ACRODERAMATITIS ENTEROPATHICA: A CASE REPORT

Vageeshwari Devuni

Pharm. D
CMR college of Pharmacy,
Kandlakoya, medchal road, Hyderabad(501401).

***Abstract*: Acrodermatitis enteropathica is a rare genetic autosomal recessive disorder, Characterized by periorificial dermatitis, alopecia, and diarrhea. It is caused by Mutations in the general that encodes a membrane protein that binds zinc. It is Characterized by inflammation of the skin(dermatitis) around bodily openings (periorificial) and the tips of fingers and toes(acral), hair loss(alopecia) and diarrhea.It occurs due Acrodermatitis to zinc deficiency.The deficiency caused by a defect of dietary zinc absorption in the duodenm and jejunum. Consequently, there are multiple signs and symptoms in severe deficiency: growth retardation, impaired immune function, and multiple skin or gastrointestinal lesions.**

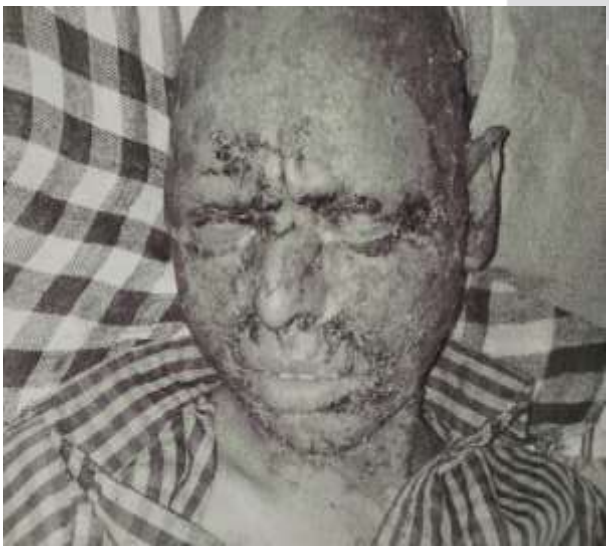
***Keywords:* ENTEROPATHICA, dermatitis, alopecia zinc, bacterial infection, Erythematous lesion.**

INTRODUCTION:

Acrodermatitis ENTEROPATHICA (AE) is an autosomal recessive condition resulting in Severe zinc deficiency. The deficiency is caused by a defect of dietary zinc absorption in The duodenum and jejunum(1). Zinc is an essential co-enzyme in metal enzymes(like alkaline phosphates); it is an important Structural component of gene regulatory proteins(required, for example, for the intracellular Binding of tyrosine kinase to T-cell receptors) and it has also a function in regulation(has the Ability to regulate gene expression). Consequently, there are multiple signs and symptoms In severe deficiency: growth retardation, impaired immune function, and multiple skin or Gastrointestinal lesions(2).'It is classified as primary zinc deficiency, genetically based deficiency, and acquired secondary deficiency(3). The genetic zinc deficiency is associated with the defects in two zinc transporters, one is involved on intestinal zinc uptake Zrt-and Irt-like protein-4(ZIP), causing classical AE and the other is responsible for zinc secretion in breast milk and transporter-2(ZnT) resulting in deficiency of lactogenic origin(4). Zinc is an important constituent of the catalytic site of many vital metalloenzymes in the body like carbonic anhydrase and alkaline phosphatase. Zinc finger proteins, like those seen in retinoic acid and vitamin D receptors play an important role in structural differentiation of many organs including the skin. Zinc also plays a role as an ionic signal regulating gene expression. Zinc transporting proteins are expressed by two gene families, ZnT and Zip that play opposite roles in Zinc trafficking. Zinc is absorbed primarily in the jejunum by the transporting protein Zip4. Zinc levels progressively decrease in human breast milk; however bioavailability is higher in breast milk than cows milk(5). It is found in good amounts in protein rich food especially animal protein. It's bioavailability is decreased by phytates and iron.

CASE:

Here is a case of 40 years adult male with painful lesions since 3 months developed crusted lesions associated with redness initially over both feet which is gradual in onset progressed to involve both knees, genitalis, axilla, back, face, scalp, neck. Pain associated with lesions photosensitivity+, excessive loss of hair, watery discharge from eyes, burning sensation+, photophobia+. On examination patient was found with absence of eyelids and eyebrows. Oral cavity-thrush+. On Cutaneous examination multiple erosions+ over face, scalp, neck, palms, forums of hand extensions aspect of both elbows, axilla, groin, both knees dorsal aspect of both feet extending onto legs associated with scaling. Multiple scaly papular lesions+, over chest and back were present. Laboratory



findings shows decreased serum zinc levels 45mg/dl, ESR-1-15min, ESR-2-29min, RBS-132mg/dl, BU-44mg/dl, Serum creatinine-0.7mg%. Past medication history shows decreased zinc since childhood which was treated with Tab.Ascazin 50mg TID, Tab.Riboflavin 2tabs TID. Based on Subjective and Objective data taking past medical history into consideration it is confirmed as Acrodermatitis enteropathica.

DISCUSSION:

Patient has low serum Zinc levels from childhood, due to Zinc deficiency lead to Acrodermatitis. Then the patient is prescribed with Zinc supplements to be taken orally (40mg) 3 times a day since six months. Instead of taking medication thrice a day patient took it once a day. Therefore it lead to reoccurrence of Acrodermatitis enteropathica. Patient and his family mem are counselled about drug use and it's importance in preventing the recurrence of ADR and advices the patient to use medication regularly. ADE was first described in 1963 by Brandt and later identified as a definitive disease in 1942 by Donbolt and Closs(6). Classical ADE is believed to be a result of a defect in a zinc transporting protein encoded on chromosome 8q24.3.5 by the gene SLC39A4. It is a histidine-rich transmembrane protein known as hZIP4, involved in Zinc uptake(7). The systemic features of zinc deficiency are diarrhea, anorexia, growth retardation, photophobia, conceal opacities, hypothesis, hyposmia, hypogonadism, amenorrhea, anemia, impaired wound healing, hoarseness, neuropsychiatric problems, and perinatal morbidity due to increased Zinc requirement in pregnancy(8). Immunologic abnormalities like decreased T-cell, neutrophils, natural killer(NK) cell and macrophage function result in secondary colonization of erosions and infections. Our patients had anemia, diarrhea and poor wound healing. In the Histopathology, necrolysis, a term describing cytoplasmic pallor, vacuolation, ballooning degeneration, and subsequent confluent necrosis of keratinocyte within the superficial stratum spinosum and stratum granulosum of the epidermis is said to be almost pathognomonic. Other features are confluent parakeratosis, hypergranulosis and psoriasis OEM hyperplasia(9). Accepted treatment now a days is with supplementation of elemental Zinc at a dose of 2mg/kg/day, atleast two or three times the recommended dietary allowance of 15mg/day. The most accurate way of establishing diagnosis is by measuring plasma or serum Zinc levels, though ADE with normal Zinc Levels has been reported(10).

Zinc is a very important micronutrient, a component of more than two-handed metalloenzymes, and essential for the proper functioning of the various metabolic and biochemical pathways of the body. AE results from mutations in the Zinc transporter gene SLC39A4(solute carrier family 39 member A4), leading to improper enteral Zinc absorption(11).

CONCLUSION:

Acrodermatitis enteropathica is a rare condition; early recognition of cutaneous manifestation being necessary, particularly because of immunodeficiency and infection concerns in these patients. This case is being reported to highlight that isolated Zinc deficiency can occur in adults and should be considered to prevent mismanagement of this otherwise easily treatable condition.

REFERENCES:

[1] Nicolai History, MD, PhD, Lavinia Ciontu, MD, and Violets Strange, MD, PhD Medicine(Baltimore). Acrodermatitis Enteropathica 2016May;95(20):e3553.
[2] Maverakis E, Fung MA, Lynch PJ,et al. Acrodermatitis enteropathica and an overview of Zinc Metabolism. J Am Academic Dermatol 2007;56:116-124.
[3] Sarabjit kaur, Ankita Sangwan, Priyadarshini Sahu, et al.Clinical Variants of Acrodermatitis enteropathica and it's Co-relation with genetics. 4 Jan 2016; Bol 17(1); 35-37.
[4] Kasana S, Din J, Maret W. Genetic causes and gene-nutrient interactions in mammalian Zinc deficiencies: Acrodermatitis enteropathica and transient neonatal Zinc deficiency as examples. J Trace Elem Med Biol2015;29:47-62.
[5] Maverakis E, Fung MA, Lynch PJ, et al. Acrodermatitis enteropathica and an overview of Zinc Metabolism. J Am Academic Dermatol 2007;56:116-24.
[6] Donbolt N, Closs K. Acrodermatitis enteropathica. Acta Derm Venereol1942;23:127-69.
[7] Kharfi M, El Fekih N, Aounallahskhiri H, et al. Acrodermatitis enteropathica: A review of 29 Tunisian cases. Int J Dermatol 2010 ;49:1038-44.
[8] Jagadeesan S, Kaliyadan F.Acrodermatitis enteropathica. Dec 5;2018.
[9] Tatlican S, Yamangokturk B, Eren C, et al. Diagnostic challenge: A case of Acrodermatitis enteropathica without hypozincemia and with maternal milk of low Zinc level. Pediatr Dermatol 2010;27:534-5.
[10] Mohan Saritha, Divya Gupta, Laxmisha Chandrashekar, et al. Acquired Zinc deficiency in an adult female. 2012; 57(6); 492-494.
[11] Jagadeesan S, Kaliyadan F. Acrodermatitis enteropathica. Dec 5;2018.