Werner's Syndrome: A Rare Cause of Hoarseness

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Summary

Werner's syndrome (WS) is a rare hereditary disorder which is characterized by clinical signs of premature aging. A 31-year-old man presented with a 12-year history of hoarseness. Also noted were diabetes mellitus, cataracts, scleroderma-like skin atrophy, osteoporosis, and hypogonadism. A clinical diagnosis of WS was made. Laryngoscopy revealed bowed vocal folds resulting in a spindle-shaped closure with glottal incompetence during phonation. We used Gortex for medialization of the middle part of vocal fold to correct the glottal gap in this patient. Despite correction of glottal incompetence in patients with WS, quality of voice could not be improved to that of age-matched normal individuals.

Key Words: Werner's syndrome; Hoarseness; Premature aging; Thyroplasty

Introduction

Werner's syndrome (WS) is a rare autosomal recessive disorder which is characterized by premature aging.[1] Only 1100 patients are reported worldwide.[2] Signs and symptoms appear when affected individuals are in their 20s or 30s and include loss and graying of hair, cataracts, atrophy of the skin and peripheral fat, and diabetes.[1], [3] and [4] Infertility,

early onset of arteriosclerosis, osteoporosis, cancer predisposition, and hyperlipidemia are other problems in these patients.[1] and [4]

In this paper, we present a case of WS with hoarseness as his initial manifestation. Then, we discuss the modalities of management of hoarseness in patients with WS.

Case report

A 31-year-old man with a 12-year history of hoarseness was referred to our clinic by an endocrinologist. He was diagnosed with diabetes mellitus and hypercholesterolemia about 4 months ago. The chief complaint of the patient was hoarseness. The patient has a history of bilateral cataract, osteoporosis, and hypogonadism. Laryngoscopy revealed bowed vocal cords resulting in glottal incompetence during phonation (Figure 1). This glottal gap led to breathy hoarseness, decreased maximum phonation time, and vocal fatigue. He was short in stature compared with his family members (Figure 2). He complained of several years of skin stiffness. On physical examination, he had stiff and shiny skin similar to those with scleroderma. Additionally, he had several skin ulcerations on his knee and toes. His visual acuity has decreased for 4 months because of bilateral cataract. The patient's diabetes was kept under control with insulin injection for 4 months.

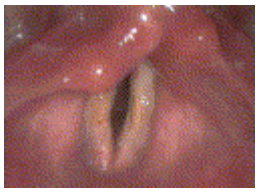 Full-size image (24K)

Figure 1. Laryngoscopic view of the larynx on phonation; note the glottal incompetence and bowing of vocal folds.

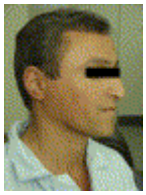 Full-size image (54K)

Figure 2. Atrophic changes in the face of the patient.

A clinical diagnosis of WS was made. The thyroplasty procedure was done for him under local anesthesia. Thyroplasty type 1 surgery approach was done with insertion of a piece of Gortex for medialization of left vocal cord. The vocal folds gap in phonation was closed and the maximum phonation time was increased by surgery but his hoarseness had improved minimally because of the atrophy and stiffness of vocalis muscle and vocal fold membrane. Voice analysis of the patient confirmed an increase in intensity of voice and maximum phonation time and a decrease in minimum, maximum, and mean voice base frequency after thyroplasty. The frequency of generation of vowel /a/ after surgery was 636 Hz compared to 739 Hz before surgery. The intensity of generation of vowel /a/ before surgery was 51 dB which increased to 70 dB after surgery.

Discussion

WS is a rare disorder and its incidence is estimated to be one in one million individuals in the US. Now, about 1100 cases have been reported in the world and more than 800 cases are in Japan.[2]

WS is characterized by clinical signs of premature aging.[1] Its cardinal manifestations are bilateral cataracts, tight and atrophic skin, short stature, premature thinning and graying of scalp hair, diabetes mellitus, hypogonadism, osteoporosis, hoarseness, and mesenchymal neoplasms.[1], [3] and [4] The diagnosis of WS is clinical and not pathological. The disease is inherited as autosomal recessive trait and both sexes are affected equally.[1] The mean survival for patients is 54 years and death usually occurs because of atherosclerosis or malignant tumors.[5]

A hoarse high-frequency voice is a well-known feature of WS.[3] Tsunoda et al reported a case of WS that presented with hoarseness. They transplanted autologous temporal

fascia into the vocal fold for correction of the bowing vocal folds *via* a laryngoscopic approach.[3] We used Gortex for medialization of the middle part of the vocal fold for correction of the glottal gap *via* an open approach. Another procedure that may correct this problem is anterior commissure advancement. So far, there is no published paper to refer to anterior commissure advancement for WS. In old patients, although the quality of voice improves after closure of the vocal fold gap, their voice still lacks youth qualities. The same applies to patients with WS. Their voice does not regain youth qualities even after successful surgery. Although we can do different procedures to close the gap of bowed atrophic vocal cords in these patients, the same quality of voice as age-matched individuals could not be achieved.

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