Supplementary table 1: Differential diagnosis of AE					
Dermatological disease	**AE[1-3]**	**Pellagroid dermatitis[4]**	**Biotinidase deficiency[5]**	**Leiner's disease[6]**	**Complement deficiency disorder[7]**
Cause	Zinc deficiency (genetic or acquired)	Niacin (vitamin B3 deficiency)	Autosomal recessive disorder is caused by mutations in the *BTD* gene on chromosome 3p25.1, leading to impaired biotin recycling and enzyme function.	The exact cause of Leiner's disease remains unknown, but biotin deficiency linked to Complement component 5 coded by the C5 gene plays a role.	Inherited deficiencies in specific complement components (e.g., C1q, C2, C4, C5–C9) or regulatory proteins (e.g., C1 inhibitor)
Clinical features	Symmetrical erythematous, squamous, or eczematous lesions around the perioral, anogenital, and acral regions	Symmetrical erythematous scaly plaque in bilateral sun-exposed areas of the body.	Eczematous, seborrheic dermatitis -like rash on the face and scalp	Severe and progressive infantile erythroderma resembling generalised seborrheic dermatitis.	C1q, C2, C4 deficiencies: Associated with lupus-like lesions, photosensitivity, and vasculitic rashes. - C5–C9 (terminal pathway) deficiencies: Increased susceptibility to Neisseria infections, which can present with petechial or purpuric rashes
Distribution	Periorificial and acral region	Malar rash, dorsum of the hands and feet	Periorifical region	Rapid-onset scaling with greasy crusting on the scalp, grey opaque scales on the trunk and limbs, and exudative, oedematous intertriginous areas.	-**Lupus-like lesions:** Typically on sun-exposed areas. - **Vasculitic rashes**: Can occur on the lower extremities or other areas.
Other features	Alopecia, diarrhoea, lethargy, neuropsychiatric problems, growth retardation	Dementia, diarrhoea, anxiety, glossitis, cheilosis, gastritis	Alopecia, conjunctivitis, seizures, hypotonia, ataxia, visual impairment, sensorineural hearing loss, developmental delay	Recalcitrant diarrhoea, malabsorption and wasting, and recurrent staphylococcal and candidal infections	Glomerulonephritis, meningitis, systemic lupus erythematosus
Diagnosis	Low serum zinc levels, low alkaline phosphatase level, and genetic testing for *SLC39A4* mutation	Low serum tryptophan, NAD, NADP, and niacin levels, with low urinary N1-methylnicotinamide (<5.8 µmol/day) and reduced erythrocyte NAD concentrations	Genetic testing for BTD mutations and lab tests, including high lactic acid, ammonia, arterial blood gas, and serum amino acids	Deficient opsonic activity in the patient's serum, with or without a deficiency in complements C3, C4, or C5	Tests like CH50 (total hemolytic complement) and AH50 to assess classical and alternative pathways.
Histopathology	Early lesions show granular layer loss, confluent parakeratosis, PMN infiltrates, and mild spongiosis. As lesions progress, ballooning, reticular degeneration, and keratinocyte necrosis develop, leading to psoriasiform hyperplasia in end-stage disease.	Stratum corneum degeneration, epidermal thickening, fibroblast proliferation, capillary dilation, and an inflammatory infiltrate of lymphoid and plasma cells.	Acanthosis, hyperkeratosis, perivascular lymphocytic infiltrate	Epidermal hyperplasia, spongiosis, and a mixed inflammatory infiltrate in the dermis.	-**Lupus-like lesions**: Interface dermatitis with basal cell vacuolisation. - **Vasculitis**: Leukocytoclastic vasculitis with neutrophilic infiltration around blood vessels.
Treatment	A lifelong supplementation with 3 mg/kg/day of elemental zinc may be required	Nicotinamide doses of 250 to 500 mg/day orally should be given	biotin replacement with a starting dose of 5 to 20 mg daily.	-**Supportive care**: Ensuring proper nutrition and hydration. - **Infection management**: Use of antibiotics for bacterial infections. - **Immune support**: Therapies like fresh-frozen plasma have been utilised.	- **Preventive measures**: Regular vaccinations (e.g., against Neisseria meningitidis). - **Infection prophylaxis**: Long-term antibiotics to prevent recurrent infections.
NAD: Nicotinamide adenine dinucleotide, NADP: Nicotinamide adenine dinucleotide phosphate, PMN: Polymorphonuclear neutrophils, BTD: Biotinidase					