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GW Ophthalmology On-Call: May 2023

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OPHTHALMOLOGY ON CALL

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George Washington University

Department of Ophthalmology

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Introduction

GW Ophthalmology On Call is a monthly case report publication presented by medical students and residents at the GW Department of Ophthalmology. The purpose of this publication series is to highlight educational content seen at the GW Department of Ophthalmology. This content is for teaching purposes only and should not be used to guide treatment. All case presentations have been written in accordance with HIPAA guidelines. Case details have been modified for patient privacy purposes.

Endophthalmitis

Priyanka Bhatnagar (MS2), Myra Zaheer (MS2), Will West, MD, David Belyea, MD, MBA

Chief Complaint
Eye pain and blurry vision

History of Present Illness
A 68-year-old female with a significant medical history including left eye Uveitis, retinal detachment in the same eye for which she underwent scleral buckle surgery in 1970 (the buckle was removed in 1996), myopia in both eyes, cataract surgery in the left eye carried out in 1996, and left eye Glaucoma treated with xpress shunt in 2010, presented with symptoms of left eye pain and irritation. These symptoms had been ongoing for 36 hours and were initially accompanied by floaters in the left eye. However, a graying of vision started the morning of her visit, and it had progressively worsened to a state of near total vision loss. The patient denied experiencing any recent trauma to the eye, any ocular discharge, or light flashes.

Ocular History
Glaucoma OS s/p xpress shunt (2008)
Retinal detachment OS s/p scleral buckle (1972)
Scleral buckle removal (1992)
Uveitis
Pseudophakia OS (1992)

Past Medical History
None

Medications
PF 1% OS BID

Allergies
Penicillin

Family History
None

Social History
None

Review of Systems

Denies fever, chills, nausea, vomiting, chest pain, SOB, diarrhea/constipation, urinary symptoms, weight loss, fatigue

Ocular Exam

Physical Exam		
	OD	**OS**
Visual Acuity	20/25-2	CF at 6 ft
Intraocular Pressure (mmHg)	10	44
Pupils	Round and reactive	Round and reactive
Extraocular Motility	EOMI	EOMI

Slit Lamp Exam		
	OD	**OS**
External	Unremarkable	Unremarkable
Lids and Lashes	Normal, no lesions	Mild eyelid edema, mild ptosis
Conjunctiva/Sclera	White and quiet	3+ injection, slight mucopurulent discharge. Shallow, yellow appearing avascular thing wall bleb with patches of epithelial erosions without leak filled with fibrin at 10:00. Pale appearance and fluorescein stain overlying the bleb. No visible sutures.
Cornea	Clear	Haze
Anterior chamber	Deep and quiet	Hypopyon, fibrin/pupillary membrane, xpress shunt seen with associated fibrin
Iris	Round and reactive	Poorly reactive surgical pupil. Peaked inferiorly with possible surgical iridectomy
Lens	1+ NS	Clear

Dilated Fundus Exam		
	OD	**OS**
Cup to Disc	0.4	No view due to anterior chamber findings
Optic Nerve	Normal	n/a
Vessels	Normal caliber and course	n/a
Macula	Normal	n/a
Periphery	Attached 360 degrees w/no RT/RD noted	n/a
Vitreous	Clear	n/a

Imaging

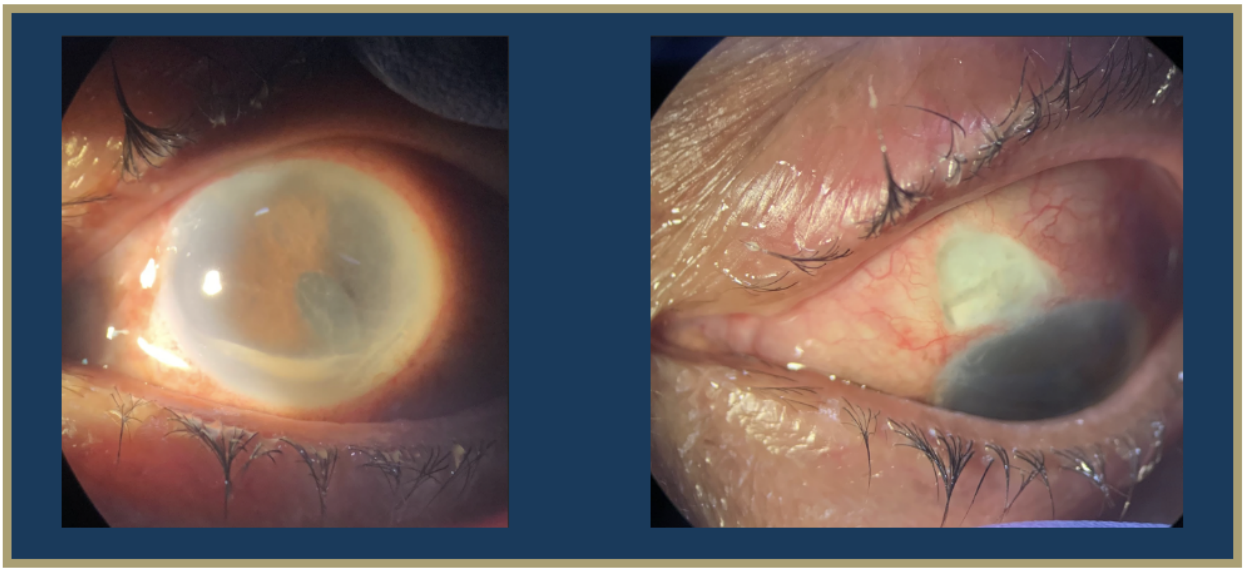

Figure 1: Slit lamp photo of left eye demonstrating corneal haze, hypopyon and conjunctival injection with inferotemporal peaking

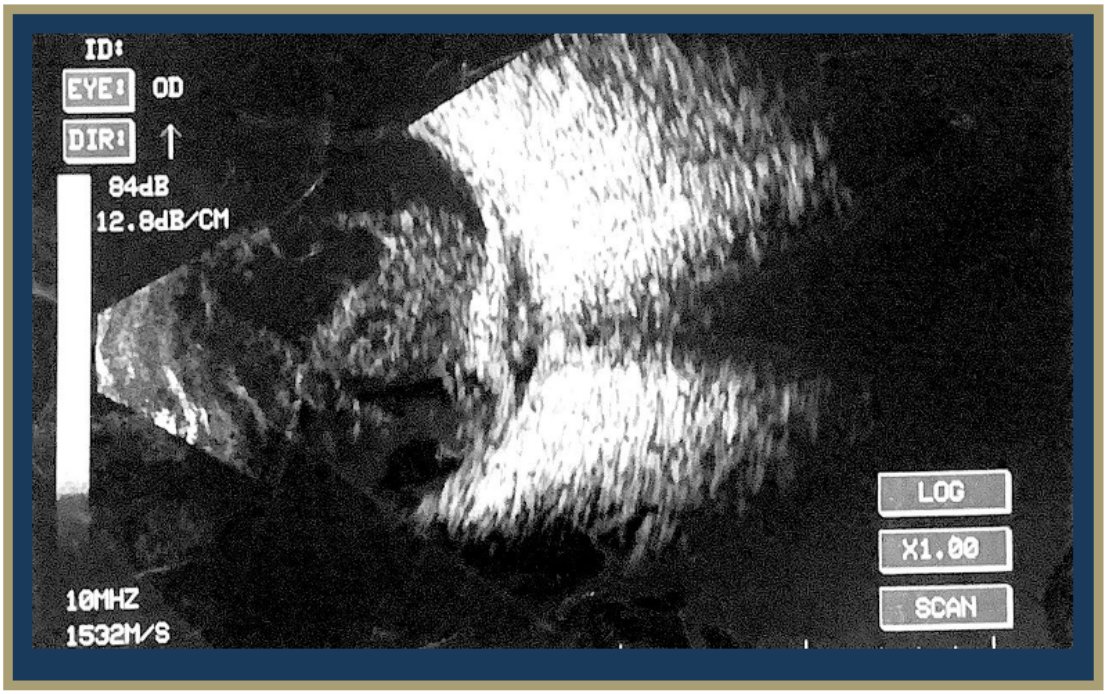

Figure 2: B-scan ultrasound OS reveals no vitritis and a small retinal detachment in the posterior pole.

Differential Diagnosis

Endophthalmitis
Uveitis
Pseudoendopthalmitis from intravitreal injections

Clinical Course

A 68-year-old female with an extensive ocular history, which included left eye uveitis, retinal detachment in the left eye (treated with scleral buckle surgery in 1970, buckle removed in 1996), myopia in both eyes, cataract surgery in the left eye (1996), and glaucoma in the left eye treated with an Xpress shunt in 2010, presented on Day 0 with acute left eye pain and vision loss. Slit lamp examination of the left eye displayed corneal haze, hypopyon, and conjunctival injection with inferotemporal peaking. Elevated intraocular pressure and additional findings of a compromised epithelium over the patient's filtering bleb with fibrin within the bleb and extending from the xpress shunt raised concerns for exogenous endophthalmitis. A B-scan ultrasound OS revealed a small retinal detachment in the posterior pole. The patient was initiated on topical antihypertensives and received intravenous Diamox for acute intraocular pressure management. An anterior chamber tap had reduced the IOP to 28 mmHg, and a vitreous tap along with an injection of intravitreal vancomycin, ceftazidime, and dexamethasone was carried out; samples were sent for culture. The patient was discharged on Vancomycin OS QID,

Tobramycin OS QID, and Cyclopentolate OS BID for endophthalmitis, as well as Cosopt OS BID and Brimonidine OS BID for intraocular pressure management.

On Day 1, the patient reported reduced pain (1/10) and light sensitivity while on brimonidine, dorzolamide, and cyclopentolate (timolol was not started). Slit lamp examination was consistent with initial findings, and the IOP had further decreased to 4. The exam showed increased corneal edema, a deep AC with unchanged hypopyon, and a bleb with decreased fibrin and increased clarity. Diamox, brimonidine, and timolol were discontinued; the patient remained on vancomycin and tobramycin Q2H and cyclopentolate BID.

By Day 2, the patient had reported stable vision and absence of pain. The slit lamp examination had shown improvement, including a visible central pupil. IOP was measured at 9, with no bleb leak. Cultures had tested positive for Streptococcus mitis oralis. The patient continued on vancomycin Q2H, tobramycin Q2H, and cyclopentolate BID.

On Day 6, the patient began experiencing intermittent pain, and the IOP was measured at 22. Cultures had indicated susceptibility to ceftriaxone and vancomycin. The patient had continued on vancomycin and tobramycin QID, and medications were reinitiated with dorzolamide-timolol OS BID and prednisone BID OS.

By Day 8, the patient had reported no significant changes in vision, pain or discomfort, or visual disturbances (flashes/floaters). Slit lamp examination had revealed a new hyphema, and a B-scan showed a vitreous hemorrhage. The patient's endophthalmitis was successfully treated over the subsequent weeks, resulting in full resolution. The patient continues to be under observation and follow-up care.

Discussion

Endophthalmitis, a severe vision-threatening condition, involves a pus-producing inflammation of the internal eye tissues, provoked by bacterial or fungal contamination of the vitreous cavity.[1] This infection contrasts with panophthalmitis where the infection extends to the orbital soft tissues.[2] The path of entry classifies endophthalmitis into exogenous and endogenous types. External injuries such as those from ocular surgery (post-operative endophthalmitis) or trauma (post-traumatic endophthalmitis) lead to exogenous infections.[1] In contrast, endogenous infections result from hematologic spread. Endophthalmitis related to filtering bleb treatment for glaucoma is termed bleb-related endophthalmitis.[2]

The frequency of endophthalmitis types varies according to geographical location and the individual ophthalmic center. Acute postoperative endophthalmitis commonly ensues after intravitreal anti-VEGF injections (0.025% to 0.2% per injection) or cataract surgery (0.1% of cases).[2] Traumatic endophthalmitis accounts for 25% of endophthalmitis cases and often occurs after an injury involving an intraocular foreign body.[3] In endogenous bacterial endophthalmitis, an extraocular source can be traced in 90% of cases, such as dental abscesses, pneumonia, endocarditis, bacterial meningitis, liver abscesses, and urinary tract infections.[1] Risk factors comprise immune system deficiencies, recent invasive medical procedures, and the presence of intravenous lines.[1]

Different pathogens are implicated in endophthalmitis based on its etiology. Coagulase-negative staphylococci and other gram-positive cocci typically cause acute post-cataract endophthalmitis, whereas Propionibacterium acnes generally cause chronic

post-cataract endophthalmitis.[2] Coagulase-negative staphylococci and viridans streptococci are common pathogens in post-injection endophthalmitis.[2] Streptococcus and H. influenzae are usually responsible for bleb-related endophthalmitis.[2] Bacillus cereus is the primary pathogen in post-traumatic endophthalmitis, while S. aureus, streptococci, E. coli, and Candida make up most cases of endogenous endophthalmitis in the US.[2]

The physical examination findings in endophthalmitis patients can be diverse, depending on the infection's cause and severity. Classic presentations of endophthalmitis include rapidly progressing pain (75% of cases), reduced vision (95%), and a red eye (80%)2. The most commonly observed signs are hypopyon formation (>80%)2 and eyelid swelling1, with progressive vitritis marking all forms of endophthalmitis.[4] Specific signs frequently correlate with the infection's origin. Traumatic endophthalmitis may exhibit enhanced ocular inflammation related to the injury. [1] Symptoms of endogenous endophthalmitis may include floaters, light sensitivity, and systemic signs such as sepsis, vomiting, nausea, and fever.[1] A prelude of headache and brow pain may signify filtering bleb-associated endophthalmitis.[1] Regular assessment and documentation of signs and symptoms are crucial for tracking disease resolution.

Imaging tools like optical coherence tomography (OCT), ultrasound, and fundus photography are integral in evaluating endophthalmitis. OCT allows for visualization of the retinal layers, assisting in identifying features like retinal thickening and subretinal fluid that are useful in evaluating disease severity and treatment response.[5] Ultrasounds can offer additional information about vitreous opacities, choroidal thickening, and retinal detachment.[6] Fundus photography records retinal findings like retinal infiltrates and hemorrhages, which are beneficial in monitoring disease progression.[7] Laboratory tests to identify the causative microorganisms should include vitreous and aqueous humor cultures, Gram stain, polymerase chain reaction (PCR), and serological studies.[8]

A combination of medical and surgical interventions is essential in treating endophthalmitis. Medical interventions often include intravitreal antimicrobial agents and systemic antibiotics. Intravitreal antibiotics such as vancomycin and ceftazidime target Gram-positive and Gram-negative pathogens respectively.[9] Surgical interventions may involve vitrectomy, the removal of infected vitreous, in severe inflammation or cases with poor visual prognosis.[10]

Prompt and precise diagnosis of endophthalmitis, a severe intraocular infection, is crucial to preserving visual function and averting complications. The assimilation of clinical signs, comprehensive history, and corroborative laboratory tests aids in diagnosing endophthalmitis. This scenario illustrates how timely intervention and continuous monitoring can enhance treatment outcomes in future endophthalmitis cases.

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Retinitis Pigmentosa Inversa

Robert Thomasian (MS4), Jacob Diaz (MS3), Marena Patronas, MD

Chief Complaint
Referral for atypical retinal changes

History of Present Illness
An 84 year old male with a 10 year history of central retinal changes presented to the ophthalmology clinic several years ago. He complained of poor visual acuity which had not worsened in the past year. He denied any eye pain, photophobia, flashes, or floaters.

Ocular History
None

Pertinent Past Medical History
Type 2 DM w/o retinopathy
Hypertension w/o retinopathy
PVD OU
Blepharitis OU
Cataracts OU - patient deferred CE due to poor underlying vision

Medications
Atenolol
Amlodipine
Aspirin
Tamsulosin
Atorvastatin

Allergies
ACE inhibitors

Family History
None

Social History
Former smoker

Review of Systems

All other systems intact

Ocular Exam

Physical Exam		
	OD	**OS**
Visual Acuity	20/400	20/400
Intraocular Pressure (mmHg)	16	16
Pupils	Round and reactive	Round and reactive
Extraocular Motility	EOMI	EOMI

Slit Lamp Exam		
	OD	**OS**
External	Unremarkable	Unremarkable
Lids and Lashes	mild MGD	mild MGD
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Arcus, otherwise clear	Arcus, otherwise clear
Anterior chamber	Deep and quiet	Deep and quiet
Iris	Round and reactive	Round and reactive
Lens	2+ NS with early cortical changes	2+ NS with early cortical changes

Dilated Fundus Exam		
	OD	**OS**
Cup to Disc	0.7	0.6
Optic Nerve	Normal	Normal
Vessels	Attenuated	Attenuated
Macula	RPE changes	RPE changes
Periphery	Vessel attenuation, diffuse central/peripheral RPE atrophy, pigment clumping 360° in periphery	Vessel attenuation, diffuse central/peripheral RPE atrophy, pigment clumping 360° in periphery
Vitreous	PVD	PVD

Imaging

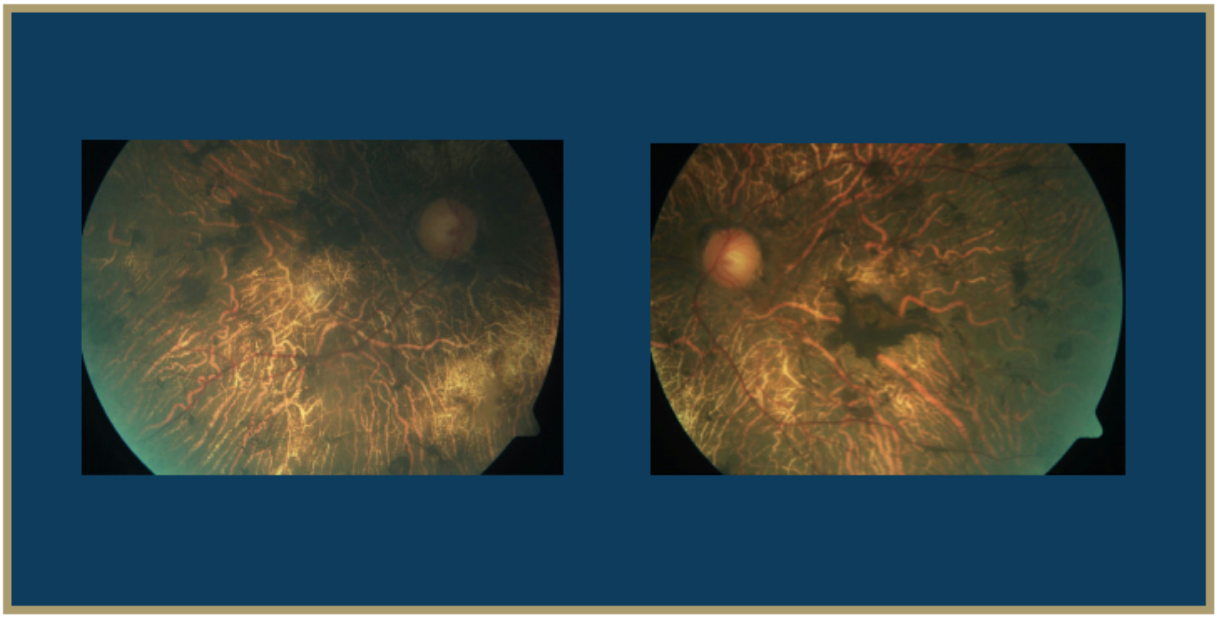

Figure 1: Fundus imaging showing sharp optic nerve with uniformly distributed pigment clumps.

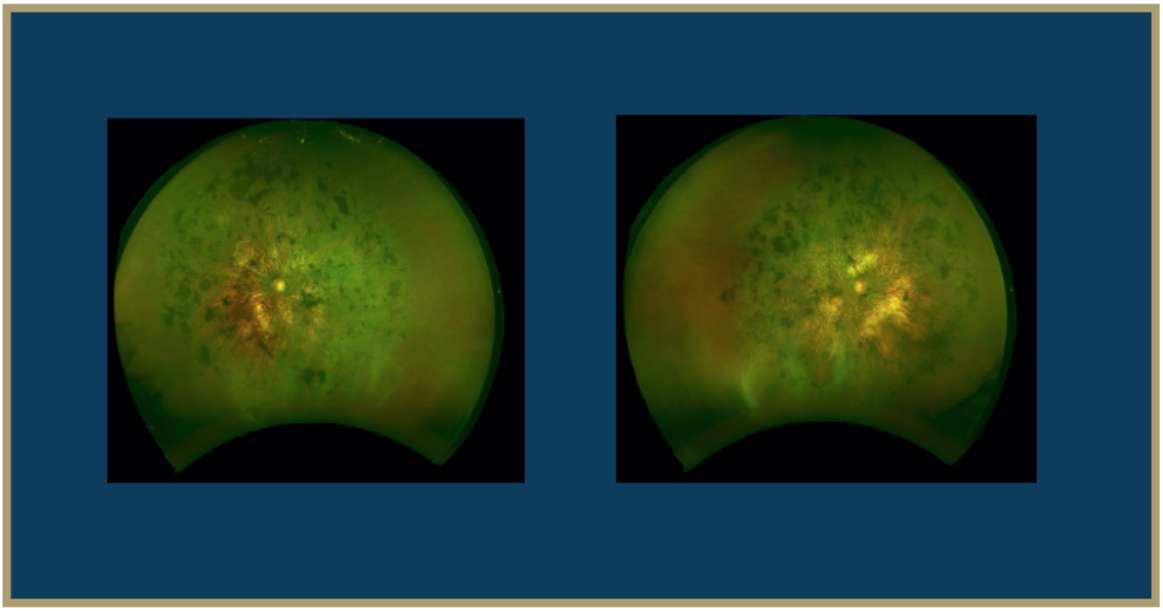

Figure 2: Optos imaging showing progression of the bone spicule-like changes with extension from the macula to the periphery.

Differential Diagnosis

Retinitis pigmentosa
Gyrate atrophy
Choroideremia
Leber congenital amaurosis

Clinical Course

An 84 year old male with a history of central retinal changes presented to the ophthalmology clinic several years ago. Fundus exam has shown progression of the bone-spicule changes from the macula with extension into the periphery, but sparing the far periphery. He was diagnosed with atypical retinitis pigmentosa. His vision continues to be stable at 20/400 OU. He continues to follow up regularly with ophthalmology and receives low vision services.

Discussion

Retinitis Pigmentosa (RP) refers to a group of inherited retinal disorders that results in the degeneration of photoreceptor cells[1]. It is the most common inherited retinal condition, affecting 1 in 4,000 people in the United States. While non-syndromic RP affects 1 in 5,000 people worldwide, RP can also be associated with syndromes such as Usher syndrome and Bardet Biedl syndrome.

The exact mechanism of retinitis pigmentosa has not been completely elucidated as there are multiple genetic mutations with different inheritance patterns that can lead to the same phenotype[3]. However, many of the mutations cause either defects to phototransduction, retinal metabolism or the trafficking of proteins from the inner to the outer segment of the retina[1]. What these mutations have in common, and what is responsible for the phenotype displayed, is that they all contribute to loss of photoreceptor cells over time. Photoreceptor cells, especially rod cells, work on a very delicate balance, and thus are likely to have little tolerance for deviations from their homeostatic norms. The loss of photoreceptor cells is due to chronic stress and inability for these cells to respond to environmental insults. This is the defining trait of retinitis pigmentosa.

RP usually progresses slowly over several decades with symptoms typically occurring bilaterally. The disease commonly presents as night blindness starting in childhood due to rod photoreceptor loss. Later stages of RP involve the loss of peripheral vision, severe photophobia, and the development of dyschromatopsia, or deficiency in color vision, which suggests cone photoreceptor involvement. The timing for this progression is not standard, with the specific mutation and homozygosity often being associated with different clinical courses[1].

RP is a clinical diagnosis that can be diagnosed through funduscopic imaging and assessing the patient's visual acuity, particularly their peripheral vision with a peripheral field test.[5] Electroretinography can be used to assess progression of disease with progression defined as greater involvement of photoreceptor cells with less signal being received over time.[5] Fundus examination of patients with RP may reveal a characteristic triad of bone spicule pigmentation,

retinal vessel narrowing, and optic disc pallor[2]. These findings may not be present in early disease and may vary depending on the severity of disease.[2] Patients with RP can also present with subcapsular cataracts and macular edema.[5] These examination findings in addition to the clinical history are satisfactory for the diagnosis of retinitis pigmentosa.[6] The diagnostic work up also includes screening for skeletal, growth, facial, or neurologic abnormalities to rule out a syndromic cause of the patient's presentation.[6] However in clinical practice, this may be superfluous as it would likely not change management unless it points to the possibility of further systemic sequelae beyond the retina[1].

To this date, no standard treatment exists for patients with RP.[7] Several dietary supplements including Vitamin A and docosahexanoic acid (DHA) have been proposed to reduce the rate of retinal degeneration[4]. DHA is an omega-3 fatty acid that is found in high concentrations in photoreceptor membranes. However, the results of these studies are controversial, and a 2013 Cochrane review concluded that there is no significant benefit of treatment with Vitamin A with or without DHA for slowing the progression of RP.[4]

More recent studies have focused on studying gene and retinal cell replacement therapies for RP. In December 2017, the FDA approved Luxturna as the first gene therapy targeting mutations in the RPE65 gene.[7] The RPE65 gene is, however, only involved in 0.3-1% of RP cases. While no standard treatment exists in patients without the RPE mutation, several additional gene and cell therapies are undergoing clinical trials.[7]

For patients with severe vision impairment and little to no light perception, retinal prosthesis devices such as the Argus II have been developed and approved by the FDA to help restore some functional vision.[4] These devices include a camera that captures visual information from the environment and an implanted electrode array that receives visual information.[4] Stimulation of the remaining retinal cells bypasses the damaged photoreceptor cells and creates a basic distinction of dark and light patterns, thus restoring some capacity of visual function.[4]

Within the spectrum of retinitis pigmentosa, retinitis pigmentosa inversa describes an atypical subtype in which the pattern of vision loss is "inverted" from the typical progression.[10] The condition presents with an initial decrease in central rather than peripheral vision. The loss of peripheral vision, if seen, develops much later on in the disease course.[10] In the case of our patient, the disease appeared to progress in this particular pattern, making retinitis pigmentosa inversa a more accurate description of his condition.

In conclusion, retinitis pigmentosa is a complex group of inherited retinal disorders characterized by progressive vision loss. Due to differences in the progression of subtypes and the lack of significant treatment options, RP remains a challenging condition to manage. As a progressively blinding disease, RP can also have a significant impact on quality of life. All RP patients, as we saw in our case, should receive regular ophthalmology follow up to monitor their individual disease progression as well as referral to low vision services for functional vision preservation. Continued efforts to improve treatment options may pave the way for potential breakthroughs in disease management in the coming years.

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Endophthalmitis

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