DEPARTMENT OF DERMATOLOGY

Clinical Meet 2023

Sr No.	Topic	Presenter
1.	Leprosy, the great masquerader	Dr. Priyanka Patil
2.	Unusual dermatoses with rheumatoid arthritis	Dr. Pooja Chaurasia
3.	Homozygous familial hypercholesterolemia with recurrent mutation	Dr. Tamanna Raman
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Leprosy, the great masquerader

Priyanka Patil

- Leprosy chronic infectious disease with predominant involvement of skin and peripheral nerves.
- A great imitator, mainly because of the wide range of clinical presentations.
- Eliminated prevalent in endemic regions.
- Shift in trend of presentation : Tuberculoid \rightarrow lepromatous
- Important to rule it out even in unusual forms to avoid misdiagnosis.
- Delay in diagnosis and treatment disability and deformities

Unusual presentations of lepromatous leprosy

- Single plaque / nodule
- Zosteriform, segmental, dermatomal leprosy
- Blaschko-like pattern
- Lymphadenopathy masquerading as lymphoma
- Long standing leg ulcer
- Spontaneous skin ulceration
- Histoid leprosy
- Lucio leprosy
- Erythema gyratum repens-like pattern
- Erythema-multiforme like lesion
- Verrucous lesions of lepromatous leprosy







Case 1 41yr/F, known case of type 2 diabetes mellitus on oral hypoglycemic drugs











We received a disc of skin from the right forearm measuring 4 mm in diameter.

The differential diagnosis given were:

- 1) Bullous fixed drug eruption
- 2) Adverse cutaneous drug reaction
- 3) Pemphigus erythematosus
- 4) Sarcoidosis









Following clinical differentials were ruled out:

- 1) Bullous FDE (fixed drug eruption)
- 2) Pemphigus erythematosus
- 3) Adverse cutaneous drug reaction
- 4) Sarcoidosis



- Extensive papular lesions without any systemic involvement.
- Morphologically lesions appeared as histoid and bacteriologically

abundance of globi was confirmatory.

• However, histoid leprosy usually has localized presentation but such a

diffuse involvement was confusing.





We received 2 skin biopsies one from the chest wall and one from the back, both measuring 4 mm in diameter.

The differential diagnosis given were:

- 1) Pityriasis lichenoid chronica(PLC)
- 2) Scleromyxedema
- 3) Epidermolysis bullosa acquisita(EBA)
- 4) Sweet syndrome
- On microscopy both showed similar histomorphological features.







Following clinical differentials were ruled out:

- 1) Pityriasis lichenoid chronica(PLC)
- 2) Scleromyxedema
- 3) Epidermolysis bullosa acquisita(EBA)
- 4) Sweet syndrome



• Such large plaques, at places succulent towards the margin, with high fever favored diagnosis of Sweet syndrome.

• Lower limb swelling and leonine facies were subtle clues pointing towards leprosy.

• Abundance of bacilli in skin biopsy and high BI and MI led to the final diagnosis.

- Leprosy can be diagnosed fairly accurately on basis of three cardinal signs.
- Spectral disease: Tuberculoid to lepromatous pole, as well as histoid, pure neuritic leprosy and reactional states.
- Atypical cutaneous presentations are reported which may lead to diagnostic dilemma.
- Causes of misdiagnosis : sudden onset of lesions, uncommon presentations,

wrong treatments and other drugs (corticosteroids, change in morphology)



<u>Unusual dermatoses</u> with rheumatoid arthritis

Pooja Chaurasia



MOST COMMON

- Rheumatoid nodules
- Rheumatoid vasculitis
- Granulomatous dermatitis
- Neutrophilic dermatoses

LESS COMMON

Psoriasis

EXTREMELY RARE

Bullous pemphigoid

Fig. 37.2 Cutaneous findings in rheumatoid arthritis. See next page for figure legend.

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Case 1:
65/F
RA since 4 years;
Pyoderma
gangrenosum since 2
years
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- We received a skin biopsy from the right buttock measuring 4 mm in diameter.
- The differential diagnosis given were:
- 1) Hypertrophic lichen planus
- 2) Pityriasis Lichenoides et Varioliformis Acuta (PLEVA)
- 3) Vasculitis
- 4) Sweet's syndrome
- 5) Pyoderma Gangrenosum







Following clinical differentials were ruled out:

- 1) Hypertrophic lichen planus
- 2) Pityriasis Lichenoides et Varioliformis Acuta (PLEVA)
- 3) Vasculitis
- 4) Sweet's syndrome

The final diagnosis offered was- Pyoderma Gangrenosum

Case 2: <u>48 Y/F</u> RA since 22 years Palmoplantar psoriasis since 2 months;














• We received a skin biopsy measuring 4 mm in diameter.









40x

PG

- 65 Y/F
- RA since 4 years
- Multiple well defined painful ulcers over lower extremities
- RA: 431
- Anti-CCP: 78.5
- Treatment:
 - Prednisolone
 - Methotrextate
 - Clobetasol ointment

Psoriasis

- 48 Y/F
- RA since 22 years
- Itchy fissured indurated scaly palmoplantar plaques
- RA: 667
- Anti- CCP: 64.2
- Treatment:
 - Clobetasol ointment + Salicylic acid
 - Methotrextate
 - Naproxen



- 54 Y/F
- RA since 25 years
- Multiple tense bullae preceded by intense itching
- RA:386
- Anti-CCP: 57.4
- Treatment:
 - Prednisolone
 - Dapsone
 - Doxepin

PG and RA: What is the link?

Ulcerative PG mimics rheumatoid vasculitis due to common predisposing factors:

- skin fragility
- arterial disease
- peripheral edema
- nutritional status
- venous insufficiency



<u>Are T- cells the common link?</u>



Autoinflammatory diseases are driven by abnormalities of the <u>innate</u> immune system

In PG lesions, <u>pattern recognition receptors</u> are upregulated

TLR overexpression has been implicated in RA and IBD.

INFLAMMATORY CASCADE IN RA



<u>Psoriasis, Psoriatic arthritis, and Rheumatoid</u> <u>arthritis:</u> <u>Is all inflammation the same?</u>

• Gut dysbiosis linked with their pathogenesis.

- HLA alleles affect disease susceptibility and severity
 - -PsA → <u>HLA-B27</u>: Enthesitis and symmetric sacroiliitis.
 <u>HLA B08</u>: Joint fusion, asymmetric sacroiliitis, dactylitis.

-RA \rightarrow <u>HLA-DRB1</u>: Severity in patients with RF and anti-CCP positive.



Bullous Pemphigoid and Rheumatoid Arthritis: Is there Disease Association?

- RA cellular immune response to an **unknown synovial membrane antigen**.
- BP-- <u>BPAg1</u>, hemidesmosomal plakin and <u>BPAg2</u>, collagen XVII :- Link the hemidesmosomes to the lamina densa.
- The **common pathogenesis** of BP and RA:
 - cross reaction between above antigens and those in the synovium, or
 - unmasking of antigens in the skin by inflammation initiated in the synovium in cases of RA.
- **Association** rather than **chance occurrence** between these two diseases.



Homozygous familial hypercholesterolemia with recurrent mutation

Tamanna Raman

 Autosomal codominant disorder characterized by lifelong elevation of LDL cholesterol (LDL-C)

- Caused by pathogenic mutations in
 - Low density lipoprotein receptor (LDLR)
 - Apolipoprotein B (APOB)
 - Proprotein convertase subtilisin/kexin type 9 (PCSK9) genes.



35 yr old / M



Dermoscopy



Fasting lipid profile

- The fasting lipid profile was found to be deranged
 - 1. Total cholesterol (TC): 500 mg/dl (TC: < 200mg/dl)
 - 2. Triglyceride: 302 mg/dL (TG: < 150 mg/d)
 - 3. LDL: > 300 mg/dL (LDL: < 100 mg/dl)
 - 4. VLDL: 60.4 mg/dL (VLDL: 5–40 mg/dl)
 - 5. HDL: 21 mg/dL (HDL: 40-60 mg/dl)

Histopathology



Next-generation sequencing

- Performed on the proband and family members, specifically targeting the exons of
 - 1. LDLR,
 - 2. APOB,
 - 3. PCSK9.

Autosomal recessive familial hypercholesterolemia

Proband – GDN 11138 (35 year male) Mother – GDN 11139 Brother – GDN 11140

Identified mutation LDLR:pS177L (missense variant in exon of LDLR gene)

Proband – Homozygous state Mother – Heterozygous state Brother – Negative for mutation



Discussion

- According to the UCL LDLR gene variant database, 3779 LDLR mutations have been reported so far
 - 77% were substitutions
 - 16%, deletions
 - 5%, duplicates
- Typical clinical picture can result from the accumulation of common cholesterol-increasing alleles (polygenic FH) approximately 80% of FH patients remain undiagnosed.

3 patients from India

1 Heterozygous

2 Homozygous

Analysis of pharmacogenomic variants

• Additionally, the analysis included screening of pharmacogenomic variants rs2306283, rs4149056, and rs2231142, which are relevant to Statin-Associated Musculoskeletal Symptoms (SAMS)

• Features of SAMS

- 1. Myalgia
- 2. Tenderness
- 3. Stiffness
- 4. Cramps

Analyzing the pharmacogenomic variants

- Proband and mother were heterozygous for the rs2306283 (c.388A>G) and rs4149056 (c.521T>C) variants in SLCO1B1 gene, leading to decreased function phenotype.
- No change in treatment prescribed to the patient by cardiologist (atorvastatin 40 mg and ezetimibe 10 mg) as complied with recent CPIC guidelines.



- This patient received a combination therapy of atorvastatin 40 mg and ezetimibe 10 mg as per CPIC guidelines with no adverse effects.
- Screening prior to treatment enables stratification of individuals at higher risk of SAMS.
- Facilitates the implementation of personalized medicine.



The use of Secukinumab in autosomal recessive congenital ichthyoses: a case

series

Shrishti Singh

INTRODUCTION

Autosomal recessive congenital ichthyosis (ARCI) has four forms

CONGENITAL ICHTHYOSIFORM ERYTHRODERMA (CIE)



erythrodermic variant

LAMELLAR ICHTHYOSIS (LI)



fish-like scales all over

HARLEQUIN ICHTHYOSIS



diamond shaped, hard thick plates

PLEOMORPHIC ICHTHYOSIS



Heterogeneous group, mild residual scaling

• The treatment of ARCI still remains unsatisfactory.

Rationale for Injecting Secukinumab



- Recent studies IL-17/23, upregulated in skin as well as blood of individuals with ARCI.
- Off label use of **Ustekinumab**, beneficial in CIE and Netherton syndrome (case reports) [2]
- Encouraged us to administer
 Secukinumab, a monoclonal antibody against IL-17, in four children with ARCI

75mg of Inj. secukinumab, s/c
Weekly x 5 weeks
F/b once monthly x 6 months.

- Treatment response was assessed using
 - The Ichthyosis Area and Severity Index (IASI),
 - The Dermatology Life Quality Index (DLQI),
 - The Itch Severity Scale (ISS)

• Follow up period – 3 months
RESULTS

CASES ONE (16/F) AND TWO (14/M)

 Siblings with lamellar ichthyosis (homozygous p.Gly218Ser variation in TGM1), presented with diffuse scales present all over, and were under intermittent treatment with topical steroids, moisturizers and oral acitretin (25 mg) x 8 years

• Unfavourable consequences of retinoids in the elder sibling (early epiphyseal closure)







Before









DLQI ISS

CASES THREE (17/F) AND FOUR (16/M)

- Siblings diagnosed clinically as CIE.
- They presented with diffuse erythematous scales all over the body.
- More severe in elder sibling.
- Her brother had a milder phenotype.



After

Before











No adverse events were reported.

All four patients exhibited sustained benefit during the follow up period of three months post therapy.

DISCUSSION

- Gene therapy for ARCI is evolving; is not yet available
- An open-label clinical trial investigating Secukinumab in ARCI; LI patients had a modest improvement in scaling, due to primarily barrier dysfunction with little inflammation
- By contrast, siblings with CIE demonstrated dramatic improvement, due to greater inflammatory component and elevated IL-17/IL-23 levels
- Larger studies evaluating long-term benefit of this treatment will help target the inflammation underlying most types of ichthyoses while using other supportive measures to reduce scaling.

CLINICAL AND EXPERIMENTAL

DERMATOLOGY THE EDUCATIONAL JOURNAL OF THE BRITISH ASSOCIATION OF DERMATOLOGISTS

REVIEW ARTICLE

The role of thalidomide in dermatology (p 667) **ORIGINAL ARTICLE**

Readability assessment of the British Association of Dermatologists' patient information leaflets (p 684)

Continuing Professional Development

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Subramani D, Sardana G, Kothari R, Gupta A. Secukinumab significantly reduces inflammation but only mildly improves scaling in four cases of autosomal recessive congenital ichthyosis. Clin Exp Dermatol. 2022 Dec;47(12):2288-2290.



WILEY



Long pulsed Nd:Yag & Er:Yag; A versatile laser with complementary wavelengths

Namratha Puttur



Fotona SP Dynamis- Our experience of 6 months

• A highly versatile, multi-purpose laser system with a wide range of

applications in **aesthetics**, **surgery** and **gynecology**.

• The department of dermatology obtained this system six months ago

and it has shown remarkable results.

Two laser wavelengths- Nd: Yag at 1064nm and Er:Yag at 2940 nm



Long pulsed non-ablative Nd:Yag laser

- Nd:YAG penetrates deep into the skin
- Selectively targets structures such as veins and hair follicles.
- Long pulse makes it safe for darker skin tones

Indications of Nd:Yag

Laser hair reduction



After 2 sessions of LHR, 4 weeks apart

 World's best and fastest technology for hair reduction

No gel needed

 No burns or post inflammatory hyperpigmentation

Vascular lesions



Haemangioma

Vascular lesions



Thread veins

Inflammatory rosacea



After 1 session

Inflammatory acne



After 1 session

Onychomycosis

 One of the only machines which can treat onychomycosis without

the need of **antifungals**



Warts



Induces coagulation of blood
vessels feeding the wart causing
subsequent necrosis

Before



Ablative Er:YAG laser

- Er:YAG is selectively absorbed by **water** in skin cells, enabling extremely precise ablation.
- Also stimulates the production of **collagen** in the dermis.
- Minimal adverse effects
- Reduced recovery time

Indications of Er:Yag

"HaiRestart"- hair restoration

- Production of heat shock proteins, growth factors & extracellular matrix proteins
- Painless, no downtime



After 6 sittings 2 weeks apart

After 6 sittings 2 weeks apart

Scar resurfacing



After 1 session

"TensorTight"-skin tightening

Minimally invasive procedure that is especially effective in saggy skin



After 2 session ,4 weeks apart

"TightSculpting"- fat loss



Non-invasive laser
treatment for sculpting and
skin tightening on all body
areas

"LineLase"-stretch mark reduction



• Effective and safe treatment

for **stretch marks** that is

appropriate for all skin types

After 1 session

"OrangeLase" - cellulite reduction



• Non-invasive treatment of

cellulite that effectively

tightens skin and

increases its elasticity

"SmoothEye" - periocular rejuvenation



Immediate tightness of the

periocular area

•Stimulates collagen remodeling

and initiates neocollagenesis

After 1 session

"LipLase" - perioral rejuvenation



- Non-invasive lip plumping
- Increase in youthful appearance
"VectorLift"- eyebrow lift



- Non-invasive laser eyebrow lifting
- Natural-looking results with no

downtime

Combined Indications of Er:Yag and Nd:Yag

4D Face Lift



4D Face Lift



Non-dermatological indications NightLase

"NightLase" - snoring and apnea treatment



- Increases the size of airway by upto
 23%
- Results seen immediately after a single sitting
- Decrease in snoring and increase in quality of sleep

Witherspoon P. Understanding the Link Between Sleep Apnea and TMJ. Dental Products Report. 2021 Oct 1;55(10). Jovanovic J. NightLaseTM–laser-assisted snoring and apnea reduction, 9 months of experience. J Laser Heal Acad. 2011;2011:S11. Sippus J. CASE REPORT: NightLase® Procedure–Laser snoring and sleep apnea reduction treatment. J Laser Health Acad. 2015;2015:1-5.

ComfortLase

"ComfortLase" - photobiomodulation and pain management



- Stimulates cells to naturally heal, relieve pain and reduce inflammation.
- Accelerated tissue regeneration
- Increased lymphatic flow
- Stimulation of cell metabolism

Regulski PA, Szopinski KT, Levičnik-Höfferle Š. Photobiomodulation Therapy for the Symptoms Related to Temporomandibular Joint Disk Displacement. Case Reports in Dentistry. 2023 Apr 13;2023.

Amaroli A, Benedicenti A, Ravera S, Parker S, Selting W, Panfoli I, Benedicenti S. Short-pulse neodymium: Yttrium–aluminium garnet (Nd: YAG 1064 nm) laser irradiation photobiomodulates mitochondria activity and cellular multiplication of Paramecium primaurelia (Protozoa). European Journal of Protistology. 2017 Oct 1;61:294-304.

Photobiomodulation and pain management



Gynaecological indications

- Minimally invasive solution for stress urinary incontinence
- Improves urethral support by photothermal stimulation of vaginal wall

- Non ablative treatment for vaginal atrophy post menopause
- Induces microcirculation and tissue regeneration



Mild and moderate stress and mixed urinary incontinence



After IncontiLase® treatment



Atrophied vaginal mucosa



Vaginal mucosa after RenovaLase treatment

