

Response to Case Report: De-Novo Ulcero-Necrotic Presentation of Untreated Leprosy

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Necrotic erythema nodosum leprosum (ENL) is an atypical presentation of type 2 reaction in leprosy patients that presents with painful ulcers and systemic symptoms. Though appearing sinister, prompt initiation of multi-drug therapy and standard ENL management leads to the resolution of the lesions. As a result, we wish to raise a few points regarding the diagnosis and management of a patient with lepromatous leprosy described by Gosavi et al (2021) in the case report: "De-Novo Ulcero-Necrotic Presentation of Untreated Leprosy."

Keywords : Erythema Nodosum Leprosum, Necrotic ENL, Ulcers

To the Editor,

We read the case report- "De-Novo Ulcero-Necrotic Presentation of Untreated Leprosy" by Gosavi et al (2021). The patient presented with a history of fever, myalgia, arthralgia and ulcerative lesions. Combined with these were features of nodular infiltration and scars on the pinna, madarosis, ulnar nerve thickening and hypoesthesia, and muscle power of 4/5 in all extremities, making this a classical description of lepromatous leprosy with necrotic erythema nodosum leprosum (ENL). The authors are reporting an 'atypical' presentation of ENL in the form of necrotic ulcers. However, we would like to raise a few more points that we felt did not offer

enough clarity. Authors have termed the lesions of this patient as *de-novo* ulcers as a presentation of leprosy in the title. The manuscript then proceeds to describe a typical case of necrotic ENL, with 'pus-filled lesions' rather than '*de-novo*' ulcer as the primary lesion. The authors finally arrive at a final diagnosis of erythema nodosum necroticans, which contradicts the very title of the report. Ulcers though uncommon due to lepromatous infiltration of the skin *per se*, are observed in leprosy patients and sometimes might even be the presenting feature. Ulcerative lesions occur commonly in the form of trophic ulcers, secondary to trauma and peripheral neuropathy, necrotic ENL, also termed ENL necroticans,

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lazarine leprosy and lucio phenomenon. Among ENL's atypical presentations, necrotic lesions in ENL have been reported in up to 40% of the cases (Alakad et al 2021). They present as deep, painful ulcers with jagged edges and a necrotic slough. On histopathological examination, apart from foamy macrophages, this variant generally shows an acute necrotising vasculitis with a dense inflammatory infiltrate responsible for severe manifestations like necrosis and ulceration (Alakad et al 2021). However, the reported case did not demonstrate obliterative angiitis, constituting the sine-qua-non of necrotic ENL. The explanation provided by the authors is that the biopsied ulcer may have been in a healing stage, which is unlikely in an untreated patient of leprosy with severe ENL who had onset of disease only 15 days before presentation. A more plausible explanation would be the failure of authors to biopsy the representative area.

Necrotic ENL is generally associated with a high bacillary load (Sirka et al 2017, Bhattacharjee et al 2019). Hence prompt diagnosis and early initiation of treatment assume importance. Standard multi-drug therapy (MDT)-multibacillary regimen is the therapy of choice, along with oral prednisolone at a dose of 1 mg/kg/day in tapering doses. In this patient, the authors initiated therapy with MDT without dapsone, understandably in view of severe anemia. However, the rationale behind intravenous dexamethasone is unclear, as it does not comply with the standard WHO recommendations (Leprosy/Hansen Disease 2020). They have also not commented regarding the subsequent oral therapy for control of ENL, oral steroids, thalidomide or clofazimine and maintenance. The rationale behind using fresh frozen plasma (FFP) for necrotic ENL is nebulous. The main contents of FFP include clotting factors, plasma proteins such as albumin and anticoagulants

such as proteins C and S. It is unclear how these would have helped in the healing of ulcers, which occur due to an immune-mediated process, vascular occlusion and necrotising vasculitis. The authors have listed purpura fulminans as a differential diagnosis, which generally occurs in a setting of sepsis and disseminated intravascular coagulation (Talwar et al 2012). It, however, presents as purpuric patches that undergo necrosis and ulceration, which is not the clinical picture in this case. Another point we would like to raise is the issue of antibiotic stewardship. The rationale behind using intravenous ciprofloxacin and metronidazole is debatable, as the authors provide no evidence of bacterial culture and sensitivity profile.

In conclusion, the published case report begins with a misleading title and presents an arbitrary management strategy for necrotic ENL. Both leprosy and leprosy reactions, sometimes with unusual manifestations, continue to be prevalent in our country. Standard guidelines for the management provided by WHO should be adhered to in clinical practice (Talwar et al 2012).

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Prof R. Chavan, the corresponding author of this article Gosavi et al was given opportunity to comment on this letter but didn't do so - *Editor*