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Leprosy

SIG Leprosy(IADVL Academy)

(ACAD Discussion 2022)

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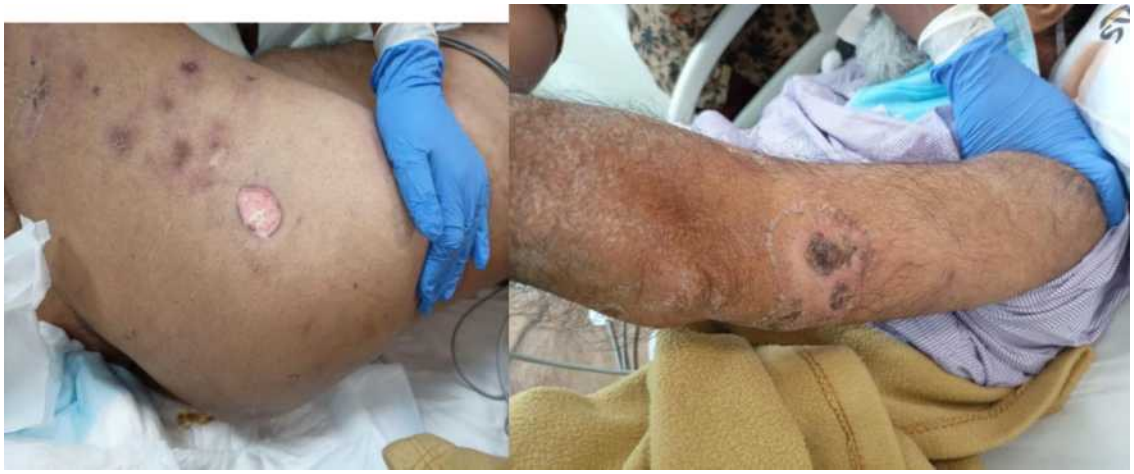
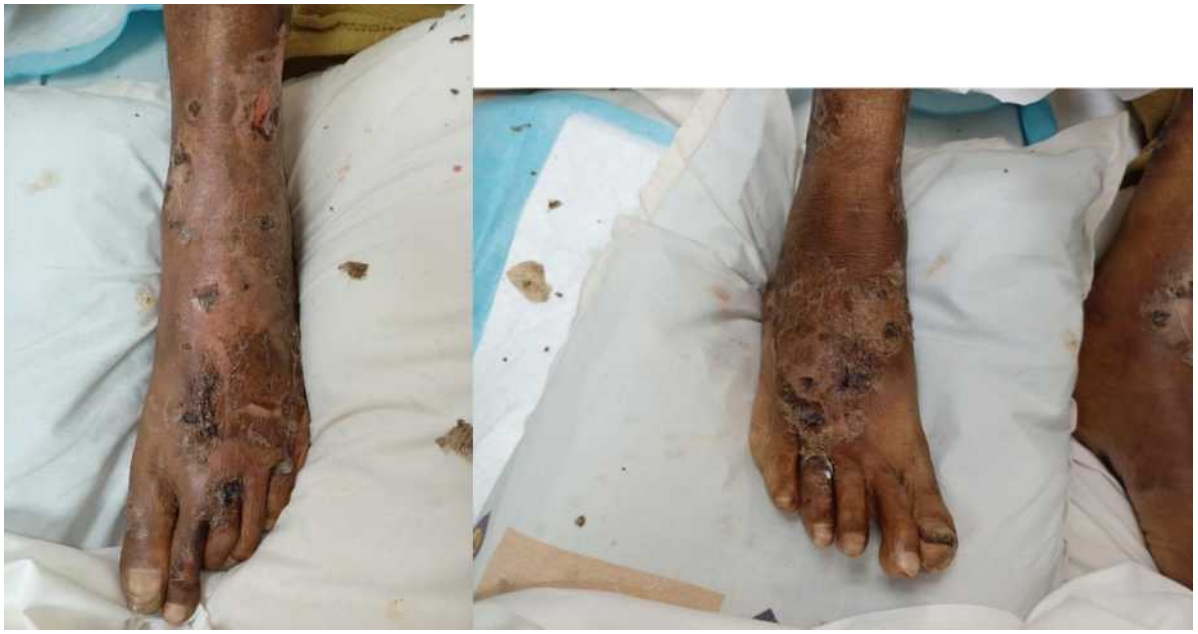
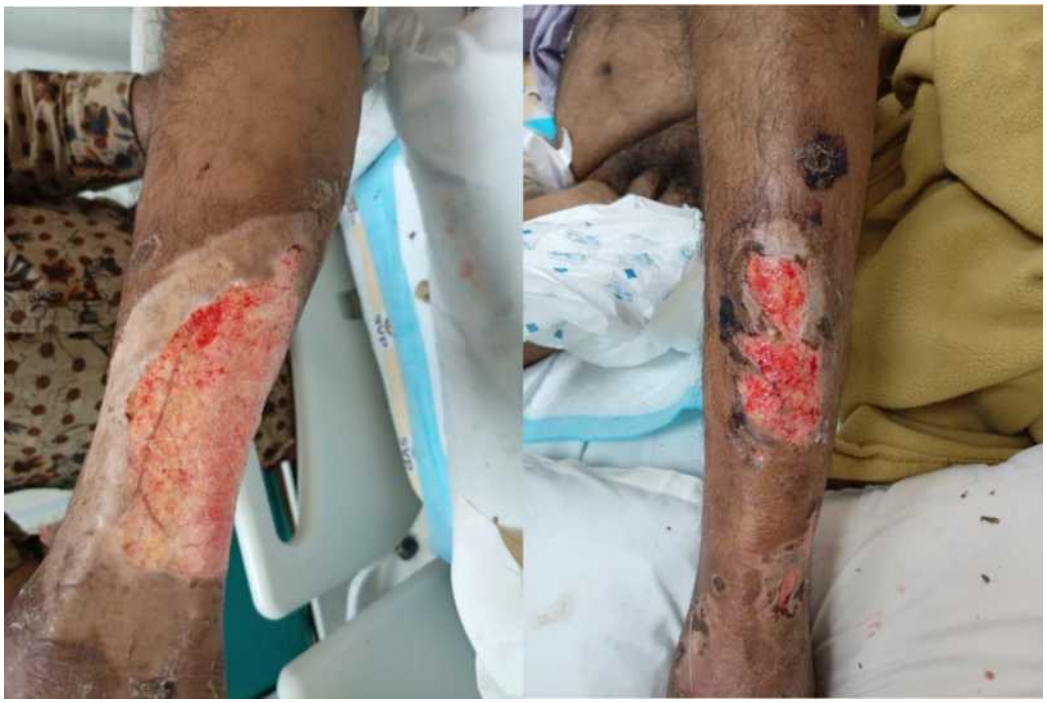
CASE 1: DR POOJA AGARWAL

- 66 yrs old male ; admitted due to COVID 19 pneumonitis, Referred i/v/o reddish lesions over malar area, Raw areas over right elbow and left leg, Healing raw area over right big toe and K/c/o DM
- Since 2 yrs patient had tingling sensation in both lower limbs and arms , f/b swelling over left leg which subsided with treatment
- No h/o fever/ joint pains at this point of time
- After 5-6 months- again developed redness and swelling over both legs, F/b development of reddish fluid filled lesions over legs
- Admitted at govt hospital - Rx was given & lesions subsided in a span of 2 months
- Developed bleeding from the nose and few raw areas without any preceding history of trauma
- associated with mucoid discharge

- Biopsy was done from the nasal mass, focal necrosis , many neutrophils with foci of microabscess, lymphocytes, plasma cells , macrophages along with with few ill formed epithelioid cells & multinucleated histiocytic giant cells .
- No well-formed epithelioid cell granuloma seen.
- AFB seen on ZN stain
- IMPRESSION = necrotizing acute and chronic inflammation s/o tuberculous etiology
- Pt was started on ATT but he left
- Differentials: AICTD with vasculitis, Covid induced vasculitis
- Investigations: CBC- Leucopenia
- RFT/LFT- WNL
- S ANA by IIF- Negative
- C ANCA - Neg (Yes we also thought of GPA!)
- Repeat workup for TB-
- Sputum For AFB = NEGATIVE
- GENXPERT = NEG
- 10 days later: Repeat referral i/v/o Pt developing multiple erythematous lesions over UL/lower abdomen/thighs/ buttocks







- New erosions over legs
- h/o joint pain in hands, elbow, and feet present
- No H/o slippage of footwear
- No H/o unnoticed trauma
- No h/o dropping things from hands
- No h/o sharp shooting pain over limbs

- No H/o contact with a person taking blister pack
- On examination: Peripheral nerves- not enlarged
- Temperature -Altered in glove and stocking distribution
- Crude/fine touch- intact
- New differentials: Erythema nodosum, Necrotic ENL
- Investigations: SSS from ear lobe & normal skin- Globi seen!
- HPE of erythematous plaque over the thigh-
- Epidermis is thinned with flattening of rete ridges.
- Superficial dermal vessels show congestion and dilatation.
- Lymphohistiocytic infiltrate collection is seen around the adnexa and superficial blood vessels.
- Deep dermal inflammatory infiltrate chiefly consisting of polymorphs and foamy macrophages are seen. Necrotizing vasculitis around the deep dermal vessels extending up to the subcutaneous fat is seen.
- AFB STAIN : Acid fast bacilli are seen
- Findings are suggestive of *Erythema Nodosum Leprosum*
- Final diagnosis: LL Hansen with Necrotic ENL

Dr Rajyalaxmi Konathan: its very interesting case. Thank you for posting. We can think of:

1. Necrobiosis lipoidica diabeticorum

2. pyoderma gangrenosum especially leg and elbow lesions

3. Hansen's disease in reaction with trophic ulcers
4. Vasculitis and connective tissue disorders.

Dr Ashok: To me it looks like a case of Necrotizing vasculitis. Were there any systemic symptoms?

Was the autoimmune profile- specially ANA,DsDNA pANCA,cANCA done? Through general medical check up should be done.Examine the lymph nodes,Urine tests, throat swab.ASO Titre, MT,CXR.

Do viral scan- HIV,HBsAg,HCV,Covid, Tb Gold, PsA,CEA, USG Whole abdomen to look for any SOL.

Smear from nose n ear lobules for AFB stain and a deep wedge biopsy from the margin of an recent ulcer which should include the ulcer wall and the base.

Dr Kisalay:

Let me try to explain why I considered Leprosy or to be more specific Lucio Phenomenon as my first d/d:

1. The patient developed spontaneous ulceration without preceding palpable lesion (papule or nodule)
2. There is a history of hemorrhagic blisters on the lower extremity.
3. The ulcers were at different stages of development. The ulcers particularly on the elbow had a geographic margin.
4. There was no associated systemic symptom related to skin finding.
5. The red face (beautiful leprosy?)
6. The histopathological finding of necrosis with neutrophilic microabscess with epithelioid cells and AFB

All these led me to think of the Lucio phenomenon as my d/d. ENL necroticans were not considered due to absence of systemic features, absence of nodular lesion preceding ulceration and absent history of MDT MB.I wanted to know whether the AFB were present inside the endothelial cell(the report mentions presence of AFB without mentioning their location.), and whether there was a feature of endarteritis obliterans. That is why I wanted to see the H/P pictures.

Biswanath Behera:

I am thinking of a vaso-occlusive episode ? Heparin-induced skin necrosis or Cryoglobulinemic vasculitis
In addition, will rule out an associated atherosclerosis as the prior ulcer was on the tip of the toe It will be better, if we can see the pathology images
The coagulation profile of the patient and the treatment received may be posted

Shyma prasad:

There are some important lessons from this case:

1. A 2 year history of tingling and numbness with impaired sensations in a glove and stocking distribution and a possible neuropathic ulcer of the toe should warrant an SSS from earlobes, as an imperative, in India
2. Acute presentations in an otherwise chronic disease can mislead spectacularly, especially in the setting of Covid.
3. A good history and examination with an SSS at the beginning itself would have obviated the need for all those expensive investigations.

Kisalay:

1. Was there any nodule or plaque?
2. Was there any systemic feature?
3. Was there any feature of neuritis?

If the answer to the questions is No, Lucio Phenomenon should be the diagnosis in place of ENL necroticans.

Attaching a file to look at the difference between the two entities.

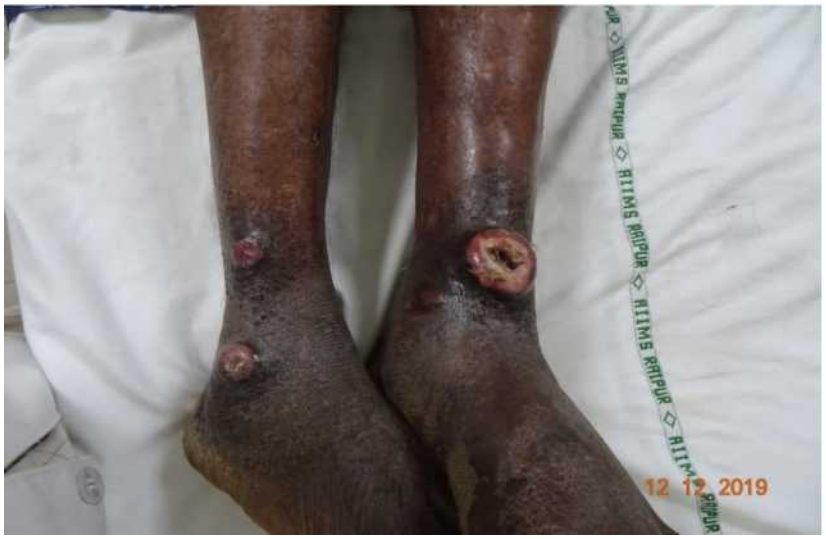
I still feel the diagnosis is not ENL; It is Lucio Phenomenon.

The new lesions which appeared were erythematous plaques and nodules which had Evanescence also. USG scrotum was done which showed epididymitis. No evidence of neuritis.

As the pt had COVID-19 pneumonitis other systemic features may be attributed to that only.

CASE - 2 DR NAMRATA CHHABRA

A 53 year old male presented with multiple hypopigmented lesions all over body, an infiltrated plaque and single shiny papule over the lower back, hard subcutaneous nodules and large ulcerated nodules around both elbows and ankles trunk since 3 years. C/O loss of sensation over back of foot for 1 year. H/O receiving MBMDT 10 years back for few months



Biswanath Behera:

There are many ill-defined copper-colored small macules (although blurry in the images), well-to-ill-defined annular plaque, papules, and nodules with and without ulceration. In addition, the papules and nodules are located around the joints.

My diagnosis will be Subpolar Lepromatous Leprosy

Xanthoma-like presentation is well known in LL and translucent papules can be seen in LL

Dr Pooia:

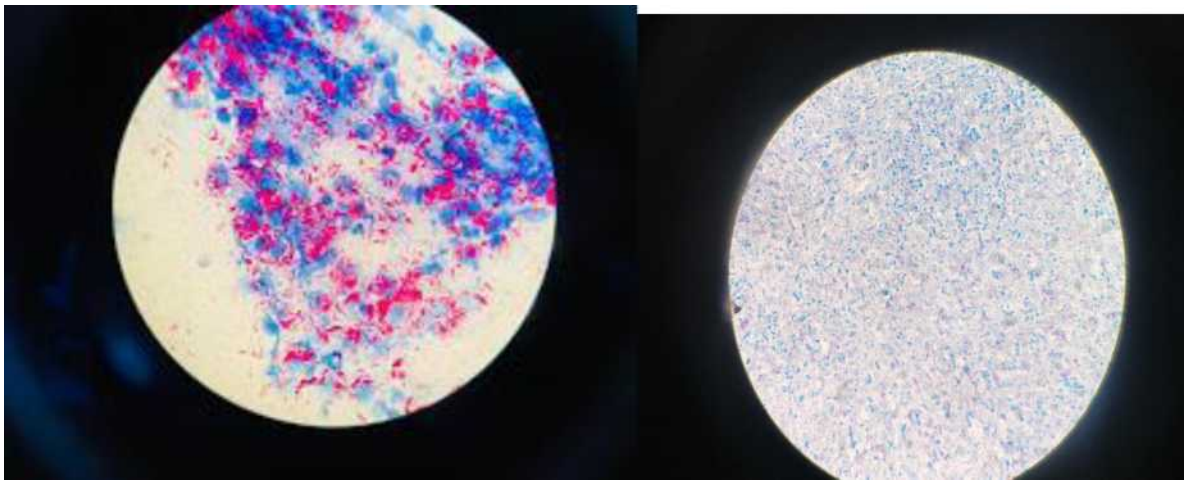
Looking at the history and clinical pictures my First impression would be Histoid Leprosy. Though written in books as a rare presentation, I have never seen ulceration over Histoid nodules or plaques till date. Can you please post SSS and HPE findings.

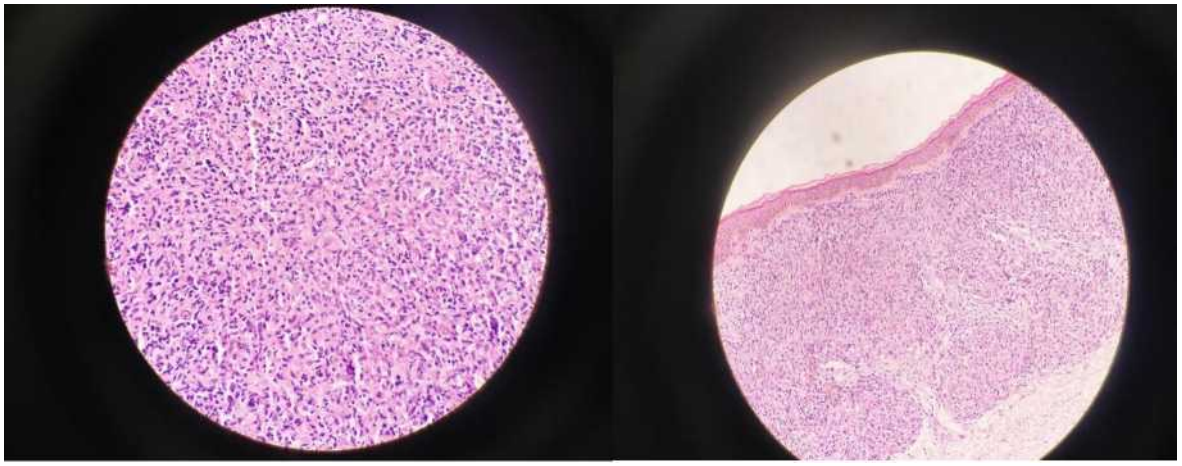
Dr Namrata chabra: that is a good differential Dr Pooja seeing the succulent papule at back, and clustered nodules near joints, although presence of small hypopigmented lesions all over body were towards LL.

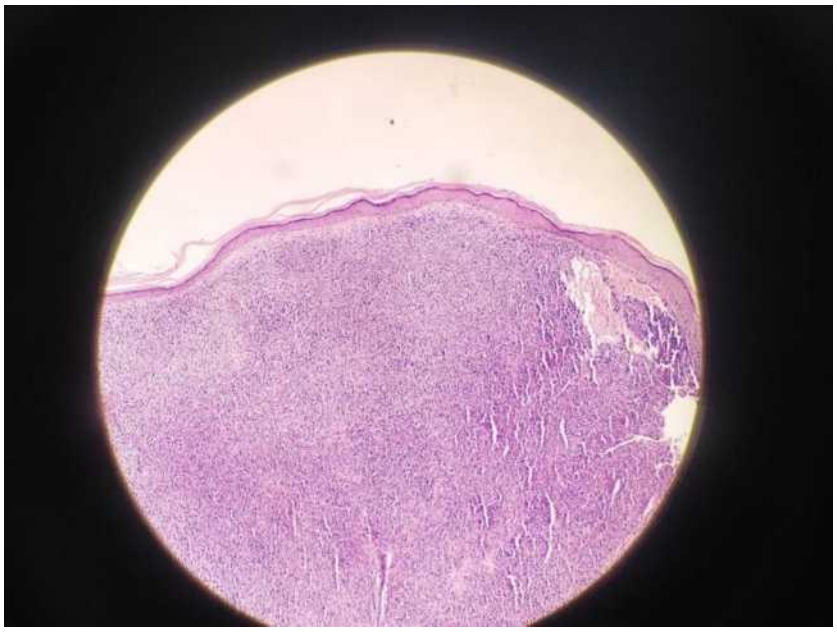
On nerve examination, multiple symmetrical thickened peripheral nerves. We kept a differential of LL Hansens with histoid like lesions. However, elderly patient with large hard nodules with ulceration and We also had a dd of Cutaneous lymphoma

Dr Shafgufta: In lepromatous leprosy, Xanthalesma-like lesions, Molluscum-like, and lepromatous leprosy with ulcerations have been reported, even when the patient is not in reaction. So, i would keep this as a case of Borderline Lepromatous to Lepromatous leprosy, with nerve abscess around the elbow

Dr Shyam: Histoid leprosy with? Dystrophic calcification/? Tumoral calcinosis







The HPE showed diffuse proliferation of spindle shaped histiocytes arranged in fascicles, whorled pattern. Special stain showed a heavy load of intact bacilli.

the HPE features from the back lesion as well as ulcerated nodules revealed similar features suggestive of Histoid. I would like the members to give their opinion regarding the diagnosis where features of Histoid leprosy and LL Hansens were present at the same time. Based on the scenario if the patient had already completed MDT 10 years back, or if there was irregular / incomplete MDT (patient history was not reliable). Sir, we have planned to give 2 year MBMDT and BI from nodules and intervening skin were both 6+.MI as shown in pic is high up to 20-25%

Kindly go through the marked area of this H/P picture

Kisalay: 1. The two larger circles seem to be well-circumscribed macrophage granuloma with a few spindle shaped cells

2. The smaller circle seems to contain a Langhan giant cell in center The slit skin smear shows Globi .

Dr Tarun Narang: Both these features are unusual for de novo histoid but in the case where we are also seeing lesions of borderline leprosy both these can be seen/ observed.

Tuberculoid contamination, a peculiar phenomenon may be seen in subcutaneous histoid lepromas. This is the occurrence of definite foci of epithelioid cells, located within the lesion substance or in the encircling fibrous tissue encapsulation. No satisfactory explanation of this occurrence is postulated, but it is likely that the epithelioid cells represent the tuberculoid component of the earlier borderline phase of the disease.- (IAL Textbook of Leprosy). Globi have also been described in histoid leprosy.

Dr Namrata:

the globi visible on SSS can be explained as it was taken from ear lobe and the infiltrated annular plaque S/O LL.However, the spindle shaped histiocytes and their pattern of arrangement s/o histoid from large nodules and the succulent papule from back.

we did not do SSS from histoid lesions, in biopsy special stain for AFB, our pathologist could show us some elongated bacilli with tapering ends, but my experience to differentiate the two is limited.

Dr Kisalay:

1. These cells in the granuloma are not epithelioid cells and are macrophages only. Textbook of IAL has mentioned epithelioid contaminants and not contamination by macrophages.

2. In the textbook of IAL, epithelioid contaminant has been mentioned along with Histoid Habitus: the parallel arrangement of lepra bacilli that is missing in SSS of this case; instead Globi are seen. In H/P, the AFB-stained slide is a little blurry.

Dr Tarun Narang:

these macrophages are the predominant cell type in histoid leproma, and three types of macrophages are described:

1.Ordinary macrophages

2. Fusiform or elongated macrophages

3. Foamy macrophages

All these are loaded with bacilli and in addition to this, transitional forms have been also seen which do not contain bacilli and may depend on the stage of evolution of the histoid leproma.

Depends on the type of lesion that the biopsy may have been taken.

(Jihe L, Qingying K, Gan-Yun Y, Mingyu X, Li H, Luenyuan Z. Observations on ultra-structure of histoid leproma. Int J Lepr Other Mycobact Dis. 1982;50:471-4.)

Dr Biswanath Behera:

the histopathology pattern of the granuloma is not expansile but rather subepidermal band-like and nodular (Need a scanning power to clarify). There is no pseudocapsule. There is no whorled or fascicular pattern of growth. In addition, the presence of spindle-shaped histiocytes in the provided pathology image is doubtful.

Dr Shyam Prasad:

Morphologically, this case has certain unique features which deserves a more permanent kind of documentation otherwise than in the ACAD group.

1. Co-existence of hypopigmented macules, smooth shiny nodules, infiltrated plaques, and large subcutaneous hard tumours, all in one patient.

2. While large dermatofibroma like or neurofibroma like lesions have been reported in histoid, the bigger subcutaneous tumours here appear visually different.

3. Ulceration of histoid nodules is also quite rare.

3. All lesions histologically leprosy

4. Good response to MDT even of the larger lesions, suggesting that they were not tumours of a different sort. Incidentally, how does an epitheliod cell differ morphologically from a macrophage, on light microscopy and H&E stain, if there is no vacuolation or foamy cytoplasm?

Dr Kisalay:

Given the super bacillated state, in my experience any form of WHO MBMDT with monthly rifampin typically fails. **Monthly** moxifloxacin and minocycline in **addition** to 2 year MBMDT would be much better. As is the case, highly bacillated lesions flatten very quickly to any anti leprosy treatment but have a high rate of treatment failure. These cases are also a high risk for patient being lost to follow up as individual lesions respond early. Is there a family history in this case?

The difference between macrophage and epithelioid cells are as follows:

Cell margin of macrophages are well maintained. But cell margins of epithelioid cells are indistinct and adjacent cells are fused to produce a symplasmic mass; pale blue vesicular nuclei of multiple cells drift like a handful of boats in the ocean of light eosinophilic cytoplasm. This gives them the epithelial cell-like appearance and hence called epithelioid cells. Due to the large size of individual cells, the distance between the nuclei of adjacent cells is also large(hence the "a handful of boats in large ocean" analogy). As a rule, the term epithelioid cell is never used as a singular number.

Dr Namrata:

Adding monthly Moxi and Mino would not add much to the cost too. There was family history suggestive in this case

Dr Tarun Narang:

All these options have been tried or are being used in patients with a high bacillary load, but there is no published literature that has shown any benefit. Immunotherapy with BCG and MIP is another option, but the question is how long should you continue MDT? because we have seen patients who had active lesions and a BI of 4/5 even after 2 years of MDT and immunotherapy. Although MI was zero, we found that the viable bacillary count by RT PCR was still high.

Merely adding monthly minocycline and moxifloxacin may not be enough in these patients. Hence some leprosy experts advise MDT or monthly ROM or PMM regimen till smear negativity.

What is the House's opinion about prolonging treatment till smear negativity, especially in patients with histoid leprosy or polar lepromatous leprosy?

Dr Ashok:

Whereas we may not have to wait for smear negativity to stop the treatment in Polar Lepromatous leprosy after 1yr of MDT therapy because immunity is rather stable in this spectrum and it is assumed that body immune system will take care of the remaining bacilli. But in Histoid Leprosy we should not take that chance because of the low immune status here and we should continue the treatment to smear negativity.

Dr Tarun Narang:

Polar lepromatous leprosy patients are also anergic and the immunity is not able to take care of bacilli in these patients as well, hence we should give MDT for a longer time in these patients as well. Moreover, majority of these patients have chronic/ recurrent ENL and are on long-term steroids which is not seen in patients with histoid leprosy. hence these patients also need a longer duration of treatment. Long-term steroids should also impair immunity in these patients and that would effect the immunity taking care of the remaining bacilli after 1 year of MDT.

Dr Ashok:

Type 2 reactions more common in Subpolar Lepromatous Leprosy rather than Polar L. But as you said host immunity is less in LL as a whole, this concept of giving MDT for 1yr in multi bacillary leprosy was recommended by WHO with the idea that the remaining bacilli would be taken care of by host immunity. This is what WHO told. But in my practice, I usually continue MDT for 2 yrs/ smear negativity which one is later to be on safe side. WHO even recommended 6 months MDT for paucibacillary Leprosy , even if the granuloma persists in biopsy. They told the granuloma will regress slowly after stopping MDT. TRUE or Philosophical? The duration of therapy I decide on case to case basis.

Dr Tarun Narang:

I agree the duration of treatment is debatable, that is why I wanted to have views of the house on this issue, WHO guidelines and the recommended duration of treatment work well in most of the patients but it is inadequate as far as patients with high bacterial load or high BI(>3) are concerned.

And in recent paper, it was observed that 55.5% to 66.6% of cases with a BI 3-4, showed viable bacilli at the time of completion of 12 months of MDT treatment. And these were also showing signs of activity on histopathology. *(Singh I, et al. Efficacy of fixed duration multidrug therapy for the treatment of multibacillary leprosy: A prospective observational study from Northern India. Indian J Dermatol Venereol Leprol doi:* 10.25259/IJDVL_915_2021)Unfortunately, we don't have recent studies which give information about the number of viable bacilli being shed by these patients, which makes them an important source of transmission despite being declared 'CURED'. Plus the morbidity and sometimes mortality due to ENL and the adverse effects of long term corticosteroids.

I think even lepromatous leprosy patients or all those with BI>3 should be offered a longer duration of treatment, minimum 2 years but can be extended based on SSS report at the end of treatment.

Dr Sunil Ghate:

Highly bacillated cases need longer duration. Minimum 2 years for BL-LL irrespective of the smear status. There is no facility to do so in many places. One can go by clinical assessment alone.

If the MI is positive at the end of Rx, it may be extended further. It's only a small pool of highly bacillated cases who need to given longer duration.

I would like to know whether giving rifampin daily would make a difference in these highly bacillated cases. In my opinion, it would be beneficial. I don't like the idea of monthly pulse. It (Rcin pulse) was tried as a pulse in late 70s early 80s before it became part of MDT. The circumstances at that time were different. We need systematic studies like THELEP now to prove or disprove it. We all know very well that dosing a drug monthly May promote resistance. Rifampin is an important component of ATT.

Dr Tarun Narang:

No evidence of daily rifampicin but there are a few studies that have shown monthly ROM or PMM to be as effective as MDT. I am attaching an old editorial on the treatment of leprosy, I found it very interesting. The National Hansens disease Program in USA use daily rifampin along with dapsone and clofazimine to their patients with hansens disease.

Dr Shagufta:

In kashmir, shortage / non availability of MDT is an issue in GMC Srinagar where most of these cases are diagnosed and enrolled.

Once diagnosed, we ask these patients to get MDT from their respective district hospitals or from Director health office.Patients have to go to many places to manage their treatment. This ultimately leads to delay in treatment or contributes to default to treatment.

Moreover, the medical officer at the district level strictly tends to adhere to guidelines, and would not issue the MDT after one year of completion, if we write or even request him that we need to extend treatment further in a highly bacillated case.Kindly advise: how can we get a free flow of MDT supply in GMC Srinagar, so that our patients get uninterrupted treatment.

Dr. Tarun Narang:

We can at least try it in our multibacillary cases with a BI>3/4, and see the response as well as the relapse rate over the next few years. And as Dr PN Rao Sir mentioned, a RCT is required to see the response, relapse and adverse events with daily rifampicin.

But the problem with such studies is that we need a long follow-up of at least 5-6 years to observe the relapse rate and most of the patients do not follow up for such a long period. Plus Funding agencies are also not very much interested in longer duration projects.

CASE-3 DR SWETALINA PRADHAN

A 45-year old male from Chhapra, Bihar presented with sudden onset of multiple vesicles which within 2 to 3 days developed crusting, associated with fever.Patient had 2 episodes of painful nodules with fever 8 months back which subsided without any treatment.There was no history of any hypopigmented skin lesions. There was h/o intermittent epistaxis.H/O slippage of foot wear was present.

Examination: There were few vesicles, Majority of the lesions had developed crusting with central

umbilication(varioliform), There was bilateral pedal edema.

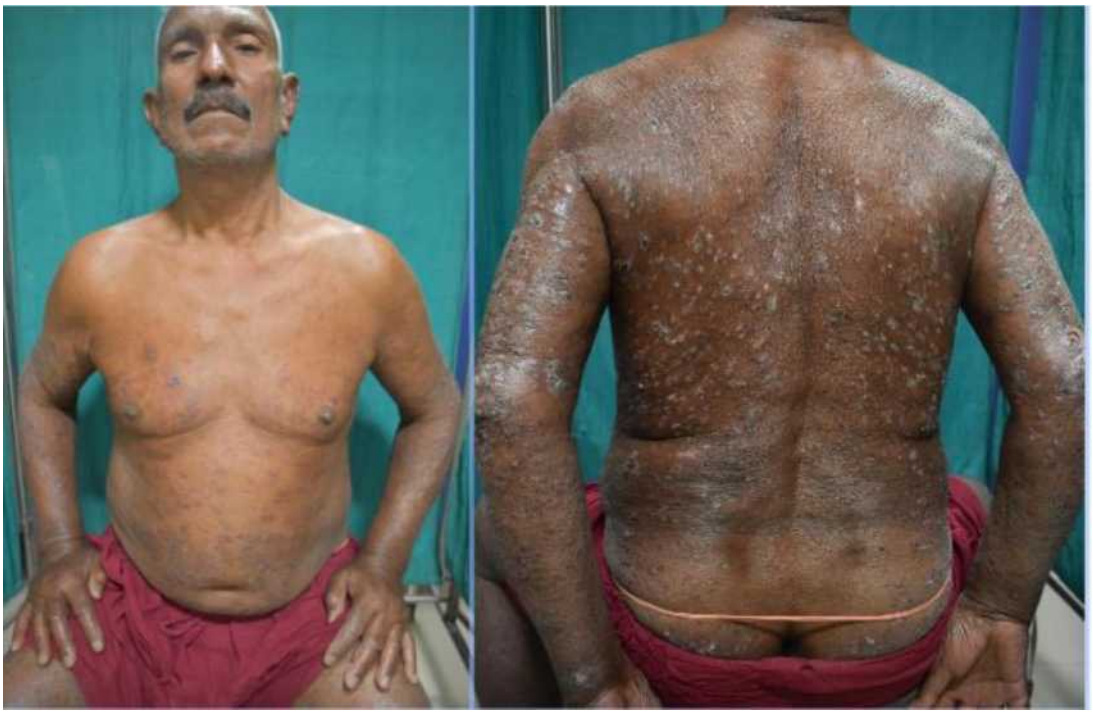
Mucosa: Normal

Bilateral ulnar nerve and Right CPN were enlarged Differentials:

VARICELLA

PLEVA

Papulonecrotic tuberculid



This is a case of Hansen's disease with blister formation(crusting is a secondary feature of bullae);

Theoretically, blister appears in leprosy in the following conditions:

1. Blister as a presentation of trauma/burn on a neuropathic area(not your case)
2. Erythema nodosum leprosum(your case)
3. Lucio pheno(presence fever and nodules in your case rule out this diagnosis)
4. Lazarine leprosy(I don't know whether we use this diagnosis any more or not; but even if we do, the lack of large ulcers rules out this diagnosis)
5. Bullous drug reaction in leprosy(a drug history would be helpful)
6. Co-existence of leprosy and AIBD(reported by Dr Thappa once, if I remember correctly)

Dr Kisalay:

Considering the morphology, I would prefer ENL as my provisional diagnosis. Will wait for SSS(from blister fluid) and histopathology

Dr Swetalina:

This patient had sudden onset of vesicles which subsequently developed crusting(varicella like). He had history of painful nodules in previous episodes . We clinically suspected varicella . However the Histopathology was suggestive of ENL. The patient had infiltration at places.

Dr Narasimha Rao:

In patients of Florid LL, due to extensive hypo aesthesia, cutaneous responses to various stimuli can be abnormal or exaggerated. (as vascular sympathetic tone is decreased). This could be just a severe reaction to insect bites or can be considered as exaggerated Papular/ vesicular reaction to similar

injury/ insult. A biopsy from an early lesion would be Interesting. A dose or two of ivermectin would be good. We can also Check for eosinophilia.

Dr Anantha:

A possibility of eczema herpeticum may also be considered if the patient has been on prolonged steroids for ENL previously. Or even disseminated and visceral varicella. Seen a fatal case of the latter in a leprosy patient on chronic steroids. A tzanck form vesicles could also be done to rule this out.

Dr Pooia:

My first clinical impression also was Kaposi varicelliform eruption like Dr Ananta mam. Though usually reported in conditions like PF/ AD, may be the diffuse skin involvement with irregular treatment of leprosy predisposed the person to same.

Dr AG:

1 Was there any mucous membrane lesion?

2 Was there any pleomorphism noted in evolving the blisters?

3 Any h/o contact with Varicella?

4 Was Tzanck test done?

5 Virus culture [if facility available]?

6 Did the bx include the blister and the deeper tissue?

7 Was the pt on Dapsone,Lasix or penicillamine or any other drugs which cause bullous reactions

8 Did the blisters heal spontaneously without treatment?

9 Was there any other symptom other than fever- eg arthritis,prostration,eye irritations-as found in ENL

Answers to these very many Qs may give clue to this interesting case.

ENL presenting with tens & hundreds of blisters{ and without any other systemic symptoms} --'which subsided spontaneously'-v v unlikely.

Insect bite reactions as suggested by Dr Rao is possible,but not probable.

Lesions are too many and they are not only on limb but on body and covered parts also.

Drug reactions [if the pt is on any-not mentioned] possible but then again should not heal unless the drug is withdrawn. Varicella and Kaposi are two strong possibilities but to be substantiated by thorough work-ups, especially when the pt is from a top academic institute like AIMS.

If it's Kaposi Varicelliform eruption, then it should be a really be a very rare case indeed.

Dr Shagufta:

Drug Rash

Kaposi's varicelliform eruption

Varicella (Should not subside in two to three days in a middle-aged man)

Some kind of vesicular rash due to Covid-19 infection.

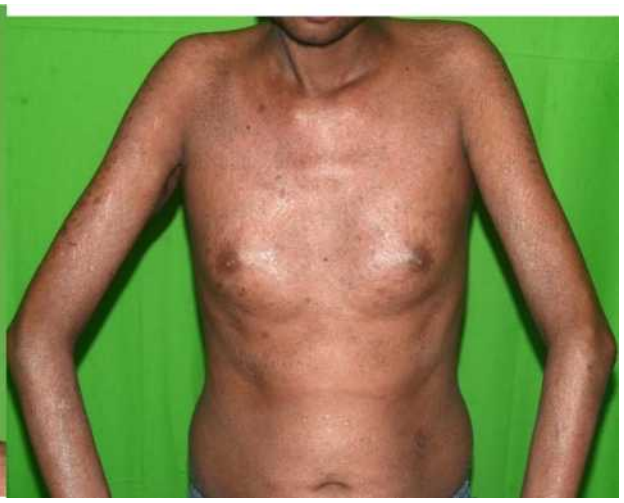
Vasculitis, as a presentation of lepra-2 reaction

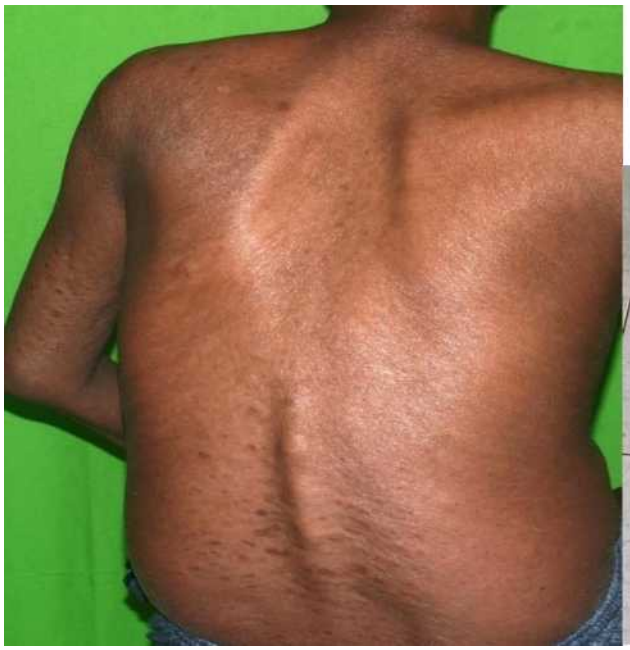
Irritant contact dermatitis? history of application of any indigenous preparation to treat lesions?

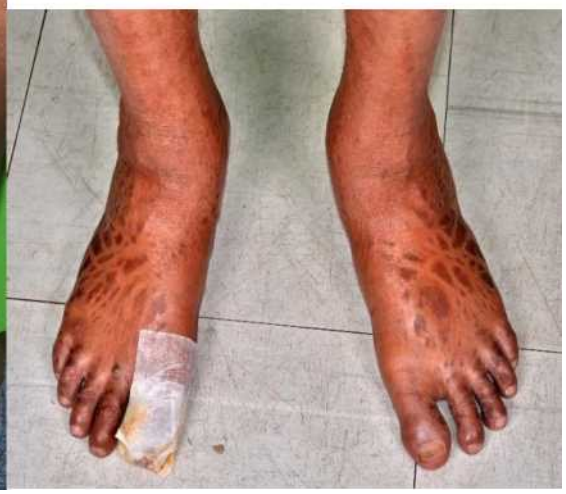
CASE-4 DR TARUN NARANG

- 32 year male , resident of Punjab, Referred from ophthalmology (gradual painless loss of vision in both eyes) for evaluation of Hyperpigmented patches on trunk and limbs x 6 months and Paraesthesias of upper and lower limbs x 6 months. Apparently well 4 years back, Developed febrile illness with multiple, red, painful nodules on face, trunk and limbs and Suspected as leprosy with ENL and started on MDT and Prednisolone. He Improved with treatment but had recurrence of symptoms after prednisolone was stopped - prednisolone was restarted along with methotrexate and Complaints of itchy red lesions with MDT so he stopped MDT himself after 3-4 months and Continued taking prednisolone 5 mg and methotrexate intermittently.MDT was restarted 2-3 times but he stopped it due to adverse effects. Cataract/ osteoporotic fractures- prednisolone stopped. He had worsening of paraesthesias, sensory loss, vision and developed hyperpigmented and scaly lesions,Weakness of hands and feet, Bed ridden and lost his job.
- No history of Hypopigmented lesions, Dyspnea, cough, chest pain, Seizures, Polyuria, polydipsia, Loss of weight, loss of appetite, night sweats, bowel and bladder complaints
- Similar complaints in family members and contacts present.
- On general examination: Avg built, Weight : 65 kg; Height- 156 cms
- Conscious, oriented to time, place and person, Afebrile , BP-100/70 mm Hg, PR-80/min, RR-20/min, No pallor, icterus, cyanosis, clubbing, pedal edema.
- **Lymphadenopathy- Bilateral inguinal lymphadenopathy, discrete, firm, non tender , mobile .**

Systemic examination- within normal limits.



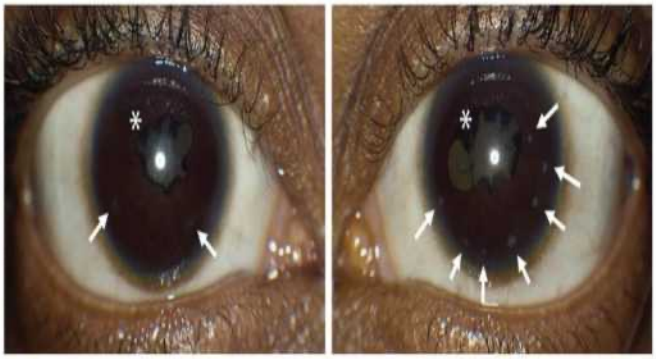




Sensory examination: HMF: Normal

Cranial nerve examination: LMN type facial nerve palsy on left side Sensory examination:

- Generalised hyperaesthesia
- Patchy sensory loss over dorsum of hands, feet, face, and trunk
- Deep sensations- intact
- Chronic anterior uveitis, Posterior synechiae, Cataract, Pre-pthisis bulbi



Shoulder	**Right**	**Left**
Abduction	3/5	3/5
Adduction	3/5	3/5
Flexion	3/5	3/5
Extension	3/5	3/5
Elbow		
Flexion	3/5	4/5
Extension	3/5	4/5
Wrist		
Flexion	3/5	0/5
Extension	0/5	0/5

Hip **Right** **Left**

Abduction	4/5	4/5
Adduction	4/5	4/5
Flexion	3/5	3/5
Extension	3/5	3/5
Knee		
Flexion	4/5	4/5
Extension	4/5	4/5
Ankle		
Dorsiflexion	3/5	3/5
Plantar flexion	3/5	3/5
Inversion	3/5	3/5
Eversion	3/5	3/5

Dr Namrata Chhabra:

Sarcoidosis is one differential as the clinical presentation is not classic for Hansens. The hyperpigmented icthyosiform lesions, ant uveitis, facial palsy, peripheral neuropathy, fever with nodules ? EN. Sarcoidosis should be ruled out ?Heer fordt syndrome.

Dr. Santoshdev Rathod:

eosinophilic granulomatosis with polyangiitis (EGPA). Has peripheral neuropathy, Heart involvement and Ocular manifestations as described in your case.

Dr Shyam Prasad:

In the Indian context, I wouldn't think of anything other than leprosy, unless slit smears are negative. BL/LL leading to peripheral nerve thickening,sensorimotor neuropathy, ypogonadism/gynecomastia, icthyosis, chronic uveitis. Type 2 reactions aggravate the condition leading to facial palsy, lymphadenopathy, and worsening orchitis and uveitis,and possible hepatic impairment. Steroid abuse leading to cataracts, osteoporosis/fractures, possibly myopathy.

Dr Shagufta:

Gradual loss of vision, Bilateral multiple nerves enlarged, paresthesias, patchy sensory loss on multiple sites, wasting of hand and feet muscles, lesions suggestive of ENL in history, response to MD T and steroids, worsening of symptoms once steroids were stopped. Only Hansen's with ENL, with grade 2 deformities.

Dr Tarun Narang

The investigations are as follows

NCS (29/9/17)	severe sensory-motor demyelinating and axonal neuropathy predominantly involving clinically affected nerves.
Nerve biopsy (S-27905)	Fibrocollagenous tissue,focal perivascular infiltration,no granulomas, no nerve fascicles.(noil representative)
PET **CT** (26329/17)	FDG avid bilateral parotid and submandibular glands. FDG avid enhancing lesions in bilateral testes and inguinal and external iliac lymph nodes
DEXA scan	Lumbar spine T-score= **-3.6,** Z-score= **-3.6, WHO** classification = osteoporosis, Fracture risk= high Left hip T-score= -2.1,Z-score = -2,**WHO** classification= osteopenia
HRCT Chest	Normal
USG Scrotum and inguinal legion	Bilateral testes-^ Increased vascularity, increased echotexture, s/o orchitis. Multiple enlarged inguinal L.N. on right side, largest 2.7 x 0.9cm
FNAC of inguinal LNs	Mature lymphocytes, few activated lymphocytes, occasional plasma cells, no granulomas or atypical cells seen
Ultra sound of peripheral nerves	Thickened median, ulnar, cpn, post tibial & small nerves.

Skin Biopsy - Thinned out epidermis

- Dermis showed well-formed granulomas composed of epithelial histiocytes with foamy cytoplasm in most cells, occasional giant cells and sparse LMN cells.
- These granulomas were entraping nerve and adnexal structures.
- No amyloid and necrosis evident.
- Lepra stain -ve

Muscle biopsy-Multifocal collection of epithelioid histiocytes admixed with lymphomononuclear cells and giant cells forming well formed epitheloid granulomas.

- These granulomas at places causing destruction of myofibres.
- Stain for AFB & M. leprae -ve.
- Features s/o sarcoidosis.

MRI brain and brachial plexus

- Diffuse mild cerebral atrophy, with atrophic globe.
- Mild scleral thickening of bilateral eyeballs.
- Diffuse thickening of bilateral brachial plexus with bilateral axillary lymphadenopathy

Brachial plexitis

Based on the investigations the final diagnosis was **Ichythyosiform sarcoidosis with neurosarcoidosis**

Constellation of features are suggestive of sarcoidosis and none of them are individually specific enough to differentiate sarcoidosis from leprosy.

- **Repeatedly negative SSS,**
- **Progressive neurological and ophthalmologic deterioration despite MDT**
- **Raised ACE levels,**
- **Brachial plexitis**
- **FDG avid bilateral parotid and submandibular glands. FDG avid enhancing lesions in bilateral testes and inguinal and external iliac lymph nodes**
- **Sarcoidal granulomas on muscle biopsy**

Treatment Patients was started on Prednisolone 40mg/day and Azathioprine 50 mg od

- After 1 month- improvement in paresthesias and burning sensation/skin lesions
- After 3 months- improvement in muscle power
- After 6 months- steroids were stopped-skin lesions cleared, power improved- patient was employed