



Lepra Reaction and neuritis



Introduction

- Reactions in leprosy: A challenge for clinician
- Incidence: 20 % – 30 % approx
- Propensity for damaging the nerves causing physical disabilities
- Reactions / Nerve damage can occur either before, during or after treatment



Suspect reactions

- Sudden appearance of symptoms
- Inflammation of existing skin lesions or appearance of painful tender nodules
- Inflammation of nerves
- Involvement of ocular tissue
- Swelling of hands, feet and pain in small joints



Types of Reactions

Type – 1 (Reversal reaction)

- Acute hypersensitivity to antigens
- Associated with alteration in CMI

Type – 2 (ENL)

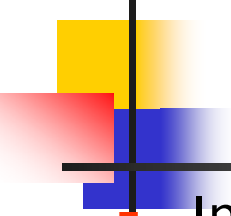
- Antigen – antibody reaction
- Immune complex syndrome



Precipitating & risk factors

- Lesion on face / Lesion on course of the trunk nerve
- Extensive disease – Multiple lesions & nerve involv.
- Inter current infection
- Parasitic infestation.
- Stress [Physical, Physiological & Psychological].
- Hormonal changes (Pregnancy, puberty / adolescence)

Clinical features of Type 1 reaction

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- Inflammation of pre-existing skin lesions with red & swollen plaques
 - Rest of the skin normal
 - Acute neuritis – Swelling with severe pain along the course of the nerve is common
 - Eyes: Ocular nerves involved
 - Oedema of extremities or face with nerve involvement.
 - Systemic manifestations are not common.
 - Occur in PB & MB
 - Usually within 6 months of MDT therapy
 - General condition fair

Type 1 reaction





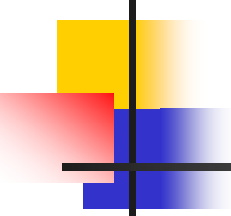
Reversal Reaction or Type 1 Reaction

- Mechanism: Delayed hypersensitivity against M.Leprae antigens
- Management: reduce the stimulating antigen with MDT, while suppressing the CMI response with steroid therapy.



Reversal Reaction or Type 1 Reaction

- Lesion on face or
- Lesion close to nerve
- Extensive disease—Multiple lesions & nerve involvement
- Inter current infection
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Erythema Nodosum Leprosum

Type 2 reaction

- Episodic: Lasting for 48 – 72 hours or a few days, poor GC
- MB leprosy: Increased bacterial load, later during Treatment
- Eruption of evanescent tender red papules & nodules most frequently occur on extremities and face
- Blanch on pressure
- Lymph nodes, liver and spleen may be painful and enlarged
- Nerves and joints may become swollen and tender
- Oedema in extremities is the common feature
- Episcleritis, Iridocyclitis, periostitis & myositis may be observed

Type 2 reaction



Pustular and Ulcerated ENL on the back





Difference : Type 1 & Type 2 reactions

Skin	Type 1 reaction	Type 2 reaction
Inflammation of the skin	leprosy patches are inflamed, but rest of the skin normal.	New, tender, red lumps, not asso. with the leprosy patches.
General condition of the patient	Good, with little or no fever.	Poor, with fever and general malaise.
Timing of presentation and type of patient	Usually early on in the course of MDT; both with PB and MB.	Usually later in the treatment; only people with MB.
Eye involvement	Weakness of eyelid closure may occur.	Internal eye disease (iritis) is possible.



Chronic ENL reactions

- Chronic ENL reactions are often troublesome to the patients and the physicians, during the course of treatment with MDT.
- Very first dose of MDT can precipitate reaction of mild nature. Occasionally severe reaction is also met with the first dose.
- It is better to give a short course and taper it .
- Steroids are not ideal treatment for ENL reaction on a long-term basis.
- Sedatives do play a favorable role.



Complications

- Arthritis
- Rhinitis
- Laryngitis
- Iridocyclitis
- Glaucoma
- Peripheral neuropathy
- Lymphadenopathy
- Myopathy
- Orchitis



Severe reactions

- Red, painful, single or multiple nodules in the skin with or without ulceration
- Pain or tenderness in one or more nerves with or without NFI
- Silent neuritis or quiet nerve paralysis
- New area with loss of sensation
- Increased or new muscle weakness noticed
- A red, swollen skin patch on the face/ overlying another major nerve trunk
- A skin lesion that becomes ulcerated or is accompanied by a high fever



Severe reactions

- Marked oedema: Hands, feet or face
- Painful &/ red eyes with/ without loss of visual acuity (Involv. of eye)
- Generalized symptoms with painful swelling of the small joints & fever
- Recurrent ENL : > four attacks in a year
- Reaction: lasting more than six months
- Mild Reaction: not responding to NSAIDs within 6 weeks.
- Enlargement of Lymph glands /testes with mild pain or tenderness
- Involvement of other organs



Treatment of reactions

Early detection of reactions
and
Adequate treatment will prevent disability !



Management of reaction & Neuritis consists of

1. Recognizing neuritis EARLY. (Acute / quite nerve paralysis)
2. Steroids
3. Splinting
4. Monitoring
5. Supportive therapy
6. SURGERY in select cases



Treatment – Reversal reaction

- Prednisolone is the drug of choice
- Action – Immunosuppressive, ↓oedema &
↓ post-inflammatory scar formation
- The duration of immunosuppressant should cover the period that the antigen load is able to trigger CMI response



Indications for steroid therapy

- Pain in the trunk nerves
- Patient with early / partial sensory loss
- Muscle paresis with or without nerve pain (Silent Neuropathy – QNP)
- Patient with red (erythematous) skin lesion or developed new skin lesions (Type-1)
- Patient with tender red spots on the skin, ulcerating nodules with swollen hands and feet (Type-2 / ENL).



Precautions before steroid therapy

Rule out the following before starting steroid therapy and refer to higher centre

- Tuberculosis
- Diabetes
- Gastritis
- Hypertension
- Nephritis
- Any infection
- Pregnancy
- Child (< 12 year)
- Eye problems
- Ulcers & osteomyelitis
- Server depression/psychosis



Precautions before steroid therapy

Start treatment for following conditions before starting steroid therapy

- Worm infestation
- Diarrhoea with blood & mucus
- Fungal infection
- Scabies
- Epigastric pain



Regimen to treat LR/N

Prednisolone regimen	Clofazimine added to treat ENL
40 mg O.D. for first 2 weeks 30 mg O.D. for weeks 3 & 4	One capsule (100mg) 3 times a day x 4 weeks
20 mg O.D. for weeks 5 & 6 15 mg O.D. for weeks 7 & 8	One capsule (100mg) 2 times a day x next 4 weeks
10 mg O.D. for weeks 9 & 10 5 mg O.D. for weeks 11 & 12	One capsule (100mg) once a day x third month

Resting Inflamed Nerves

Ulnar nerve : Elbow flexed to an angle of 90°
Median nerve : Wrist extended to 40°



Common peroneal nerve : Knee flexed to 10°



Posterior tibial nerve : Ankle in neutral position of 90°



Side-effects

- Moon face
- Osteoporosis
- Peptic ulcer
- Cushingoid
- Glaucoma
- Diabetes



General Instructions

- Not to take medicine on empty stomach
- Restrict salt intake
- Ensure treatment compliance
- **DO NOT STOP STEROID THERAPY ABRUPTLY**
- Regular NFA: Risk group
- Re-evaluate patient: who develops serious S/E
- Refer: Pain does not subside
nerve function deteriorates



Counselling

- Reason for treatment
- Duration of treatment & correct doses
- **DO NOT STOP STEROID THERAPY ABRUPTLY**
- Conditions to be reported immediately
- Follow up visit



Treatment - ENL reaction

- High dose of clofazimine (300 mg) is able to control reactions.
- Even clofazimine is not free from side effects.
- Thalidomide (a teratogenic drug) is used mainly to wean away patients from steroid dependency/steroid adverse effects.
- ENL patient should be subjected to thorough investigation to find out the precipitating cause.
- We should aim at treating the precipitating cause, rather superficially treating ENL.

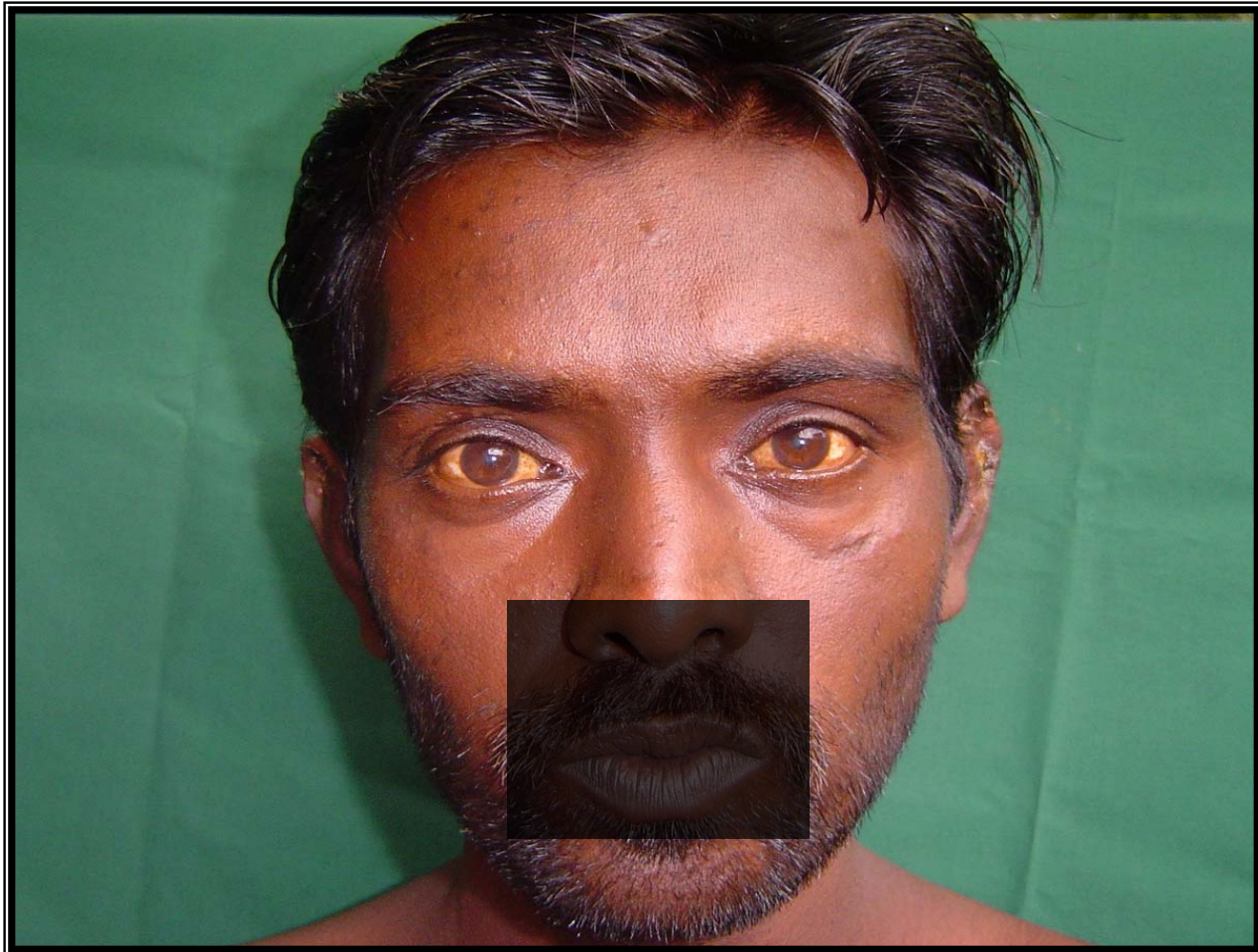


Conditions for use of Thalidomide

Women with severe ENL, can be given TLD under the following conditions

- Control of the ENL reaction: First try steroids, usually for few weeks.
- Menstrual & Sexual history must be taken to determine the pregnancy and must use two reliable forms of contraception simultaneously while on TLD.
- Give counseling regarding the dangers of thalidomide and consent should be obtained.
- A pregnancy test may be done after an appropriate time since the last sexual contact.

Severe Type 2 reaction



Swelling of face, hands & feet





Before and after treatment



Type – 1 Reaction

Response to steroids and Clofazimine

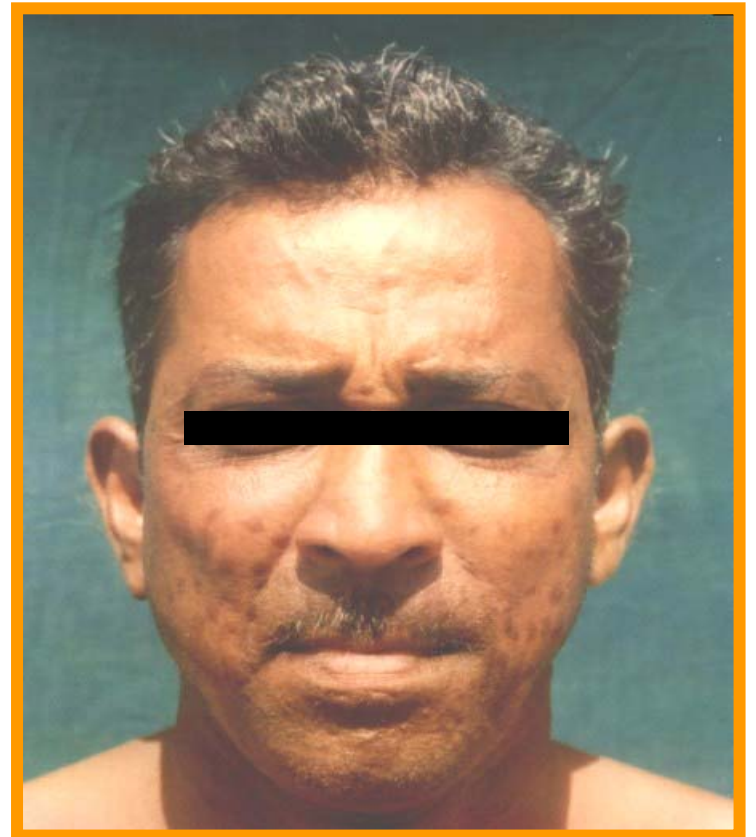


Type – 1 reaction



Type 2 reactions

Treated with Prednisolone





Referral

■ Before Starting treatment

- Systemic illness
- Nerve abscess
- Ocular involvement
- Type 1 reaction occurring after MDT therapy

■ During Steroid Therapy

- No improvement in condition in 2 weeks
- Worsening of NFI/ reaction
- Complications not manageable at PHC
- Deterioration in general physical condition



Referral

- After Steroid therapy
 - Lag - opthalmos
 - For reconstructive surgery
 - Rehabilitation



Differential diagnosis of Type 1 Reaction

- **Palmar Abscess:** when more is warmth, tenderness and oedema of the extremities
- **Erysipelas:** An acute inflammatory condition of the skin caused by streptococci. Penicillin is rapidly effective in this condition
- **Cellulites:** An acute inflammatory oedematous condition of the sub-cutaneous tissues due to Streptococcal infection-responds to penicillin



Differential diagnosis of Type 1 Reaction contd..

- **Angioneurotic Oedema or Giant Urticaria** – It is an acute evanescent circumscribed oedema that usually affects the lips, ear lobes and the mucous membranes of the mouth. The treatment is similar to that of Urticaria.
- **Cutaneous Lupus Erythematosus or D.L.E.** – In severe reversal reaction the skin lesion can ulcerate, as a result of which there may be scarring and atrophy of skin. It resembles Cutaneous Lupus Erythematosus.



Differential diagnosis of Type 2 Reaction

- Erythema Nodosum
- Iritis
- Orchitis
- Arthritis
- Lymphadenitis
- Erythema Multiforme

Standard steroid schedule - WHO

