CASE REPORT

Type 1 reaction masquerading clinically as ENL: A Case Report

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Summary  Attention is drawn to a Type 1 reaction masquerading clinically as ENL. Histology showed no evidence of ENL but suggested heightened T-cell activity (CMI), a characteristic feature of Type 1 reaction. We present a case of a 29 year old man diagnosed as lepromatous leprosy with recurrent Type 2 reaction treated with thalidomide for 2 years. The patient was referred to our institute from a teaching hospital. Skin biopsies were carried out during two separate eruptive episodes 2 months apart. Histopathology showed heightened T-cell activity, but no evidence of ENL.

Introduction

Erythema Nodosum Leprosum (ENL) is the dermal component of Type 2 lepra reaction, and occurs in BL-LL leprosy, in the presence of a high bacillary load. ENL may occur at any stage, but is most frequent during MB Multi-Drug Therapy. ENL incidence ranges from 0.7–4.6% of BL-LL cases and late recurrences occur up to 8 years after release from treatment. ENL episodes may recur an average of 2.6 times.1

Presented here is a young man with BL leprosy, who had several recurrences of skin lesions answering to the description of ENL. Two lesional biopsies obtained on two separate occasions, showed histological evidence of Type 1 reaction.

Case Report

A 29 year old policeman was referred to our institution from a teaching hospital in August 2013, for periodic fever accompanied by painful crops of small discoloured skin lesions, accompanied by nerve function impairment (NFI) and painful neuritis of 3½ years’ duration. At the time of the reference he was on 100 mg Thalidomide on alternate days prescribed by a dermatologist.

HISTORY

He was well till January 2010 when he developed painful nodules on both forearms and numbness in his right hand. A biopsy of one of the lesions done in his home town was...
reported as ‘lepromatous leprosy.’ He was given MB-MDT by a private practitioner in February 2010, which was discontinued by the patient in July 2010. The painful lesions recurred in August 2010 for which he was referred to a leprosy hospital in Mumbai where a course of corticosteroids (starting dose 40 mg) was prescribed along with MB-MDT. He was released from MDT in August 2011. During this period he experienced over five episodes of painful skin lesions, accompanied by NFI, jaundice and anaemia. At his home town hospital he was prescribed thalidomide 100 mg thrice daily from July 2011. The thalidomide dose was tapered to 100 mg once daily in May 2012.

There were two further episodes of painful nodules during the course of thalidomide, between July 2011 and August 2013, for which the dose was increased to 100 mg thrice daily.

Electrophysiological studies in May 2012 showed slowed motor conduction velocities and reduced compound muscle action potential amplitudes in the Peroneal, Tibial and Ulnar nerves bilaterally. Distal motor latencies were also increased. No sensory potentials were detected in the Ulnar and Sural nerves bilaterally. These findings were reported as ‘Sensory-Motor Polyradiculoneuropathy.’

The status of his disease just prior to the reference to us was: -BI 1·3+ with MI 0%. Repeat nerve conduction studies at the teaching hospital were reported as ‘Mononeuritis Multiplex’ involving all limbs.

On examination severe tenderness of the ulnar nerves and right greater auricular nerve were noted. The median, lateral popliteal, posterior tibial and superficial peroneal nerves were mildly tender bilaterally.

Monofilament testing showed severe sensory impairment in the distribution of all the nerves. voluntary muscle testing showed Grade 0 power in the thenar and hypothenar muscles bilaterally. A few painful erythematous crops (Figure 1) were also seen on forearms and legs bilaterally.

Skin biopsies of two painful skin lesions were carried out during two separate eruptive episodes two months apart (Figures 1 and 2).

Fig: 1

Fig: 2

Figures 1 and 2. Fig: 1 Painful crops on left forearm in August 2013 and Fig: 2 right forearm two months after in October 2013.
Results

The first skin biopsy dated August 2013 showed poorly differentiated cells and no intact bacilli (Figures 3 and 4). The second biopsy dated October 2013 showed heightened T cell activity, a few macrophages and a few beaded and solidly-stained acid fast bacilli. This picture suggested BL leprosy with Type 1 Reaction. There were no features of typical ENL in either biopsy (Figures 5 and 6).

M EDICAL INTERVENTION

In view of severe neuritis and NFI, thalidomide was stopped and a course of corticosteroid 60 mg was started. He improved gradually in NFI. However, 8 weeks later, at a corticosteroid dose of 30 mg, there was a recurrence of severe neuritis of the median and ulnar nerves with NFI. The corticosteroid dose was increased to 40 mg which relieved the symptoms.

When the tapered corticosteroid dosage reached 25 mg he again developed a few erythematous crops in the skin, accompanied by severe painful ulnar neuritis. At this point clofazimine 300 mg daily was added, tapered by 100 mg every 8 weeks, combined with 25 mg corticosteroids. Since December 2013 the patient no longer gets painful crops but there is
recurrent left ulnar neuritis if the steroid dosage is tapered below 20 mg. As of July 2014 he is on a steroid maintenance dose of 20 mg, and clofazamine 100 mg daily, for the ulnar neuritis.

Discussion

According to the literature the most conspicuous histological feature in acute ENL is the infiltrate of polymorphs in the reaction centre. In the common ‘pink node’ type of ENL this centre is usually in the sub–cutis, while more severe or ulcerating forms of reaction are associated with a more superficial, diffuse distribution of polymorphs in the dermis. Although our patient presented with clinical features suggestive of acute ENL there was evidence of Type 1 reaction, especially in the second biopsy. For rational treatment of lepra reactions histological confirmation is advisable. It is reported that the presence of the T Cell co-stimulatory molecule B7-1 is reported in ENL lesions, although its strongest expression is in Type 1 reaction.

Conclusion

Attention is drawn to a Type 1 reaction masquerading clinically as ENL. Histology showed no evidence of ENL but suggested heightened T-cell activity (CMI), a characteristic feature of Type 1 reaction.

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