Letter to the Editor

Coexistence of Retinitis Pigmentosa and Coats-Like Vitreoretinopathy

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Dear Editor,

This letter shares the multimodal imaging findings, genetic profile, and therapeutic approach of a case of retinitis pigmentosa (RP) presenting with signs of Coats-like retinopathy. RP is a hereditary, progressive degenerative disease of the retina. The disease initially manifests with night blindness and peripheral visual field loss, while severe central vision loss may develop in later stages.[1] Although RP is generally observed as an isolated ocular condition, it may also be associated with systemic diseases and other ophthalmologic pathologies. A well-known example is Usher syndrome, in which RP is accompanied by hearing loss.[2] Another ophthalmologic pathology reported in association with RP is Coats-like vitreoretinopathy, a combination referred to as Coats-like RP.

Coats-like RP can present with abnormal telangiectatic vessels in the retina and subretinal lipid exudation. Although the exact etiology and genetic background of Coats-like RP have not been fully elucidated, it has been reported to be associated with mutations in various genes, such as Norrie disease protein and Crumbs cell polarity complex component 1 (*CRB1*).[3] While several cases of Coats-like RP have been reported from Türkiye and worldwide, data regarding the genetic background and treatment approaches of such cases remain limited.

A 38-year-old woman presented to our clinic with complaints of bilateral low vision since childhood. The patient reported also having hearing loss since childhood, but no other known systemic disease. There was no parental consanguinity or family history of similar complaints.

The patient underwent a detailed ophthalmologic examination. Her best-corrected visual acuity (BCVA) according to Snellen chart was 3/20 in the right eye, while vision in the left eye was limited to light perception. Intraocular pressures and anterior segment examination findings were normal in both eyes. Fundus examination revealed a blunted macular reflex, peripheral retinal atrophy, and bone spicules bilaterally. Additionally, bilateral subretinal lipid exudation and telangiectatic changes were noted inferior to the optic disc and along the inferior arcuate vessels (Figure 1).

Swept-source optical coherence tomography (OCT) (Topcon, Tokyo, Japan) demonstrated a central macular thickness of 507 µm in the right eye and 363 µm in the left eye, as well as bilateral diffuse intraretinal cystic spaces and disruption of the ellipsoid zone (Figure 2). Wide-field fundus fluorescein angiography (Optos, USA) revealed bilateral telangiectatic vessels and vascular leakage in the inferior retina (Figure 3). The patient's findings were consistent with Coats-like RP, and a variant of Usher syndrome was suspected due to her concomitant hearing loss. Whole-exome sequencing identified a homozygous *USH2A* mutation (c.12706T>G). Audiological evaluation confirmed sensorineural hearing loss.

Keywords: Retinitis pigmentosa, Coats-like vitreoretinopathy, telangiectasia, photocoagulation

Cite this article as: Gönül Ş, Erdem A, Saylam H, Marzioğlu Özdemir E. Coexistence of Retinitis Pigmentosa and Coats-Like Vitreoretinopathy. Turk J Ophthalmol. 2026;56:283-286

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Received: 31.10.2025
Revision Requested: 07.01.2026
Last Revision Received: 19.01.2026
Accepted: 27.02.2026
Publication Date: 26.08.2026

DOI: 10.4274/tjo.galenos.2026.39038

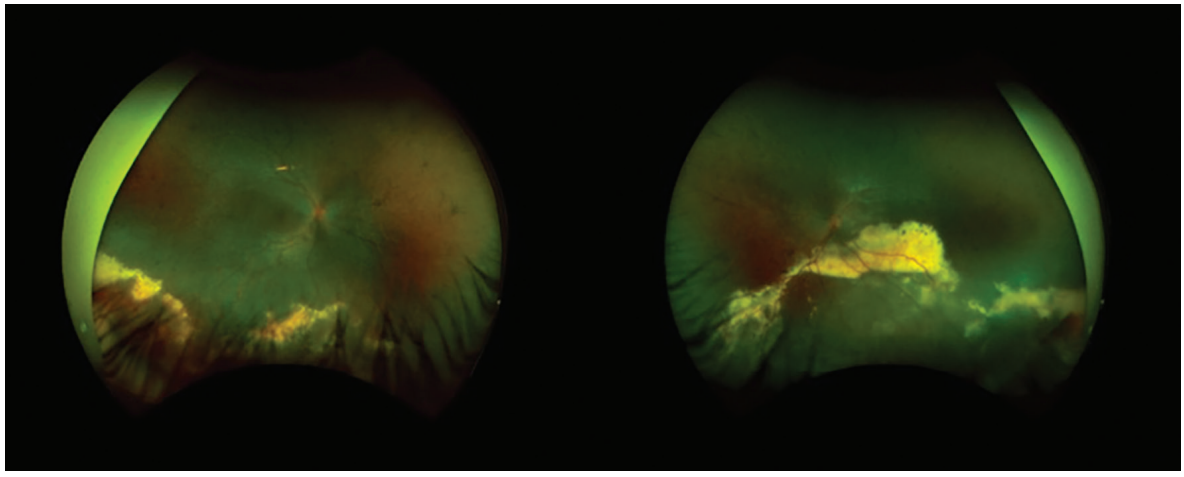

Figure 1. Bilateral subretinal lipid exudation and telangiectatic changes in the inferior retina and bone spicules in the peripheral retina were observed. The exudation extended to the macula in the left eye (left)

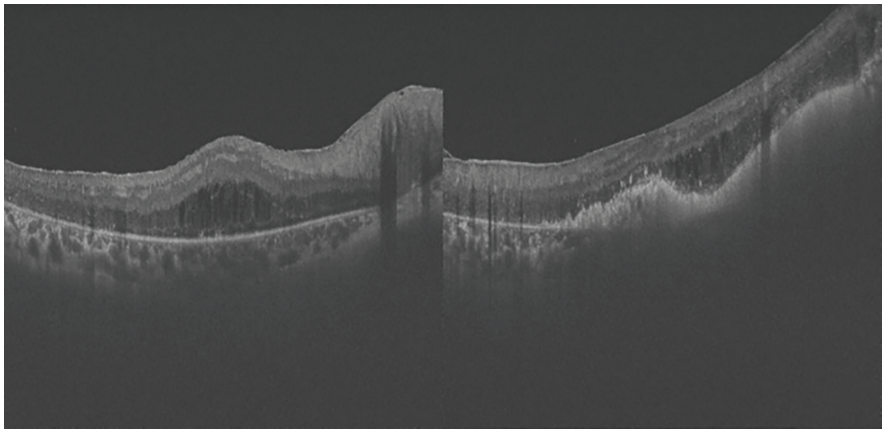

Figure 2. Swept-source optical coherence tomography revealed bilateral diffuse cystic cavities, atrophy of the outer retinal layers, and deposition consistent with subretinal lipid exudation on the left

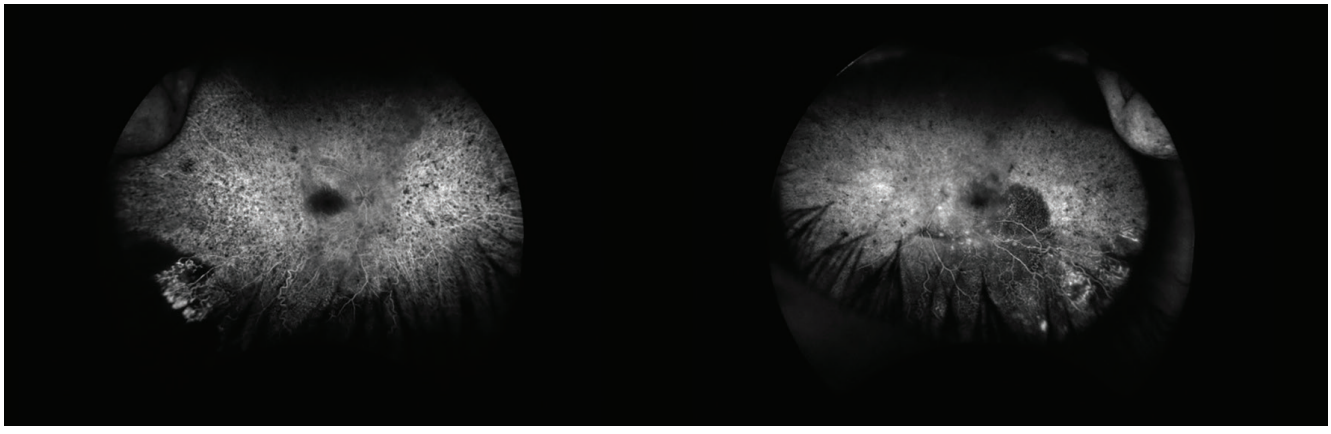

Figure 3. Wide-field fundus fluorescein angiography demonstrated telangiectasia and vascular leakage in the inferior retina bilaterally

Monthly injections of intravitreal bevacizumab (Avastin; Genentech, USA) were administered three times to the right eye and twice to the left eye. Fifteen days after the injections, photocoagulation was applied bilaterally to areas with telangiectatic vessels and vascular leakage in the inferior quadrants and ischemic areas in the superior quadrants.

At the 1-month follow-up after photocoagulation, bilateral serous retinal detachment was detected in the inferior quadrants, with elevated areas of persistent telangiectatic vessels. Consequently, 180-degree transscleral cryotherapy was applied to the inferior retinal quadrants of both eyes in a single session. Cryotherapy was applied to the affected areas using a double freeze-thaw technique in a single session per eye.[4]

At the one-month follow-up after cryotherapy, Snellen BCVA was still 3/20 in the right eye and light perception in the left eye. On fundus examination, photocoagulation and cryotherapy scars were present in addition to the initial findings, and subretinal exudation was observed inferior to the optic disc. Photocoagulation was performed on the retinal area between the inferior arcuate vessels and the ischemic zone in the right eye (Figure 4).

At the patient's most recent examination, after two years of follow-up, BCVA remained unchanged at 3/20 in the right eye and light perception in the left eye. Fundus examination revealed persistent exudation. Central macular thickness was 524 µm in the right eye and 291 µm in the left eye, and OCT findings were similar to those observed in the initial examination (Figure 5).

Aflibercept was considered as an alternative anti-vascular endothelial growth factor (anti-VEGF) agent during follow-up. However, it could not be administered because the patient could not afford the medication. Furthermore, follow-up and treatment intervals could not be optimized because the patient was unable to attend scheduled examinations regularly due to socioeconomic limitations.

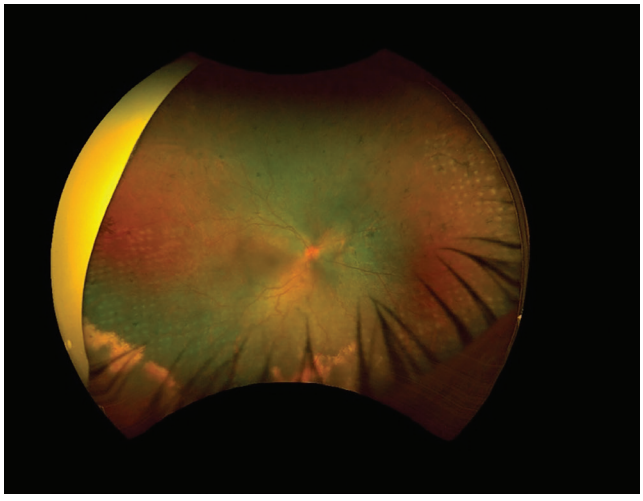

Figure 4. Wide-field fundus image of the right eye following photocoagulation

RP can coexist with a variety of systemic and ocular pathologies. When associated with systemic disease, the coexistence is referred to as syndromic RP. The association of RP with Coats-like lesions was first described in 1988.[5] Coats disease typically affects males in early childhood and is usually unilateral.[4] In contrast, Coats-like lesions associated with RP are usually detected in adulthood, exhibit bilateral involvement, and present with telangiectatic changes and lipid exudation in the inferior retina.[3] Consistent with previous reports in the literature, our patient was an adult woman with RP who exhibited telangiectasia and exudation involving the inferior retina.

Although our patient's retinal findings resembled those in previously reported cases, the presence of sensorineural hearing loss suggested the possibility of a syndromic condition such as Usher syndrome. In a cohort study conducted by Daich Varela et al.[6], Coats-like retinopathy associated with hereditary retinal diseases was reported to have a considerably heterogeneous genetic basis, occurring in the context of various gene mutations. The most common associations were with *CRB1*, *RHO*, and *USH2A* mutations, respectively.[6]

To the best of our knowledge, there are limited data on the underlying genetic mutations and treatment approaches in similar cases reported from Türkiye. Demirel et al.[7] reported a case with clinical findings similar to ours but provided no information regarding the genetic basis, and the patient was managed conservatively with preservation of visual acuity. Another three-case series from Türkiye reported clinical findings similar to ours but stated that genetic testing could not be performed.[8] In our patient, a homozygous *USH2A* mutation was identified, although it was classified as a variant of uncertain significance. Given the consistency between the mutation and the clinical findings, we believe the patient may have a variant of Usher syndrome with associated Coats-like RP. However, broader genetic analyses including the patient's parents or confirmation through experimental animal model studies, would be required to establish this mutation as a novel variant. Another case report described Coats-like RP findings in a patient with nanophthalmos.[9] These reports suggest that Coats-like RP may occur in association with systemic syndromes as well as other ocular pathologies, which complicates both the diagnosis and the management of the disease.

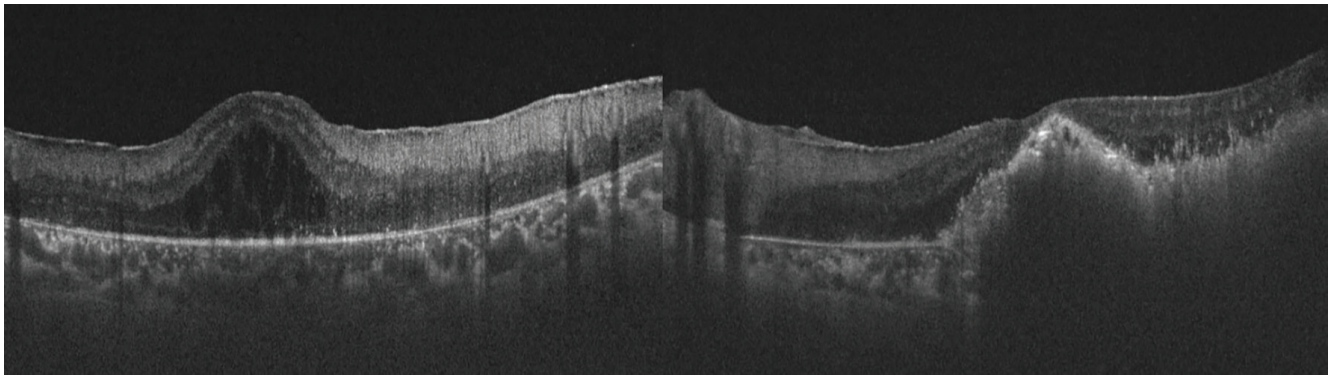

Figure 5. At the patient's final follow-up examination, swept-source optical coherence tomography images of the right (right) and left (left) eyes appear similar to the initial findings

The mainstay of treatment in RP patients with Coats-like retinopathy is ablation of all telangiectatic vessels and ischemic retina, sparing the parafoveal region. For this purpose, laser photocoagulation may be applied to non-elevated retinal areas, whereas cryotherapy is preferred for areas with extensive subretinal fluid and dense exudate. Anti-VEGF agents may be used as adjunctive therapy. Vitreoretinal surgery may also be required in patients with massive serous retinal detachment.[8] Although anatomical improvement following treatment has been reported in some cases, functional recovery remains limited.[3] In our case, treatment consisted of laser photocoagulation, intravitreal anti-VEGF injections, and cryotherapy. Over the two-year follow-up, no significant functional improvement was achieved due to retinal damage from both RP and the associated vascular pathology. Nevertheless, potential complications secondary to the vasculopathy were prevented, and anatomic integrity was preserved.

In conclusion, we report a case of Coats-like RP and sensorineural hearing loss associated with a homozygous *USH2A* gene mutation. However, larger case series and more detailed genetic analyses are needed to corroborate the pathogenicity of this mutation. Although such cases pose diagnostic challenges due to their relative rarity, they may carry a poor prognosis due to the multifactorial retinal damage involved. In similar cases, accurate diagnosis requires detailed multimodal imaging assessment, evaluation for additional systemic pathologies, and effective genetic counseling. It should also be kept in mind that combined treatment modalities may be necessary to halt disease progression.

Ethics

Informed Consent: Informed consent was obtained from the patient for all findings, genetic and laboratory results, and images related to this case.

Declarations

Authorship Contributions

Surgical and Medical Practices: Ş.G., A.E., Concept: Ş.G., A.E., H.S., E.M.Ö., Design: Ş.G., A.E., H.S., E.M.Ö., Data Collection or Processing: Ş.G., A.E., H.S., E.M.Ö., Analysis or Interpretation: Ş.G., A.E., H.S., E.M.Ö., Literature Search: Ş.G., A.E., H.S., E.M.Ö., Writing: Ş.G., A.E., H.S., E.M.Ö.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Liu W, Liu S, Li P, Yao K. Retinitis pigmentosa: progress in molecular pathology and biotherapeutical strategies. Int J Mol Sci. 2022;23:4883.
2. Janáky M, Braunitzer G. Syndromic retinitis pigmentosa: a narrative review. Vision (Basel). 2025;9:7.
3. Moinuddin O, Sathrasala S, Jayasundera KT, Branham KH, Chang EY, Qian CX, Recchia FM, Fahim AT, Besirli CG. Coats-like exudative vitreoretinopathy in retinitis pigmentosa: ocular manifestations and treatment outcomes. Ophthalmol Retina. 2021;5:86-96.
4. Sen M, Shields CL, Honavar SG, Shields JA. Coats disease: an overview of classification, management and outcomes. Indian J Ophthalmol. 2019;67:763-771.
5. Khan JA, Ide CH, Strickland MP. Coats'-type retinitis pigmentosa. Surv Ophthalmol. 1988;32:317-332.
6. Daich Varela M, Conti GM, Malka S, Vaclavik V, Mahroo OA, Webster AR, Tran V, Michaelides M. Coats-like vasculopathy in inherited retinal disease: prevalence, characteristics, genetics, and management. Ophthalmology. 2023;130:1327-1335.
7. Demirel EE, Dikci S, Genç O, Yılmaz T. Coats-like retinitis pigmentosa. Ret-Vit Özel Sayı. 2015;23:187-190.
8. Kan E, Yilmaz T, Aydemir O, Güler M, Kurt J. Coats-like retinitis pigmentosa: reports of three cases. Clin Ophthalmol. 2007;1:193-198.
9. Urgancioglu B, Ozdek S, Hasanreisoglu B. Coats'-like retinitis pigmentosa variant and nanophthalmos. Can J Ophthalmol. 2007;42:877-878.