INTRODUCTION

The term *Staphyloma* is derived from “Staphylos”, meaning grape in Greek. It is generally described as scleral ectasia due to the localized thinning and expansion of the posterior scleral wall. Posterior staphyloma is usually seen in pathological myopia, but it might also be seen congenitally as an isolated developmental anomaly.1

Retinitis pigmentosa is characterized by the classical triad of waxy optic disc, arteriolar narrowing and bone-spicule pigmentation in the retina. The main complaints are nyctalopia and progressive visual loss. The diagnosis of RP is supported by full-field flash ERG. In this report, we present a case whose examination unveiled an association of posterior staphyloma and Retinitis Pigmentosa (RP).

CASE REPORT

A 22-year male presented with visual deterioration and progressive nyctalopia that started from the early childhood. Anterior segment biomicroscopy was unremarkable in both eyes. Slit lamp examination revealed bilateral vitreous degeneration. Fundus examination revealed mid-peripheral bone spicule pigmemtations and large posterior staphyloma in both eyes (Figures 1A and 1B). Axial length measured by A-mode ultrasonography was 27.05 mm in OD and 27.80 mm in OS. B mode ultrasonography revealed localized ectasia resembling posterior scleral staphyloma in both eyes (Figures 1C and 1D). Cycloplegic refraction was -6.00 (-1.25 x 175) dpt in the right eye and -7.00 (-1.50 x 165) dpt in the left eye. Slit lamp examination showed normal anterior segment. Fundoscopic examination revealed bone-spicule pigmemtations, waxy optic disc, arteriolar narrowing and posterior staphyloma, with 5 optic disc in diameter in the right eye and 4 optic disc diameter in the left. Ocular ultrasonography confirmed staphyloma. Full-field ERG showed evidence of a generalized retinal dysfunction involving both rod and cone responses, supported the diagnosis of retinitis pigmentosa.

![Figure 1: (A,B) Posterior staphyloma with apparent chorioretinal atrophy and mid-peripheral bone spicule pigmentation in fundus images. (C,D) B-mode ultrasonography showing localized posterior expansion of the posterior pole of both eyes.](image-url)

**Key Words:** Posterior staphyloma. Retinitis pigmentosa. Moderate myopia.

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DISCUSSION

Retinitis pigmentosa is a generic term used for a group of hereditary retinal diseases that are characterized by degeneration of photoreceptors and pigment accumulation in the retina. RP is mostly an isolated disease in the retina. However, it may also be a component of a syndrome such as Bardet-Biedl syndrome or Bassen-Kornzweig disease.

Pathological myopia is defined as the progressive elongation of anterior-posterior diameter of scleral wall (more than -6.00 D or axial length longer than 26 mm). The mechanical stretching of the tissues leads to secondary pathologic ocular changes. One of these changes is posterior staphyloma. Posterior staphyloma is a type of ectasia, localized usually at the posterior pole of the eye, which results from localized expansion and thinning of the sclera. It can be seen in 90% of pathological myopia patients. It is a distinctive feature of pathological myopia and its prevalence increases with the age. Posterior staphyloma also increases the risk of visual loss, chorioretinal atrophy, subretinal neovascularization, macular retinoschisis and macular hole formation. Ocular B-mode ultrasonography is a rapid and effective method in detecting posterior staphyloma.

A few case reports describe MRCS syndrome (microcornea, retinal dystrophy, cataract, and staphyloma). It is inherited as an autosomal trait and patients complain night blindness and poor vision due to the cataract during teenage. Chronic angle closure glaucoma can be seen in older individuals. Retinal abnormalities are present ranging from peripheral retinal pigment epithelium atrophy to retinal pigmentation. Genetic source of the syndrome is likely to be a mutation in a major developmental regulator gene and it may lie in the nanophthalmos locus on chromosome 11, possibly allelic with the NNO1 gene. It can be speculated that this case may be a clinical subtype of MRCS syndrome. The bone spicule pigmentation, waxy optic disc and arteriolar narrowing together with non-recordable full-field flash ERG responses, in this patient led to the diagnosis of retinitis pigmentosa. In MRCS syndrome, peripheral retina pigment epithelium atrophy, pigmentary abnormalities and ERG depressions are not significant, especially in the first two decades. Absence of microcornea and cataract are other distinguishing features of this case from MRCS syndrome. So, the authors cannot attribute this association to MRCS syndrome. To the best of authors' knowledge, this is the first report presenting such an isolated coexistence of posterior staphyloma, retinitis pigmentosa and moderate myopia.

REFERENCES