Case Report

Type II Lepra Reaction-An Unusual Presentation
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Abstract
Type II lepra reaction usually present with skin lesions. We report a 23 years old male patient presented with fever for two weeks with no visible skin lesion suggestive of leprosy and with no history of either completion or concurrent anti leprosy drug treatment was eventually turned out to be a case of Hansen’s presenting with type II lepra reaction.

Introduction
Leprosy reactions occur as sudden exacerbations of clinical manifestations of leprosy superimposed on preexisting cardinal clinical features. Type-I reactions take place in patients with borderline forms of the disease (BT, BB, BL). Type II reactions or erythema nodosum leprosum (ENL) occur exclusively in patients near the leprous end of the leprosy spectrum (BL, LL). Though lepra reactions can occur before initiation during institution or after completion of antileprosy drugs treatment, in 90% patients’ type II reactions follow initiation of therapy.1 Here we present a patient who had no visible skin lesions of leprosy, there was no history of anti leprosy drug treatment and the only clinical evidence of Hansen’s disease was in the form of thickened peripheral nerves and sensory deficit that were asymptomatic and presented to us as a patient of prolonged fever.

Case History
A 23 years old nonsmoker, non-alcoholic, non-diabetic, non-hypertensive male patient was admitted with fever for two weeks. He was referred from Health Centre with the provisional diagnosis of acute nephritic syndrome with high grade fever.

To start with he used to have fever only at night but later it became continuous with temperature surge at night when it started rising with chill and rigor to peak at 102°F to 103°F. At the same time he had body aches and aches especially in the lower limbs. He developed swelling of both legs three days prior to admission. Urine output as stated during this period was bit diminished.

He got admitted to a nearby Health Centre and was referred to us the next day. He had no major illness in the past. Others members of the family were apparently of normal health.

O/E: There was mild bilateral symmetrical pedal oedema. Neck vein pressure was not raised.

Pulse: 84/min., BP-150/90mmHg, Resp-20/min, Temp.101°F.

He had bilateral thickened great auricular nerves (Figure 1) detected when the cervical lymphnodes were being palpated and were barely visible on profile. Detection of thickened ulnar nerves followed. No skin lesions in the form of hypopigmented macules or plaques, papules or nodules facial skin thickening, loss of eye brows, pendulous ear lobes were seen.

There was diminished pain and temperature sensation over both pinna, medial aspect of both hands, and antero lateral aspect of both legs and dorsum of feet almost in a symmetric way. Corneal sensation was diminished and corneal reflex sluggish. Vibration and joint position sense were normal. Deep tendon reflexes were normal. No wasting of muscles detected. There was no lymphadenopathy or hepatosplenomegaly. Other systemic examinations revealed no abnormalities. Urine output was found to be of normal volume.

Initially he was put on inj Ceftriaxone but subsequently with the provisional diagnosis of Type II lepra reaction and after slit skin smear was taken he was put on tab prednisolone-40mg with MDT. The fever subsided promptly the next day.

Laboratory Workups: Hb-10.0gm%, Total leucocytes count -10200/cmm with normal differential count, Platelets – adequate, ESR-25 mm./ 1hr. Malarial parasites –not found, malarial antigens (P. Vivax and P. Falciparam) –negative. Serum creatinine -0.8mg/dl, urea -26mg/dl, FBS-97mg/dl. Liver function test: Total bilirubin - 0.57mg/dl,Total protein -7gm / dl, Alb-3.5gm/dl,Glb -3.5gm/dl, SGPT -105.2U/L, SGOT-89.3, Alkaline phosphatase -76.3 U/L. Blood Culture after 48 hours of aerobic incubation at 37°C showed no growth. Serum FSH-15.06mIU/ml[Ref-3.3 – 13.2mIU/ml], LH-16.17mIU/ml[Ref -3.12mIU/ml], HIV(I and II) – Neg.

Urine RE: showed trace amount of protein, no RBC, Pus cells- 3-4/Hpt. Urine Culture [aerobic] showed no growth after 48 hrs incubation at 37°C urine proteins were 29gm/3500ml.

USG Abdomen showed mild increased bilateral renal cortical echotexture and generalized urinary bladder wall thickening. X-Ray Chest was normal and ECG was within normal limit Normal Kidney function study showed abnormal nerve conduction in both ulnar, common peroneal, posterior tibial and sural nerve suggestive of axonal sensorimotor neuropathy. Skin slit smear preparation [5% H2SO4] showed presence of lepra bacilli [AFB].

Histopathological examination of excision skin biopsy specimen from right ear lobule with H and E (10×). (Figure 2 a and b): stain showed mild keratosis, hyperkeratosis, parakeratosis, grenz zone, mononuclear cell infiltrate around appendages and vessels in dermis and foam cells. Abundant collagenization and decreased dermal appendage were seen. Acute inflammatory cells were lacking. Z.N Stain showed plenty of AFB (M. lepra) with globi (large clumps) (Figure 2 c and d).

Discussion
Leprosy should be suspected when a patient from endemic areas has suggestive skin lesions or peripheral neuropathy. The diagnosis should be confirmed by histotopathology.1

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Lepra reaction are immunologically mediated acute reactions. Type I lepra reactions occur in borderline forms of leprosy (i.e. BT, BB and BL). Type II reactions (erythema nodosum leprosum) occurs exclusively in patients near the lepromatous end of the leprosy spectrum (BL, LL).

Most of the lepra reactions take place following start of antileprosy drug treatment; some of these reactions may precede diagnosis and the institution of effective antimicrobial therapy or even after completion of multi drug therapy.1,6

Type II lepra reaction or enL patients commonly present with fever, malaise and crops of painful erythematous nodules. They may experience symptoms of uveitis, neuritis arthritis, orchitis and glomerulonephritis and may develop anaemia, leucocytosis and abnormal liver function tests (particularly increased aminotransferase levels).1

Male with lepromatous leprosy often manifest mild to severe testicular dysfunction, with an elevation of leuteinizing and follicle stimulating hormones, decreased testosterone and aspermia or hypospermia in 85% of LL patients but in only 25% of BL patients. LL patient may become impotent and infertile.1

This patient had no visible skin lesions suggestive of Hansen’s disease. The clue to the diagnosis of leprosy was thickened great auricular nerve detected as an offshoot of clinical examinations of cervical lymphnodes.

Inspite of objective sensory loss over both pinnas, dorsum of both hands and anterolateral aspect of legs almost in a symmetrical fashion but no such complaints were volunteered by the patient. The skin hypoaesthesia was associated with intact joint position and vibration sense and intact deep tendon reflexes characteristics of lepromatous leprosy.2

Though there was involvement of bilateral ulnar, common peroneal and posterior tibial nerves corroborated by nerve conduction study, the almost symmetrical sensory loss in the legs was possibly due to associated involvement of cutaneous nerves in a symmetric distribution.2

Though various forms of glomerulonephritis can occur in borderline lepromatous and lepromatous types of leprosy and in type II lepra reactions1 there was no proteinuria or haematuria in this patient.1

The oedema of ankle and legs in this patient is due to increased capillary permeability and stasis and may be due to involvement of autonomic nerves.3

That this patient had features of neuritis (thickened great auricular and ulnar nerves, objective sensory loss with intact joint and position sense and preserved deep tendon reflexes suggestive of lepromatous neuropathy and corroboration sensory motor neuropathy in nerve conduction study) systemic features in the form of fever, anaemia, elevated liver enzymes (SGPT-105.2U/L and SGOT-89.3U/L), orchitis /testicular dysfunction (elevated LH and FSH), presence of plenty of AFB (M. leprae) with globi and prompt remission of fever with institution of prednisolone with MDT typified he had type II lepra reaction.

So this is a type II lepra reaction presented with prolonged fever in the back ground of no skin lesions typical of leprosy or ENL.

Though low grade fever can happen less commonly in Type I reactions, fever often profound in nature can occur in type-II reactions.1

Absence of typical skin lesion is uncommon in ENL. Paolo Fiallo, Carlo Pesce et al reported in 1995 a case of isolated erythema nodosum leprosum lymphadenitis involving the paravertebral, intercostal and cervical lymphnodes without concomitant skin involvement in a 62 year old male patient under treatment for lepromatous leprosy.4

Occurrence of type II reaction in leprosy without typical nodules of enL has also been recognised by authors of a large cohort study from Hyderabad, India.5

The local inflammatory reactions of lepra bacilli vary within wide limits. In one patient the disease may be much localised to affect one small skin area or its nerve supply. In contrast some cases show involvement of almost the whole body, so that histopathology done from any part of the skin may reveal numerous bacilli, although the patient is not acutely ill and is able to go about and work normally. The nerves may not be noticeably thickened and superficially the skin may appear normal. At any stage during invasion sudden exanematous reactions may appear, accompanied by fever and general symptoms.3

Though lepromatous patients on treatment presenting erythema nodosum leprosum lymphadenitis without concomitant skin lesions was earlier reported4 and ENL without typical skin nodules was a recognised entity5 in our case the patient manifested type II lepra reactions preceding the introduction of treatment and without any skin lesions of leprosy or of lepra reactions. A prolonged fever on presentation without skin lesion typical of leprosy was an unusual clinical feature.
Conclusion

Type II lepra reactions can have a sinister prognosis as they can affect many systems. These inflammatory reactions have characteristic skin lesions which rarely may be absent. Physicians should have a high degree of suspicion of “Leprosy with reaction” in a patient who does not have the characteristic cutaneous lesions but present with fever with varied systemic features.

References

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3. Manson’s Text Book of Tropical Medicine, 2oth.Ed.pp1025- 1043.