CASE REPORT

Lichen scrofulosorum in a patient with lepromatous leprosy after BCG immunotherapy

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Summary  Lichen scrofulosorum is a rare form of tuberculid seen in children and young adults. The cutaneous lesions are typically symptomless papular eruptions, associated with a strong Mantoux reaction, tuberculosis of lymph nodes and/or other organs or rarely following BCG vaccination. We describe an unusual case of occurrence of lichen scrofulosorum following BCG immunotherapy in a patient with lepromatous leprosy.

Introduction

Tuberculids develop as hypersensitive immunological reactions in the skin to an occult internal focus of tuberculosis or to introduction of mycobacterial antigens from outside. Lichen scrofulosorum (LS) is one of the recognized tuberculids, usually seen in children and young adults suffering from systemic tuberculosis. With the increasing use of BCG vaccination/inoculation for the prevention of tuberculosis or as an adjunct for various diseases and even malignancy, several complications have been described but LS remains one of the rarest.1 To date, there have been only nine case reports of LS occurring secondary to BCG vaccination.1–6 Recently, BCG has been reported to have a profound beneficial effect as an immunotherapeutic agent in patients with multibacillary leprosy having high bacillary load.7 It is now well established that vaccination of humans with BCG offers a degree of protection from the occurrence of clinical leprosy.8 Reports on the side effects of BCG when used as an adjunct with MDT in the treatment of leprosy are not there. We herein describe a case of lepromatous leprosy who developed LS following BCG immunotherapy.

Case report

A 33-year-old man, diagnosed case of lepromatous leprosy and on treatment with multidrug therapy (MDT-MB) and BCG, presented with 1 month history of asymptomatic, papular
eruptions over the trunk. Two months prior to this, he had received his first immunotherapeutic dose of BCG because of his high bacillary index (4+) (as our leprosy clinic protocol, multibacillary patients with BI ≥2+ are randomly allocated to receive either BCG or Mw immunotherapy along with 12 months WHO MDT MB). At the time of reporting, there were no symptoms suggestive of type 1 or type 2 lepra reaction, any systemic complaints suggestive of active tuberculosis or a family history of similar eruption.

Clinical examination revealed multiple small (1–3 mm) erythematous to skin colored mostly perifollicular papular lesions, discrete but arranged in aggregates, located over the back, chest, abdomen, arms, thighs and buttocks (Figure 1). A large Becker’s nevus was present over the left scapular region and incidentally the papular lesions did not involve the nevus. Minimal scaling was seen over the lesions on the abdomen. Routine investigations, including complete blood cell counts, biochemical profile, including liver and renal function tests, and chest X-ray were normal. The Mantoux test was positive (12 mm induration). Skin biopsies were obtained from the papular eruption and the nevus. Histology of the papular eruption revealed non-caseating epithelioid cell granulomas in the dermis with a normal epidermis (Figure 2). Stain for AFB was negative. Histology of the nevus showed increased melanin in the basal layer of the epidermis along with increased number of melanocytes.

Figure 1. Skin colored to erythematous tiny papules over the back relatively sparing the nevus.
The dermis showed melanin incontinence along with melanophages and there was no evidence of any nerve destruction or granuloma formation and no AFB was seen (Figure 3).

Discussion

LS is a rare tuberculid, initially described by Hebra in 1860, which is most commonly reported in children and young adults, in relation to pulmonary or extrapulmonary tuberculosis and very rarely after BCG vaccination. The clinical and the histopathological findings remain the same whether LS is caused by infection with mycobacterium tuberculosis or BCG. LS is reported to occur 1–4 months after BCG vaccination. It is clinically characterized by tiny, skin coloured, perifollicular papules arranged in groups; normally, they have a smooth surface but occasionally spiny projections with fine scales may be seen. Histology shows non-caseating, epithelioid cell granulomas in upper dermis and around dermal appendages. Tubercle bacilli are almost never seen in the histology specimen, neither can they be cultured. However, rarely antigen of mycobacterial tuberculosis has been demonstrated in papulonecrotic tuberculid, another type of more frequently seen tuberculid. It has been postulated that skin reactions occur as a result of activation of a latent tubercular focus by BCG. Some have reported it to be caused by hematogenous dissemination of the BCG bacilli in tuberculin-sensitive persons. However, in our case there was no evidence of tubercular infection presently or in the past. Another explanation is that the eruption might be mediated by a delayed hypersensitivity reaction to BCG vaccine antigens.

Multibacillary leprosy patients with high BI continue to harbour dead bacilli and viable persisters, which lead to immunological complications such as recurrent reactions and late relapses respectively. To achieve faster killing of viable bacilli and clearance of dead bacilli, various immunotherapeutic agents (vaccines and cytokines) are being evaluated as an adjunct to MDT MB therapy. Antigens of various mycobacteria have been observed to cross-sensitize the immune response to mycobacterium leprae (M. leprae) and this might help in augmenting

Figure 2. Dermis shows perivascular infiltrate and epithelioid cell granuloma admixed with foam cells. (H&E x 140). Inset shows higher magnification.
CMI in leprosy. Prominent among these are BCG, BCG plus killed \( M. \text{leprae} \), mycobacterium \( w \) (\( Mw \)), and Indian Cancer Research Centre (ICRC) bacillus.\(^{13}\)

BCG is commonly used as an immunotherapeutic agent in India in patients with multibacillary leprosy patients, due to its affordability and easy availability. Despite the increasing use of BCG, the frequency of generalized complications is as low as 1–2 per million.\(^1\) The complications due to BCG may be local or systemic. The local reactions are induration, blister formation, chronic discharging ulcer, development of lupus vulgaris and regional lymphadenopathy with or without suppurative drainage. Systemic reactions include erythema nodosum, erythema multiforme, generalized maculopapular eruption, exfoliative dermatitis, lichen scrofulosorum, lichen nitidus and papulonecrotic tuberculid.\(^{12,14}\)

Although severe side effects to BCG vaccination are well known, reports of LS in patients with leprosy are absent. To the best of our knowledge, our patient is the first reported case of LS occurring in leprosy, after BCG immunotherapy. One peculiar observation in our case was, the sparing of the Becker’s nevus by the LS lesions, even though there was a diffuse involvement of the back. The histology of the nevus showed features only of a melanocytic nevus, but no signs of leprosy granuloma or AFB positivity. Like nerves, melanocytes are neuroectodermal in origin and so should also be more preferentially invaded by \( M. \text{leprae} \). One of the reasons for the hypopigmentation in leprosy is direct destruction of the melanocytes by the lepra bacilli;\(^{15}\) it was curious that no such destruction was seen in the nevus, rather the disease activity was also absent. The reasons for this specific sparing are unknown.

In view of shortened MDT-MB regimens (12 months), immunotherapy with BCG, \( Mw \) or other mycobacteria may achieve a stronger foothold in future treatment of leprosy. BCG is being more commonly used due to its easy availability and low cost. Hence, more case reports on side effects of BCG including LS in patients with leprosy are likely to be seen in future.

References

8 Smith WC. What is the best way to use BCG to protect against leprosy: when, for whom, and how often? Int J Lepr Other Mycobact Dis, 2004; 72: 48.