeminal nucleus caudalis thalamus and hypothalamus) and deflagrate the HC.

Conclusion

Herein, we report one case of headache fulfilling the IHS criteria for HC, presented during the course of leprosy. We hypothesize that the local injury on the face of this patient predisposes a mechanism of central sensitization, resulting in trigeminal autonomic cephalgia. Relation between TACS and leprosy provides insights into craniofacial pain mechanisms.

References

1. Garbino JA, Naafs B, Salgado MH et al (2011). Association Between Neuropathic Pain and A-Waves in Leprosy Patients With Type 1 and 2 Reactions. *J Clin Neurophysiol*. **28:** 329-332.
2. Gourie-Devi M (2008). Cranial Neuropathy in patients with leprosy. *Neurol India.* **54:** 248-256.
3. Matharu MS and Goadsby PJ (2005). Functional brain imaging in hemicrania continua: implications for nosology and pathophysiology. *Curr Pain Headache Rep*. **9:** 281-288.
4. Prakash S and Dholakia SY (2008). Hemicrania continua-like headache with leprosy: casual or causal association? *Headache*. **48:** 1132-1135.
5. Rodrigues LC and Lockwood DNJ (2011). Leprosy now: epidemiology, progress, challenges, and research gaps . *Lancet Infect Dis*. **11:** 464-470.
6. Vernadakis AJ, Koch H and Mackinnon SE (2003). Management of neuromas. *Clin Plast Surg.* **30:** 247-268.

How to cite this article : Ferreira KS, Freitas DJ and Speciali JG (2012). Is there a relation between Hemicrania continua and Leprosy ?. *Indian J Lepr.* **84** : 317-320.