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Oculo – Orogenital Syndrome: A Rare Case

Dr. Bushra Khan[1], Dr. Sushil Pande[2], Dr. Milind Borkar[3], Dr. Ramesh Sharma[4]
[1]Post Graduate Resident, Department of Dermatology, NKP Salve Institute of Medical Science and Research Centre, Digdoh Hills, Nagpur, Maharashtra, India
[2]Associate Professor, Department of Dermatology, NKP Salve Institute of Medical Science and Research Centre, Digdoh Hills, Nagpur, Maharashtra, India
[3]Professor and Head, Department of Dermatology, NKP Salve Institute of Medical Science and Research Centre, Digdoh Hills, Nagpur, Maharashtra, India
[4]Professor and Guide, Department of Dermatology, NKP Salve Institute of Medical Science and Research Centre, Digdoh Hills, Nagpur, Maharashtra, India

***Corresponding author**
Dr. Bushra Khan
Email: dr.bushrakhan@hotmail.com

Abstract: A 50-year-old man, old case of Type 2 diabetes mellitus with peripheral neuropathy with chronic pancreatitis, presented with a 20 days history of inflamed tongue and angles of mouth, oral ulcers, irritated, edematous and itchy genital skin, and dry burning eyes. He had a history of fever with chills, loss of appetite, painful defecation and weight loss. On physical examination, he had mainly mild Erythema over inner canthus and medial sclera, a bright red and smooth tongue, and edematous dermatitis of the genitals. Other findings included pallor (++), pitting type pedal edema, multiple painful erosions of variable sizes with well defined margins present bilaterally over buccal mucosa. The patient had been consuming large quantities of alcohol daily since the past 25 years but hardly any cereals or vegetables. On routine laboratory testing, the hemoglobin level was 10.2 g perdeciliter, the white-cell count was 16000 per cubic millimeter with an abnormal differential count (86.0 percent neutrophils, 1 percent eosinophils, 1 percent basophils, 0 percent monocytes, and 12 percent lymphocytes). Mean corpuscular volume was 92 Units and Mean corpuscular hemoglobin concentration was 32 Units. Sr Alkaline phosphatase was mildly raised (197 units/liter) and Sr Proteins were low (Total proteins 4 g/deciliter, Albumin 1.9 g/deciliter, Globulin 2.1 g/deciliter) as were Sr Sodium levels (124 mEq/liter). On the basis of clinical findings and investigations, diagnosis of Oculo-Orogenital Syndrome was made. The patient was treated with 2 cc of Vitamin B complex injections per day intravenously and an oral multivitamin once per day. All lesions responded within 7 days. Nutritional instructions were given, but the patient refused treatment for his alcoholism.
Keywords: Oculo–Orogenital Syndrome, oral ulcers, fever.

INTRODUCTION

Oculo-orogenital syndrome is a deficiency disorder caused due to the deficiency of Riboflavin (Vitamin B2). Long before the discovery of Ribo-flavine, Stannus described a group of symptoms in-cluding soreness of tongue and lips, with a sodden excoriated condition at the angles of the mouth, and pal-pebral fissures and a characteristic lesion at the free border of the prepuce, vulva and anus [1]. Scott described a condition among the Jamaican coolies that shared some of the symptoms described by Stannus and in addition central neuritis, photophobia, indistinctness of vision, ulceration, discharge and burning sensation in the eyes. After the discovery of Riboflavin by Warburg and isolation of its crystals by Khun [8] Sebrell and Butler [4] in experimentally produced ariboflavinosis in men noted a "Scaly slightly greasy, desquamative lesion on a mildly crythematous base in the nasolabial folds, on the alae nasi, in the vestibule of the nose and occasionally at the ears and around the eyelids especially at the inner and outer canthi [1].

This disorder was noted prominently amongst prisoners of war in Japanese war camps in 1942 by Col. Eugene C. Jacobs (U. S. Army). In 1945-46, reports from Singapore captivity prisoners by Smith and Woodruff [7] mention about the occurrence of keratitis, retro-bulbar neuritis and scrotal dermatitis occurring due to riboflavine deficiency.

In India Aykroyd and Verma [5] described superficial keratitis occurring in ariboflavinosis. Later Verma [6] described angular conjunctivitis of Morax-Axenfeld type occurring in patients suffering from stomatitis and other signs of ariboflavinosis [1].

Visible light phototherapy, Chlorpromazine, Borates etc. causes decreased levels of riboflavin in the body [2]. Reports from the prisoner of war camps in the

Far East showed that blindness can occur as a result of vitamin B-complex deficiency [3]. With few exceptions, the resulting lesions cannot be ascribed definitively to the deficiency of a particular vitamin,as in most cases all the B-complex factors are deficient. This polyhypovitaminosis, caused due to decreased intake or inadequate absorption, combines various aforementioned signs and symptoms. This constellation of signs and symptoms has been termed as Oculo-Orogenital Syndrome.

Acute Clinical findings include a deep red erythema, epidermal necrolysis and mucositis. The severity of these depends on the severity of the deficiency [2].

Chronic clinical findings begin 3 – 5 months after the initiation of causative factor. Predominantly skin and mucous membrane involvement is seen. Angular stomatitis, macerated bleeding fissures that extend laterally and may ulcerate are present initially. Later cheilosis may be pronounced with xerosis, erythema and vertical fissuring of lips [2].

Tongue becomes smooth, swollen, magenta colored with loss of lingual papillae [2]. (glossitis). Dermatitis resembles seborrheic dermatitis with involvement of nasolabial, posterior auricular, cheeks and forehead as well as flexural areas of limbs, genitals, perianal and buttocks. More commonly seen in males as red, confluent, crusty dermatitis of the scrotum, which often spreads to involve the groin. In infants it is more pronounced in inguinal areas and in adults it is seen in nasolabial folds/wrinkly areas [2].

Ocular findings include photophobia, conjunctivitis, keratitis, corneal vascularization. Oculo-Orogenital Syndrome is endemic in India, China and Iran [2] Men and women are equally affected. Low socioeconomic status (e.g., poverty), and advanced age increase the risk of developing vitamin B_2 deficiency. Individuals who do not eat meat or dairy products (vegans) are at increased risk for vitamin B_2 deficiency. Individuals who follow fad diets (e.g., eating only a few select foods) or those who suffer from alcoholism or eating disorders (anorexia nervosa, bulimia nervosa) have an increased risk of developing this disease. The exact incidence is not known. The case is being reported for the rarity of its occurrence in Vidarbha region.

CASE REPORT

A 50-year-old man, old case of Type 2 diabetes mellitus with peripheral neuropathy with chronic pancreatitis, presented with a 20 days history of inflamed tongue and angles of mouth; oral ulcers; irritated, edematous and itchy genital skin, and dry burning eyes. He had a history of fever with chills, loss of appetite, painful defecation, and weight loss. There was no history of loose stools, burning micturition or loss of memory. He was a chronic alcoholic since 25 years having approx. 100 – 150 g of alcohol daily with a poor appetite. He had a history of taking unprescribed Tab. Ofloxacin twice daily for 3 days.

On physical examination, the patient had pallor, was afebrile to touch and vitally stable. Pitting type of pedal edema was present bilaterally and mild tremors were present. Systemic examination was normal with no abnormal heart of lung sounds and no organomegaly.

On cutaneous examination, few erythematous patches were seen over the lower abdomen and diffuse erythema with oedema was present over scrotum and penis. Oral hygiene was poor and multiple (5-6) painful erosions of variable sizes with well defined margins were present over buccal mucosa bilaterally. Single, non-indurated, 2 x 1 cm size tender ulcer with well defined margins and healthy granulation tissue present over left buccal mucosa. Tongue was erythematous, smooth and oedematous with central fissuring.

On routine laboratory testing, the hemoglobin level was 10.2 g perdeciliter, the white-cell count was 16000 per cubic millimeter with an abnormal differential count (86.0 percent neutrophils, 1 percent eosinophils, 1 percent basophils, 0 percent monocytes, and 12 percent lymphocytes). Mean corpuscular volume (MCV) was 92 Units and Mean corpuscular hemoglobin concentration (MCHC) was 32 Units. RBCs were 3.40 million/mm^3, Platelet Count was 2.73 lakh/mm^3. Sr. Bilirubin was 0.5, S.G.O.T was 46, S.G.P.T was 54. Sr. Urea was 10 , Sr. Creat was 0.6 , Sr. Potassium was 2.8. Sr Alkaline phosphatase was mildly raised (197 units/liter) and Sr Proteins were low (Total proteins 4 g/deciliter, Albumin 1.9 g/deciliter, Globulin 2.1 g/deciliter) as were Sr Sodium levels (124 mEq/liter). Urine examination showed presence of sugar, 2 – 3 pus cells, 5 -6 epithelial cells. Glycosylated haemoglobin was raised (10.5%) Ultrasound Sonography was suggestive of scrotal and penile oedema ?cause.

The patient was placed on 2 cc of Vitamin B complex injections per day intravenously and an oral multivitamin once per day. For symptomatic relief, tablet loratidine 10mg once daily, tablet pheniramine 25mg once at night, choline salicylate and tannic acid containing topical oral mucosal gel thrice daily, tablet Lidocaine 2 tablets thrice daily and tablet fluconazole 150mg once a day for 7days.

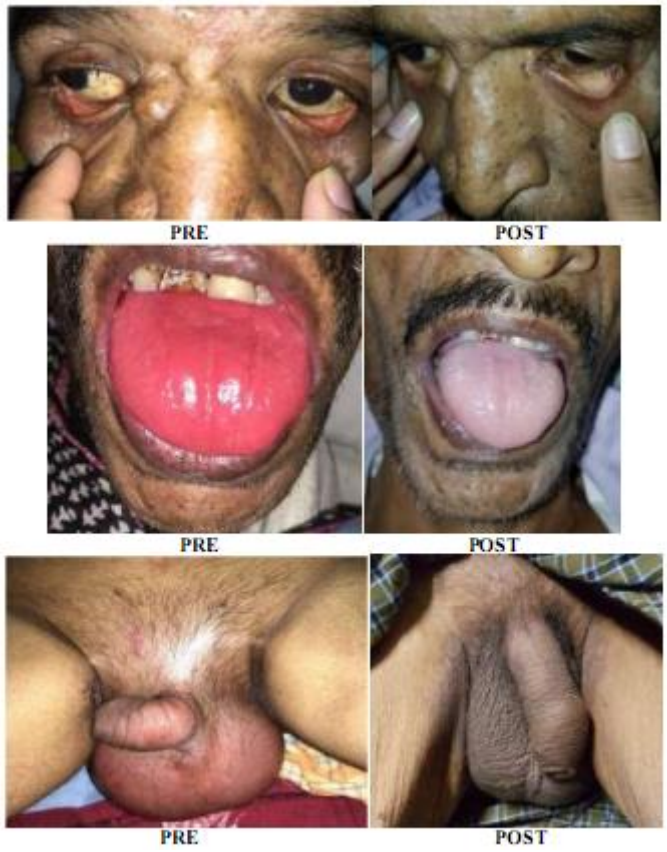

Fig-1: Clinical Examination

DISCUSSION

Oculo- Orogenital Syndrome affects individuals with decreased dietary intake and malabsorption. Many of these patients are chronic alcoholics as was the case with this patient as well. The classical features of mucositis, glossitis and scrotal dermatitis were seen in this patient. Males and females are equally affected with no sexual preponderance, although genital involvement is more common in females as seen in this case.

Features include: Papulosquamous lesions; ulcers; Seborrheic dermatitis like changes in peri-orificial sites; angular stomatitis; magenta red, smooth, edematous tongue; lichenified dermatitis of the scrotum and conjunctivitis with photophobia; anaemia, mental retardation.

Generally, the diagnosis of Oculo-Orogenital Syndrome is based on history and clinical findings but erythrocyte glutathione reductase activity as a screening test and urinary levels of riboflavin (which are usually below 30 mcg/24 hours in riboflavin deficiency) may also help determine the correct diagnosis. In this patient, a diagnosis was made on the history and clinical findings in this patient, along with normocytic normochromic anemia and change in MCV. Differential diagnosis was kept as ofloxacin induced fixed drug eruption, but no history of recurrence, history of chronic alcoholism, presence of classical lesions, no fluid filled lesions and response to multivitamin therapy favoured the diagnosis of Oculo-Orogenital Syndrome.

Patient was managed on 2 cc of Vitamin B complex injections per day intravenously and an oral multivitamin once per day as definite treatment. The recommended daily value of riboflavin is 0.6mg/1000 kcal. Treatment dosage for deficient infants and children is 1 – 3 mg/day and for adults is 10 – 20 mg/day [2]. It should be taken as part of a B-complex supplement because clinically it is rather difficult to pinpoint to a specific deficiency.

Establishing and maintaining a healthy, well-balanced diet is another important facet of treatment. Dietary counseling with a focus on good eating habits is a must. This patient was given a dietary counseling. Treatment of the underlying cause, if present, must be taken care of. The patient was a chronic alcoholic but refused to undergo treatment for deaddiction.

With vitamin supplements and a nutritious, well-balanced diet, the prognosis is excellent. The vast majority of individuals who follow this treatment regimen (supplements and diet) will achieve a full recovery within a matter of weeks.

CONCLUSION

Oculo-Orogenital Syndrome is a deficiency disorder caused due to deficiency of vitamin B2

(riboflavin) which responds well to multivitamin therapy. Duration depends on the severity of the disease and response to treatment. If the patient is unable or unwilling to follow dietary and therapeutic instructions, disability may be permanent. Cases in which the disorder is related to an underlying illness, treatment of that illness will affect the duration. Special care has to be taken to counsel the patient regarding dietary habits. And psychiatric counselling for patients with eating disorders and addictions has to be given from time to time.

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