Sporotrichoid Nerve Abscess in Borderline leprosy: Rare presentation of downgrading Type 1 reaction

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Summary  Nerve abscesses are seen in upper pole of leprosy as a manifestation of upgrading type 1 hypersensitivity reaction. We describe a case of borderline Hansen with sporotrichoid distribution of nerve abscesses, as a manifestation of type 1 downgrading reaction.

Keywords: Hansen; Nerve abscesses; Borderline tuberculoid leprosy; Downgrading Type 1 reaction

Introduction

Nerve abscess is an infrequently reported complication of leprosy. It is seen along the tuberculoid pole as a sign of upgrading Type 1 hypersensitivity reaction. We describe a patient of borderline Hansen with multiple symmetric nerve abscesses, as a manifestation of Type 1 downgrading reaction.

Case Report

A 40 year old female presented to our Outpatient Department with the chief complaint of having had multiple erythematous hypoesthetic plaques on her bilateral forearms and hands for 1 year. On nerve examination, the right ulnar and left common peroneal nerve was enlarged. A skin biopsy revealed well defined epitheloid cell granulomas in the upper dermis with grenz zone. No acid-fast bacilli (AFB) were visualised on Fite staining (Figure 1).

The patient was diagnosed with borderline-tuberculoid (BT) type of leprosy and started on WHO multibacillary multidrug therapy (MBMDT). The patient was compliant with her
treatment and on regular follow-ups in the leprosy clinic. Six months later the patient complained of the sudden bending of the little finger of her right hand and the onset of multiple painful swellings over her bilateral forearm. On cutaneous examination, she had multiple swellings along the flexor aspects of her bilateral forearms in an apparently linear distribution. These nodules were skin coloured to erythematous, non-tender, soft to fluctuant and extending along the course of the superficial cutaneous nerves (Figure 2).

The ulnar nerves and common peroneal nerves on both sides were significantly thickened and had multiple swellings along their course. A large fluctuant, non-tender swelling was seen on the postero-medial part of the lower two-thirds of the right arm. The right ulnar nerve was greatly thickened, slightly tender and merged indistinctly with the swelling mentioned above.

**Figure 1.** Well defined granuloma in the upper dermis with grenz zone (Haematoxylin and eosin 400x).

**Figure 2.** Multiple nerve abscesses along flexor aspects of bilateral forearms in an apparently linear distribution along the course of superficial cutaneous nerves.
There was partial mobile clawing of the right little and ring fingers accompanied by anesthesia of the right forearm and hand along the distribution of the ulnar nerve.

Fine needle aspiration cytology (FNAC) from these swellings revealed features of an acute suppurative lesion and was Fite stain positive for acid fast bacilli (AFB). Ultrasonography (USG) revealed altered echotexture (heterogeneously hypoechoic) with thickening of bilateral ulnar and right median nerves in forearm. MRI scan of the right forearm revealed multiple irregular peripherally enhancing lesions along the ulnar and median nerves suggestive of neural and perineural abscesses. A repeat biopsy from the plaque revealed features of Hansen disease in reaction (Figure 3).

Fite stain revealed multiple fragmented acid fast bacilli in the biopsy specimen (Figure 4).
The examination of rest of the nerves and sensations was within normal limits. The routine investigations including complete blood count, renal and liver function tests, urine analysis, X-ray chest and forearms revealed no abnormality. The diagnosis of Borderline leprosy downgrading with Type I reaction was confirmed and she was continued with WHO MBMDT along with prednisolone 40 mg orally daily. In view of the extensive nerve involvement, surgical decompression was performed. As the lesions started resolving, the dose of prednisolone was tapered by 10 mg/d every 28 days. She was released from treatment after completing 1 year of MBMDT and is under regular follow up.

Discussion

Bacterial parasitisation of peripheral nerves is a unique characteristic feature of leprosy resulting in formation of neural granulomas. In some cases when immunity is high, this granuloma progresses to nerve abscess, which is most commonly seen in patients with tuberculoid pole of leprosy. An exaggerated delayed hypersensitivity reaction due to the cell mediated immune response results in liquefaction of old caseous nerve lesions leading to abscess formation. The resulting abscess is, as in tuberculosis, a chronic abscess.1 Ramesh et al. opined that demonstration of the organisms in tissue smears is exceptional as the localised nerve abscess presentation is a manifestation of upgrading or reversal reaction which is more likely to be paucibacillary.2 Though nerve abscesses are rare in lepromatous leprosy due to low cell mediated immunity, multiple nerve abscesses are more common in lepromatous leprosy than in tuberculoid leprosy.3 In lepromatous leprosy, abscesses may be due to an ENL reaction in a nerve, exacerbation of existing lepromatous lesions, necrosis in lepromatous granuloma, iatrogenic or as exacerbation nodules in nerves. Nerve abscess in lepromatous leprosy could be acute or chronic in onset, with or without ENL reaction and may be clinically obvious or microscopic.1,4 Cold abscesses are associated with high immunity and the upper pole of leprosy while hot abscesses due to acute ENL are associated with low immunity and the lower pole of leprosy.3

Sporotrichoid presentations in leprosy have been reported in the past (Table 1).2,5–9 Nerve abscesses in the upper pole of leprosy are associated with an upgrading Type 1 reaction and demonstration of AFB is uncommon.2,3,5–7 Increased cell mediated immunity (CMI) results in upgrading and control of disease; decreased CMI results in downgrading and disease exacerbation. A shift toward the tuberculoid pole is called upgrading or reversal while a shift toward the lepromatosus pole is termed downgrading, both of which are aspects of delayed hypersensitivity, or Type 1, leprosy reactions. Clinically the distinction between upgrading and downgrading reactions is difficult to make and may require repeated examinations. The history, clinical examination and bacillary index indicate whether the underlying disease is upgrading or downgrading, and thus the prognosis. In downgrading reactions, necrosis is much less common, and over time the density of bacilli increases. In all patterns of leprosy, the major peripheral nerves undergo parallel pathogenesis, as the cutaneous lesions. The inflammation is similar, and the same classification system is applied. However, the density of acid-fast bacilli in nerves is often a logarithm higher than in the nearby skin.10 All the previous reports on nerve abscess were ambiguous regarding the management of nerve abscess.1–9 Medical therapy of nerve abscesses involves complete rest to the affected part, continuing anti leprosy therapy and administration of anti-inflammatory drugs with
Table 1. Summary of cases of sporotrichoid nerve abscesses reported in literature.

<table>
<thead>
<tr>
<th>Study</th>
<th>Spectrum of Leprosy</th>
<th>Treatment/past History</th>
<th>Site of nerve abscess</th>
<th>Demonstration of AFB in nerve abscess</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ramesh et al²</td>
<td>Borderline tuberculoid Hansen</td>
<td>MDT-PB X 2 months</td>
<td>Right ulnar nerve</td>
<td>Negative</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>Ramesh et al²</td>
<td>Pure Neuritic Hansen</td>
<td>Absent</td>
<td>Right ulnar nerve</td>
<td>Negative</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>Guha et al⁵</td>
<td>Borderline tuberculoid Hansen</td>
<td>Absent</td>
<td>Left ulnar and medial antebrachial nerves</td>
<td>Fragmented bacilli</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>Mehta et al⁶</td>
<td>Borderline lepromatous leprosy</td>
<td>MDT-MB X 2 weeks</td>
<td>Right ulnar nerve</td>
<td>Positive from lesion</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>Ramesh et al⁷</td>
<td>Tuberculoid Hansen</td>
<td>Absent</td>
<td>Posterior cutaneous nerve of forearm branch of radial nerve</td>
<td>Negative</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>Ghorpade et al⁸</td>
<td>Tuberculoid Hansen</td>
<td>Absent</td>
<td>Right superficial peroneal nerve</td>
<td>Globi with solid staining AFB</td>
<td>Caseous necrosis of granuloma</td>
</tr>
<tr>
<td>Garg et al⁹</td>
<td>Borderline tuberculoid Hansen</td>
<td>MDT-PB X 2 months</td>
<td>Superficial cutaneous nerves left side</td>
<td>Negative</td>
<td>Upgrading type 1 reaction</td>
</tr>
<tr>
<td>This case</td>
<td>Borderline tuberculoid Hansen</td>
<td>MDT-MB X 6 months</td>
<td>Positive with fragmented bacilli</td>
<td>Upgrading type 1 reaction</td>
<td>Downgrading type 1 reaction</td>
</tr>
</tbody>
</table>
corticosteroids for associated neuritis and early paralysis only. The nerve abscesses (except the giant abscesses) if not drained, tend to resolve gradually over months on anti leprosy therapy only. Indications for surgical intervention in nerve abscess depend on the type of abscess (hot v/s cold) and the functional state of the nerve trunk affected. Hot abscesses due to acute ENL related neuritis rarely need surgical intervention and resolve mostly with the administration of steroids. Cold abscesses management depends on nerve function: if the nerve function is clinically normal, anti-leprosy treatment is continued and nerve function closely monitored. Surgical intervention is required only if the nerve function deteriorates. If nerve paralysis is long standing or complete and unlikely to recover, surgical intervention is not required, but if nerve paralysis is of recent onset or incomplete urgent surgical intervention should be undertaken to relieve the compression on fascicles. Nerve abscesses also need drainage for cosmetic purpose and the risk of breakthrough of the skin to form a chronic sinus.

In our case repeated biopsy from the presenting lesion demonstrated a log increase in the number of bacilli, thus a manifestation of Type 1 downgrading reaction. Multiple nerve abscesses with multiple nerve involvement is uncommon in the upper pole of leprosy. Our case is unique in this regard in that sporotrichoid presentation of nerve abscesses was symmetrical and involved the multiple superficial cutaneous nerves of both forearms and legs simultaneously as a manifestation of a downgrading Type 1 reaction.

References

5 Guha PK, Pandey SS, Singh G. Sporotrichoid nerve abscess. *Ind J Leprol*, 1987; **59**: 469–471.