

An Atypical Sclerodermatous Condition Revealing Werner Syndrome

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Abstract

Case Report

Sclerodermatous conditions are common and variable; they can mimic systemic sclerosis and reveal underlying internal diseases. Adult progeria, or Werner syndrome, is a rare genetic disorder responsible for multi-organ premature aging and an increased predisposition to neoplasms and cardiovascular diseases. We report a case of Werner syndrome revealed by an atypical sclerodermatous state.

Keywords: sclerodermatous condition, progeria, adult, premature aging.

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INTRODUCTION

Werner syndrome (adult progeria) is a rare autosomal recessive disorder caused by mutations in the DNA helicase gene (WRN) and characterized by features of premature aging. We report a case of Werner syndrome presenting as an atypical sclerodermatous condition.

CASE REPORT

A 41-year-old male patient—with a 15-year history of type 2 diabetes and prior surgeries for bilateral cataracts (8 years ago) and bilateral hydrocele (4 years ago)—was admitted to our department for diagnostic workup and management of cutaneous sclerosis involving the upper and lower extremities.

Dermatological Examination: Short stature, sclerosus-like skin changes on the arms and forearms with hyperpigmentation and limb atrophy, and ulcerations on both feet, without sclerodactyly or other features of systemic sclerosis. The facial appearance was suggestive of Werner syndrome (alopecia, prematurely aged face, dental protrusion, corrective eyeglasses).

Paraclinical Findings: Mixed hyperlipidemia, lumbar spine osteoporosis, and femoral osteopenia. Cardiovascular function was normal, and the paraneoplastic workup was negative.

Genetic Inquiry: Parental consanguinity was noted; testing for the WRN gene could not be performed due to financial constraints.

Outcome: The diagnosis of Werner syndrome was retained based on clinical and paraclinical criteria. The patient received symptomatic treatment with emollients, UVB phototherapy sessions, and local wound care, alongside cardiological, endocrinological, and rheumatological monitoring. The patient was subsequently lost to follow-up.

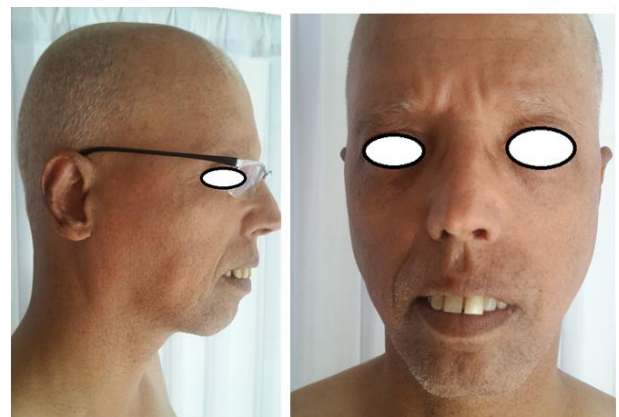


Figure 1: Alopecia, thin gray hair, and dental protrusion



Figure 2: Sclerotic and pigmented skin



Figure 3: Foot ulcers

DISCUSSION

Werner syndrome typically presents in adults during their thirties or forties, although initial signs emerge during adolescence and early adulthood [1][4]. Additional manifestations include a high-pitched voice, ophthalmic complications, hypogonadism, early multifocal atherosclerosis, and benign or malignant tumors. These complications reduce life expectancy, with death generally occurring in the fourth or fifth

decade of life. Inheritance follows an autosomal recessive pattern [2][3].

CONCLUSION

Sclerodermatous conditions are diverse and should be recognized by dermatologists alongside systemic sclerosis and morphea. Werner syndrome is straightforward to diagnose when presenting in its complete form.

Conflicts of Interest: The authors declare no conflicts of interest regarding this work.

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