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Case Report

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A RARE CASE REPORT ON WERNER'S SYNDROME (PREMATURE AGING SYNDROME)

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ABSTRACT

A Werner's syndrome also known as premature aging syndrome is a rare autosomal recessive disorder caused by genetic change in WRN gene. In this case, a 27-year-old male patient came to a multispecialty hospital with complaints of loss of hair, skin lesions over body with itching, loss of weight, loss of appetite, urinary hesitancy, ulcers in mouth, inability to eat, and difficulty in walking. On examination, multiple hyperkeratotic plaques with scaling all over body with itching, multiple depigmented patches extending to feet, whitish deposit seen over tongue, flat feet, short stature, greying and balding (alopecia) were found to be similar with Werner's syndrome. Patient is diagnosed with PEM (protein energy malnutrition), nutritional anaemia and alopecia universalis. Werner's syndrome is not curable, only symptomatic treatment is provided. This case report provides an insight of Werner's syndrome and its pharmacotherapy on symptomatic basis according to hospital admission guidelines. Early recognition will help for the further complications related to Werner's syndrome and helpful for the further consequences.

KEEYWORDS: Werner's syndrome, hyperkeratotic, autosomal, protein energy malnutrition.

INTRODUCTION

Werner's syndrome was described in 1904 by Otto Werner working at ophthalmological clinic, he studied on 4 brothers and sisters suffering from syndrome and then it was being named Werner's syndrome.[1] Werner's syndrome also known as adult progeria is autosomal recessive inherited disorder characterised by premature aging.[2] It is caused by null mutation of WRN gene, which leads to encoding of RNA helicases. The region between RQC and HRDC has a single strand DNA anneal which has an impact on WRN protein.[3] Mutation in 8Pwrn gene causes Werner's syndrome which leads to increase in cancer risk. The cancer risk is of five tumour types: soft tissue carcinoma, osteosarcoma, lentiginous melanoma, thyroid calcinoma and meningioma.[4] It is characterised by shortness of stature, juvenile cataracts, baldness, greying of hair, tendency to occur in sibling or parental consanguinity, ulcers of legs, high pitched voice, flat feet, metastatic calcifications, osteoporosis, hypogonadism and irregular dental impression.[2] The cause of WS with cell aging is concerned with increased rate of cell production which becomes senescent compared to normal cell growth which leads to increase in senescent WS cell production.[5] The most prominent feature is atrophy and wasting of soft tissue and muscle all over body and scleroderma like lesions on skin. Characteristics such as calcification of cardiac valves and atrophy of testicles and skin appendages are also been observed.[6] Japanese registry has symptoms like bird like face, low bogy weight, short stature, cataract(bilateral) and cardinal sign and symptoms.[3] The diagnostic criteria are based o9n the complications associated with it. The higher the complications associated more is probability. The most significant Achilles' tendon can be seen with calcification, progeroid face, cataracts(bilateral), intractable ulcers and atrophic skin.[7] There is no specific cure for Werner's syndrome. only symptomatic treatment is given. Preventing secondary complications and screening for diseases are done. mTOR signalling and basal autophagy in WS cells, rapamycin had shown increase in growth rate and reduced DNA damage. p38 nitrogen-activated protein kinase are also investigated. hiPCS (human induced pluripotent cells) has shown reversion of senescence related cellular phenotypes.[3] p38 is found have shown increase in replicative life span compared to untreated cells.[8] This syndrome is mostly reported in ophthalmologic or dermatologic department due to its characteristics.[9]

CASE REPORT

A 27-year-old male patient came to multispecialty hospital (date of admission: 22 January 2021) with complaints of loss of hair in the past 10 years, skin lesions over body with itching in the past 6 years, loss of weight from 2 months, loss of appetite in the past 6 days,

urinary hesitancy from 2-3 days, ulcers in mouth from 1 year, inability to eat from 5-6 days, and difficulty in walking from 2 months. Patient developed lesions in the oral cavity (inner lips and buccal cavity) which was associated with burning sensation. Multiple joint pain involving mainly large joints. On the basis of past history included inability to open mouth, and earlier patient had taken treatment for scabies. Patient consulted many local doctors for the same but got only temporary relief. On examination, multiple hyperkeratotoic plaque seen over axilla, abdomen, arms, back, hand, lower limbs, web spaces and forearm. Multiple depigmented patches seen over soles extending to dorsum and feet. Fine scaling seen over periorbital area, cheeks, chin, ears and neck. Whitish deposit seen over dorsum of tongue. On USG ABDOMEN and PELVIS, fatty liver was found. Patient is poorly built and malnourished. He is detected with severe PEM (protein energy malnutrition), nutritional anaemia and alopecia universalis.

Figure 1: Facial feature showing bird like face.

Figure 2: skin lesions all over body.

The patient was admitted for 7 days for suspected symptoms. He was given with following symptomatic treatment: Inj.ceftriaxone (1g) IV 12 hourly, INJ. Fluconazole (400) IV, IVF NS (0.9%) (500) Plus 2 amp optineuron IV 12 hourly, INJ.pheniramine IV 12 hourly, Inj. Pantoprazole (40), inj. Ondansetron(4cc), tab.shelcal (500), richmoist cream 3-4 times a day, tab.limcee(500), vitamin d3 (60,000IU) Sachet once a week, Clotrimazole, tab. Nicoglow, salicylix-sf 6% 2 times a day ,clotrin mouth paint, chlorhexidine mouth gargle ,syrup potklor(2 tsp).Inj.ceftriaxone is prescribed as antibiotic for infection. Inj. Fluconazole is given as antifungal. INJ.pheniramine and tab.pan is given as antiallergic used for skin rashes and itching and to treat acid reflux. Shelcal as calcium supplement. Optineuron is multivitamin with B12 complex. Limcee as vitamin C supplement. Ondansetron as antiemetic. Clotrin and chlorhexidine are given for fungal infection in mouth and germicidal mouth wash. Clotrimazole is given for prevent skin infection. Nicoglow to prevent lack of niacin in body and to lower cholesterol and triglyceride level. Rich moist cream is used to prevent dry and scaly skin. The patient was discharged on 28 January 2021 with following discharge medications: tab. cefixime (100), tab. pheniramine (10), tab calcium plus D3 for 10 days, vitamin D3 sachet once a week, tab. Nicoglow, tab. limcee, richmoist cream, Clotrimazole, salicylix-sf, clotrin, chlorhexidine mouth wash, syrup potklor. After proper patient counselling and providing symptomatic treatment follow up is to be taken after 1 month.

DISCUSSION

A rare case of Werner's syndrome came to our multispeciality hospital with complaints as shown in case report. On examination he was determined with multiple hyperkeratotic plaques with scaling seen over finger, web spaces, knees, popliteal fossa, feet and palms. Few scanty grey hair were seen over scalp. He was diagnosed with Werner's syndrome, protein energy malnourished, hyperalbuminemia, and fungal infection. It was diagnosed from signs and complications which were similar to Werner's syndrome. This case was reported from ophthalmologic department where no ophthalmic abnormalities were detected. He was provided with symptomatic treatment. since there is no specific cure for Werner's syndrome, according to patient's complaints and laboratory data patient was treated. He was advised for high protein diet as he was severely malnourished. With the ongoing treatment patient had a significant improvement in the condition. All the signs are seen during adolescence period but serious complications

such as cataracts, osteoporosis and malignancy are not seen until now. Patient was provided with the patient counselling about the disease and advised for the check-ups in intervals as the disease may progress.

CONCLUSION

In this case study it was observed that the patient was suffering from premature aging. The patient admission guidelines, sign, symptoms and disease progression were found to be similar to Werner Syndrome. The patient was being prescribed with drugs for symptomatic treatment with precautionary drugs as per hospital.

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