Enlarging plaque on the face with enlarged supraorbital nerve

Cesare Massone, M.D.1, Andrea Clapasson, M.D.2, Sergio Gennaro, M.D.3, Enrico Nunzi, M.D.4

1Department of Dermatology, Division of General Dermatology, Medical University of Graz, Graz, Austria
2Unit of Social Dermatology, National Reference Center for Hansen’s Disease, Azienda Ospedaliera Universitaria “San Martino,” Genoa, Italy
3Department of Neurosurgery, University of Genoa and Azienda Ospedaliera Universitaria “San Martino,” Genoa, Italy
4Department of Dermatology, Universidad Técnica Particular de Loja, Loja, Ecuador

Key words: tuberculoid leprosy, supraorbital nerve, downgrading reaction
Citation: Massone C, Clapasson A, Gennaro S, Nunzi E. Enlarging plaque on the face with enlarged supraorbital nerve. Dermatol Pract Conc. 2013;3(1):5. http://dx.doi.org/10.5826/dpc.0301a05.
Received: October 23, 2012; Accepted: November 15, 2012; Published: January 31, 2013
Copyright: ©2013 Massone et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
Funding: None.
Competing interests: The authors have no conflicts of interest to disclose.
All authors have contributed significantly to this publication.
Corresponding author: Cesare Massone, M.D., Department of Dermatology, Medical University of Graz, Auenbruggerplatz 8, A-8036 Graz, Austria. Tel. +43.316.385.13235; Fax: +43.316.385.14957. Email: cesare.massone@klinikum-graz.at.

Case report

A 38-year-old Nigerian man, who had been living in Italy since 2008, presented with an annular lesion on the right temporal and frontal region that appeared two months prior to his visit. He reported a slow centrifugal enlargement of the lesion. The man complained about headache and pain radiating to the right frontal region. He was not taking any drugs. Physical examination disclosed an annular plaque of almost 10 x 8 cm in diameter (Figure 1). The margins were raised and slightly edematous, while the skin in the center was not infiltrated but dry and anæsthetic. Edema was observed around the right orbit. The supraorbital nerve was thickened and palpable (Figure 1, white arrow).

Skin biopsy showed a multifocal superficial and deep granulomatous dermatitis with epithelioid granulomas (Figures 2 and 3) with perineural distribution. The granulomas were also focally “touching” the epidermis (Figure 4). Multinucleated giant cells, discrete edema within the granulomas, and dilated superficial vessels were also observed. PAS and Grocott stains were negative. Fite-Faraco stain did not show Mycobacteria. Polymerase chain reaction (PCR) for Mycobacteria was not performed.

Figure 1. Annular anesthetic plaque with raised margins on the right temporal and frontal region. [Copyright: ©2013 Massone et al.]
Leprosy's clinical manifestations are determined by a dynamic interactionary process between *M. leprae* and cell-mediated immunity (CMI) of genetically predisposed subjects. According to Ridley and Jopling, leprosy patients are placed into a spectrum of clinico-pathological manifestations with polar tuberculoid (TT), lepromatous (LL) and intermediate types of borderline tuberculoid (BT), mid-borderline (BB) and borderline lepromatous (BL) leprosy. The spectrum is characterized by the balance between CMI and mycobacterial load: high CMI response means low number of bacilli (paucibacillary leprosy: TT and part of BT). Low CMI response means high number of bacilli (multibacillary leprosy: LL, BL, BB and part of BT) [1-6]. As Ridley wrote: “the spectrum is uninterrupted and there may be patients with an intermediate position among two groups” [5].

Anesthetic unilateral single lesions characterize TT leprosy, while bilateral asymmetrical distribution of lesions (macules, papules and plaques) characterize BT leprosy. Multiple symmetrically arranged macules and small or large nodules and plaques characterize BL and early LL. Loss of sensation is typical in TT and BT lesions, while it appears only late in BL and LL lesions. Early damage to autonomic nerve fibers impairs sweating and causes dry skin in TT and BT leprosy [1].

Leprosy reactions, divided into type 1 (called also reversal reaction or RR) and type 2 (called also erythema nodosum leprous or ENL) reactions, are severe acute episodes that are common in immunologically unstable borderline patients, and involve an up-regulation of the host response to *M. leprae* antigens. RR is more frequent in BT patients during treatment and are due to an improvement of CMI against *M. leprae* (so-called upgrading RR) [1,7,8].

During RR, all three components of the peripheral nervous system are affected: sensory, motor and autonomic.
Sensory loss causes anesthesia, analgesia and inability to discriminate hot and cold. Motor deficit causes muscle weakness, paralysis and atrophy with irreversible nerve damage, leading to impairments and permanent disability [1].

The most common nerves involved in RR are the median, radial and ulnar nerves, the sural, posterior tibial and perineal nerves, great auricular and the facial nerve. In all leprosy patients, accurate neurological examination includes testing for loss of sensation on skin lesions, palpation of commonly involved peripheral nerves, evaluation of sensory function, and muscular strength. When involved, nerves appear enlarged at palpation [1,2].

Nerve involvement can be confirmed using electrophysiology and echography or MR. RR has to be immediately treated with steroids before nerve damage occurs. Moreover, nerve involvement may be caused by compression due to edema, and the granulomatous inflammation affecting the nerve and can be treated surgically by opening the anatomic tunnel and performing an external neurolysis [1,9].

Concerning our patient, the clinical differential diagnosis included, among others, mainly sarcoidosis, skin lymphoma, granuloma annulare, tinea faciei, discoid lupus erythematosus and lupus vulgaris. A centrifugally enlarging anaesthetic plaque with dry skin in a subject coming from an endemic area favors leprosy. A unilateral anaesthetic lesion is typically seen in TT. The presence of multifocal epithelioid granulomas without necrosis but with perineural distribution confirmed leprosy and excluded tuberculosis and the other differential diagnosis. According to Ridley and Jopling the patient was classified as TT. The negative Fite-Faraco stain is not surprising because in more than 50% of TT patients M. leprae are not seen in TT biopsies because the CMI has already eliminated the pathogens. For the same reason, also PCR is unremarkable in more than 50% of TT patients, and it was not performed by us because a negative PCR does rule out the diagnosis [1,10].

The presence of multinucleated giant cells on histopathology, discrete edema within the granulomas, and dilated superficial vessels were histopathologic signs of RR [11], are compatible with the clinical enlargement of the supraorbital nerve and the headache reported by the patient. The supraorbital nerve is a branch of the frontal nerve, it is a pure sensory nerve and the headache reported by the patient. The supraorbital nerve can rarely cause headache and pain in the orbital cavity [1,12].

TT is a stable form of leprosy with a stable CMI against M. leprae. In contrast with borderline patients (BT, BB and BL), TT patients usually do not suffer from RR and do not shift to another form of the disease [1,8]. RR occurs more frequently in BT or BB and has been only rarely reported in TT [1,13-15]. Our case is interesting because the patient presented classic clinical and histopathological TT but also developed an RR of the supraorbital nerve. These cases have been previously reported as “reactional tuberculoid leprosy” or “low resistant tuberculoid leprosy.” This unusual form of leprosy is represented by a solitary TT lesion with signs of RR (edema of the lesion and enlargement of the nerve) and can be explained by a changing of the CMI, in particular with decrease of CMI against M. leprae and development of RR [1,13-15]. If not promptly and correctly diagnosed and treated, our patient probably would have progressed in the spectrum of the disease and shifted to BT with the development of more lesions and maybe even more extensive nerve damage.

Multidrug therapy as recommended by the WHO for paucibacillary leprosy was started and the patient received rifampicin 600 mg/month, and dapsone 100 mg/day. Dapsone was subsequently substituted by minocycline 100 mg/day because of the onset of anemia. The therapy was continued for 6 months. Prednisone 30 mg/day (for 3 months with progressive tapering) was added to treat the concomitant RR. Because the headache and pain did not improve with the corticosteroid therapy, surgical enlargement of the frontal notch was performed with improvement of the pain. The therapy was well tolerated, and at 10-month follow-up the plaque disappeared and the supraorbital nerve was not enlarged anymore.

Leprosy patients have to be categorized correctly before starting therapy: during RR, multidrug therapy and steroids are needed, in order to control the disease and prevent irreversible nerve damage.

References
1. Nunzi E, Massone C. Leprosy: A Practical Guide. Berlin: Springer, 2012.
2. Ridley DS, Jopling WH. Classification of leprosy according to immunity. A five-group system. Int J Lepr Other Mycobact Dis. 1966;34(3):255-73.
3. Ridley DS. Nature of the leprosy spectrum. In: Ridley DS (ed). Pathogenesis of Leprosy and Related Diseases. London: Wright, 1988:93-105.
4. WHO Enhanced Global Strategy for Further Reducing the Disease Burden Due to Leprosy (2011-2015). Operational Guidelines (Updated), 2009.
5. Talhari S. Bangkok Workshop on Leprosy Research. Diagnosis, classification and prognosis. Int J Lepr Other Mycobact Dis. 1996;64:S13-4.
6. Lockwood DN, Sarno E, Smith WC. Classifying leprosy patients—searching for the perfect solution? Lepr Rev. 2007;78(4):317-20.
7. Pfalzgraf RF, Bryceson A. Clinical leprosy. In: Hastings RC. Leprosy. Edinburgh: Churchill Livingstone, 1985:140-176.
8. Naafs B. Treatment duration of reversal reaction: a reappraisal. Back to the past. Lepr Rev. 2003;74(4):328-36.
9. Van Veen NH, Schreuders TA, Theuvenet WJ, Agrawal A, Richardus JH. Decompressive surgery for treating nerve damage in leprosy: A Cochrane review. Lepr Rev. 2009;80(1):3-12.
10. Weedon D. *Weedon’s Skin Pathology*. 3rd ed. London: Churchill Livingstone, 2009.

11. Lockwood DN, Lucas SB, Desikan KV, Ebenezer G, Suneetha S, Nicholls P. The histological diagnosis of leprosy type 1 reactions: identification of key variables and an analysis of the process of histological diagnosis. J Clin Pathol. 2008;61(5):595-600.

12. Desikan KV, Anbalagan J, Maheshwari PK. Pure neuritic leprosy of supraorbital nerve as unusual presentation. Indian J Lepr. 2001;73(1):47-50.

13. Alfieri N, Fleury RN, Opromolla DV, Ura S, de Campos I. Oral lesions in borderline and reactional tuberculoid leprosy. Oral Surg Oral Med Oral Pathol. 1983;55(1):52-7.

14. Leiker DL. Low-resistant tuberculoid leprosy. Int J Lepr Other Mycobact Dis. 1966;34(1):72-3.

15. Noto S, Clapasson A, Nunzi E. Classification of leprosy: the mystery of “reactional tuberculoid.” G Ital Dermatol Venereol. 2007;142:294-5.