Management of cataract in Werner syndrome

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Werner syndrome (WS) is a rare progressive disorder. It is characterized by the appearance of unusually accelerated aging (progeria) including bilateral senile cataract. Here, we report a successful management of hypermature cataract in WS.

Key words: Cataract, progeria, Werner syndrome

Werner syndrome (WS) is also known as progeria adultorum or progeria of the adult. It is a rare, autosomal recessive disorder characterized by the appearance of premature or accelerated aging.[1] The occurrence of bilateral cataract at an early age is one of the disabling features of this syndrome. Here, we report a case of hypermature or morgagnian cataract in a case of WS managed by phacoemulsification uneventfully.

Case Report

A 25-year-old female presented with a complaint of diminution of vision both eyes (BE) for the past 6 months and changes in facial appearance for the past few years. The medical history identified no other family members with similar features and no family history of consanguinity. The patient's parents reported that she was of short in stature since childhood and her hair turned grey at around 20 years of age. She had never visited any clinician before this consultation.

General physical examination on admission revealed that she was only 129 cm in height and 28 kg in weight [Fig. 1a]. Her extremities were thin with markedly atrophied skin and decreased subcutaneous fat [Fig. 1d]. Her face appeared older than her age with noticeable sagging and wrinkling of loose facial skin. Her voice was high-pitched and hoarse, and she also had missing teeth with poor oral hygiene [Fig. 1b]. Ocular examination revealed small palpebral aperture with sparse eyelashes in both the eyes [Fig. 1c]. Her distant-corrected visual acuity was the perception of light in BE. The intraocular pressure was within normal limits in BE. The anterior segment showed bilateral cataract in BE. White cataract with multiple calcification spots on anterior capsule of the right eye (RE) and wrinkled anterior capsule of the left eye (LE) was seen consistent with a hypermature [Fig. 2a] or morgagnian cataract [Fig. 2b]. Based on the history and clinical findings, a provisional diagnosis of WS was made.

Biochemical investigations were normal except marginal increased blood glucose, increased erythrocyte sedimentation rate, decreased follicular stimulating hormone, and mild dimorphic anemia.

Cataract surgeries of BE were performed under general anesthesia at the interval of 15 days. The biometric measurement

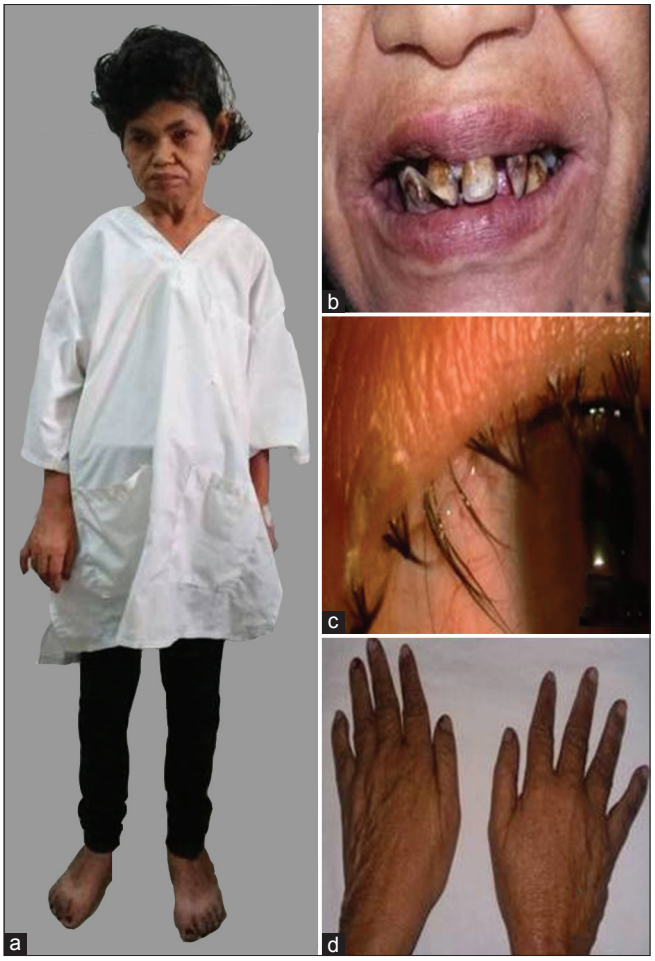

Figure 1: External photograph of a 25-year-old patient with WS showing (a) short stature, (b) poor dentition, (c) sparse eyelashes, (d) wrinkled skin

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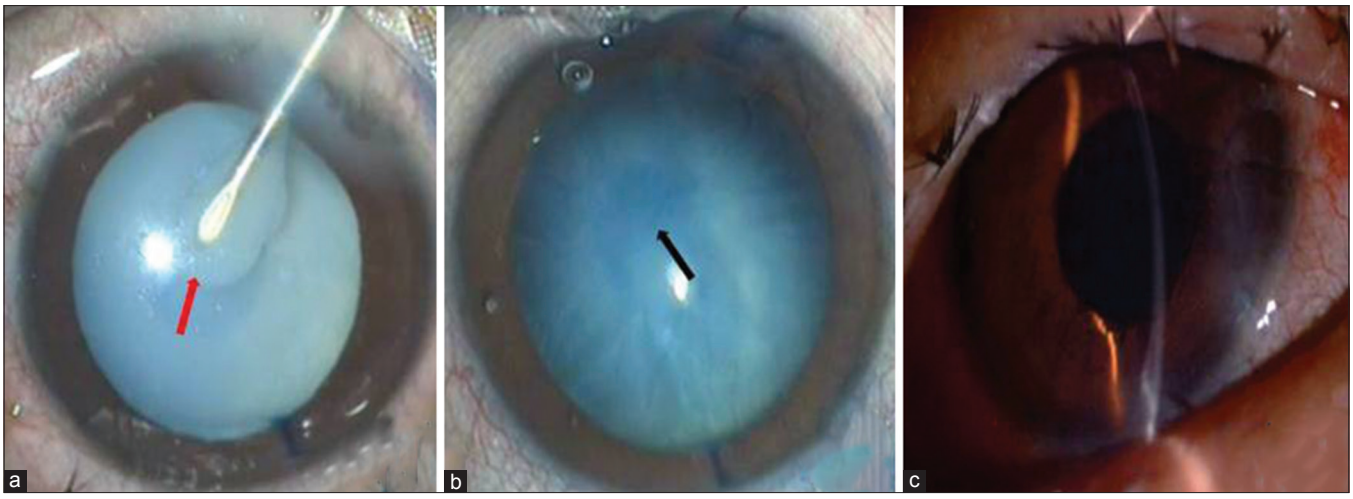

Figure 2: (a) Right eye white cataract with liquefied cortex (red arrow), (b) left eye cataract with wrinkled anterior capsule (black arrow), (c) postoperative photograph showing clear cornea and anterior chamber (right eye)

showed axial lengths of BE to be 21.96 mm (RE) and 22.09 mm (LE), respectively. Sanders-Retzlaff-Kraff II formula was used to calculate intraocular lens power (+22.5 D BE). Temporal clear corneal approach was used in view of the prominent superior orbital rim and small palpebral aperture. Anterior capsulorrhexis was started with needle capsulotome and successfully completed using utrata forceps under a high-viscosity ophthalmic viscosurgical device (OVD). Endothelium was protected using viscodispersive OVD and phacoemulsification (Signature, AMO) was performed under low parameters using the stop, chop, and stuff technique.[2] Bimanual cortical aspiration was done under low aspiration and flow rate followed by foldable acrylic intraocular lens implantation in the bag. Corneal tunnel closure was done using 10/0 nylon suture to prevent wound dehiscence. Postoperative regimen consisted of tapering doses of antibiotic-steroid drops over 6 weeks and followed up for 1 year. Endothelial counts (2454 and 2506 cells/mm^3 in RE and LE, respectively) and normal fundus examination was found at 6 weeks of the follow-up period. Distant-corrected visual acuity of. 22 RE and. 20 LE in LogMAR with surgical-induced astigmatism, against the rule 0.25D was seen in BE.

Discussion

Werner's syndrome was initially described by German scientist Otto Werner in 1904 when he identified the syndrome in four siblings with premature aging.[3] This syndrome rarely appears before puberty, but thereafter is obvious. WS case reports have documented worldwide, the majority being from Japan[1] and only few cases from the Indian subcontinent. Therefore, we believe that our case will add valuable information of disease and management.

WS is a genetic disease transmitted by autosomal recessive inheritance. In Japan, the familial occurrence has been found infrequent since 1996. Hence, most cases recently reported are sporadic.[1] The WRN gene plays an important role in aging and is known to have role in DNA replication, repair of DNA damage, gene transcription, and telomere maintenance. WRN disruption causes WS. Many organs in patients with WS prematurely undergo changes that are usually associated with aging.

In accordance with the diagnostic criteria on the International Registry website, the patient had short stature, bilateral cataract, and typical dermatological pathology including pigmentary alterations, atrophic skin, and graying of the scalp hair.[4] The patient also had hypogonadism, osteoporosis, and hoarse voice. These characteristics in our patient met the diagnostic criteria of "Probable WS."

There are many comorbid conditions in WS, and bilateral cataract is one of them which presents early.[4] Although posterior subcapsular cataract is considered to be the most common, our case presented with hypermature cataract with rapid decrease in vision and increase in clouding of the lens BE.

Before small incision cataract surgery era, Jonas *et al*. reported complications of cataract surgery in WS; postoperative wound dehiscence presented in 10 of 18 eyes that underwent intracapsular and manual extracapsular cataract surgery.[5] The researchers suggested that decreased fibroblast proliferation could be related to that high rate of wound-related complications and that the use of corticosteroids could be a worsening factor. Few studies suggested in aging or premature aging, the mitogenic and stimulatory effects of fibroblast growth factors and other agents such as stimulatory cytokines are significantly reduced and increased response to inhibitory cytokines. These changes may result in significant healing problems in the elderly.[6] In addition to impaired wound healing, secondary glaucoma with bullous keratopathy was also reported in few reports, and subsequently, these patients needed penetrating keratoplasty.[7] Keeping the occurrence of impaired wound healing in WS, we closed corneal wound with suture to prevent wound dehiscence.

We have highlighted that small incision phacoemulsification through temporal approach is safe in these cases. To the best of our knowledge, only few cases were managed with phacoemulsification with uneventful follow-up.[8-10] We performed phacoemulsification with 2.8 mm small temporal incision along with all the precautionary measures to avoid endothelial damage and wound dehiscence [Fig. 2c].

Conclusion

In conclusion, we put emphasis on the importance of safe small incision cataract surgery and modern phacoemulsification to get well-constructed corneal wounds in WS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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