CASE REPORT

SECONDARY GLAUCOMA DUE TO MUCOPOLYSACCHARIDOSIS: A CASE REPORT

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ABSTRACT

Introduction: *Mucopolysaccharidosis (MPS) is a rare, autosomal recessive inherited lysosomal storage disorder involved in glycosaminoglycans (GAG) degradation. Glaucoma, as a leading cause of irreversible blindness, in MPS patients has a very low occurrence rate, indicating that this condition needs attention and more research to improve MPS patients' quality of life.*

Case Report: *A 15-year-old boy came with a chief complaint of progressive onset of blurred vision in both eyes 4 years prior to admission. He was being treated as hypermetropia since then. Opacity was noticed in both eyes in addition to deteriorating vision for the last 3 months. Genetic test supported the possibility of MPS type VI. Hence this patient was diagnosed as secondary glaucoma due to MPS. Glaucoma medications were unable to control the intraocular pressure, thus trabeculectomy was planned. After the surgery, the patient was given antibiotic, analgetic, steroid, and anti-glaucoma medications. Releasable suture was removed on the 14th day post-surgery due to elevated IOP. Unfortunately, the patient did not make any visit afterward for further follow-up.*

Discussion: *Our case faces several difficulties with anterior and posterior ophthalmological examinations. Corneal opacity with corneal thickening often occurs in MPS type VI. The exact mechanisms underlying corneal opacities remain elusive. IOP measurements can be helpful for the diagnosis and monitoring of glaucoma. Changes in corneal thickness can affect IOP measurements, and corneal opacity can prevent accurate visualization of the optic nerve and cornea-sclera angle, as well as supportive diagnostic devices such as Funduscopy, Ultrasound Biomicroscopy (UBM), and Optical Coherence Tomography (OCT), are difficult to perform.*

Conclusion: *The underlying syndrome was suspected only after glaucoma occurred. Early detection and regular assessment play an important role in MPS. Routine follow-up is needed to ensure IOP control and determine the long-term outcome of IOP after trabeculectomy.*

Keywords: *Glaucoma, Mucopolysaccharidosis, trabeculectomy*

INTRODUCTION

Glaucoma is a group of progressive optic neuropathies that is defined by alterations in the optical nerve head caused by degeneration of the retinal ganglion cells and retinal nerve fiber layers. It affects over 79.6 million people worldwide and remains the leading cause of irreversible blindness.[1] Glaucoma in children is a rare condition that has the potential to cause blindness and is characterized by ocular structural damage and visual impairment associated with increased intraocular pressure (IOP) and is caused by a variety of conditions. The clinical

picture may vary depending on the age of onset of the disease.[2]

Impaired outflow through the trabecular meshwork is the fundamental pathophysiology of all pediatric glaucomas. The first step in developing diagnostic and screening tools that could identify individuals at risk for the condition before irreparable optic nerve damage occurs is characterizing the underlying genetic abnormalities that cause glaucoma. Additionally, genetic counseling and the risk assessment of subsequent pregnancies depend on it.[3]

For the rare conditions associated with glaucoma in children, determining the mechanism of glaucoma produces the best therapeutic outcomes. Mucopolysaccharidosis (MPS) is a heterogeneous group of multisystem disorders resulting from the accumulation of glycosaminoglycans in ocular and systemic tissues. MPS is inherited in an autosomal recessive manner except for MPS II, which is inherited in an X-linked manner. Frequent visual disturbances can be caused by corneal opacities, optic neuropathy, retinopathy, or cerebral vision disorders.[3,4]

This case report aims to determine the relationship between clinical symptoms, systemic abnormalities, and supporting examinations to be carried out, as well as establish the correct etiology and diagnosis in the patient.

CASE ILLUSTRATION

A 15-year-old boy came with the main complaint of blurry vision in both eyes. Blurred vision in both eyes has occurred for 4 years. The patient has been examined and treated as a case of hypermetropia since then. Vision became progressively blurry and opacity began to appear in the center of the patient's eyes which became more extensive, which led his family to take him to see a doctor to change glasses 3 months ago. Afterward, the patient was referred to the glaucoma department because of high intraocular pressure (IOP) and was given latanoprost, timolol 0.25%, artificial tears, and eye vitamin.

There is no history of infection or other eye problems. The patient has had a history of growth and development delays since the age of 5 years and started wearing glasses since the 5th grade of elementary school using S + 2.25D glasses. History of using other medications was denied. The patient is a child who was adopted at the age of 3 days so the biological family's medical history is unknown. The patient was born spontaneously vaginally, at term with a birth weight of 1900 grams, and immediately cried at birth. The patient's vaccination history is complete. There was no history of allergies.

On physical examination, his general condition was good, with compos mentis, and adequate nutrition (body weight of 21 kg and height of 106 cm). Vital signs are within normal

limits.

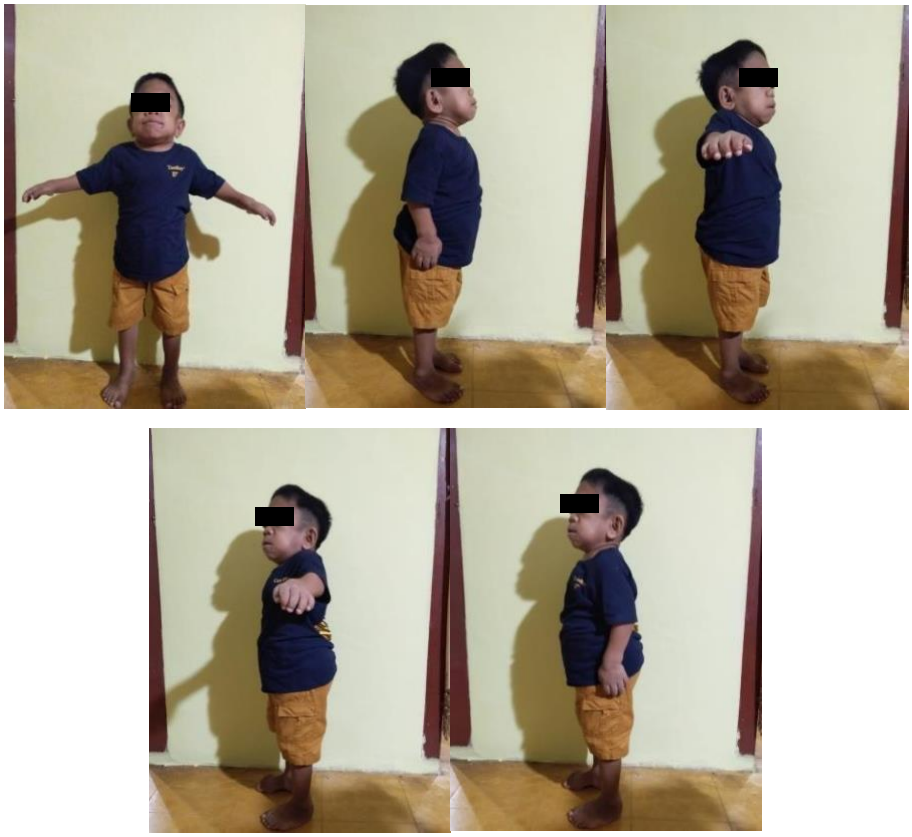

Figure 1. Clinical photo of the patient

On ophthalmological examination of the right eye, visual acuity was 1/60 and intraocular pressure was 20.7 mmHg. Ocular movement was good in all directions. The cornea was cloudy. The anterior chamber depth was Van Herick (VH) I-II. Iris was brown, crypts (+), with minimal iridodonesis. The pupil was round and light reflex (+). The lens was difficult to assess. The palpebra and conjunctiva were both normal. On ophthalmological examination of the left eye, visual acuity was 1/60 and intraocular pressure was 20 mmHg. Ocular movement was good in all directions. The cornea was cloudy. The anterior chamber depth was VH I-II. Iris was brown, crypts (+), with minimal iridodonesis. The pupil was round and light reflex (+). The lens was difficult to assess. The palpebra and conjunctiva were both normal (Figure 2).

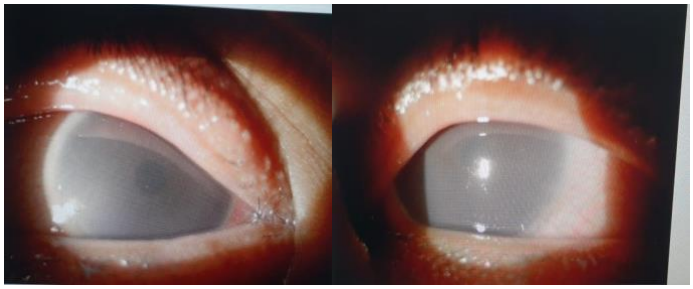

Figure 2. Anterior segment in both eyes

During examination of the posterior segment, fundus reflex, papillary, retina, and macula were all difficult to assess in both eyes (Figure 3).

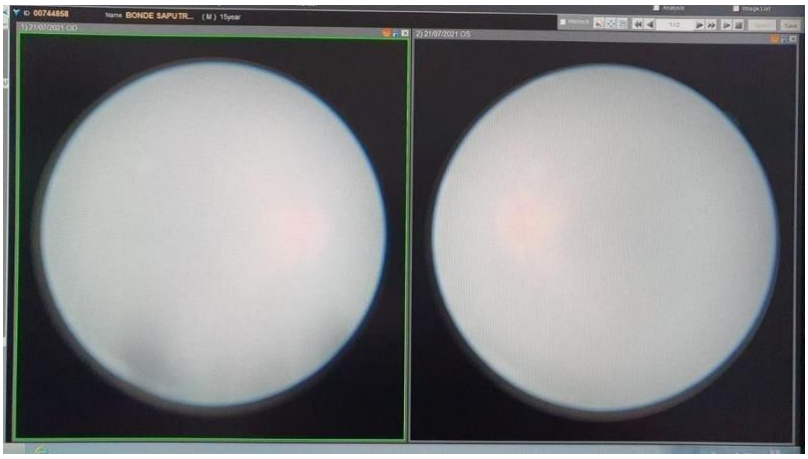

Figure 3. Fundoscopic findings

Gonioscopy was performed and it was difficult to assess in both eyes. Similarly, perimetry, UBM, papillary OCT, macular OCT, and specular microscopy were all difficult to assess. Ocular ultrasound showed echolucent features in vitreous cavity ODS (Figure 4). The biometry result showed that the right eye axial length was 21.87 mm and the left eye 21.61 mm (Figure 5). The CCT was 602 μm and 582 μm in his right and left eye, respectively. The corneal diameter was 12.6 mm in both eyes.

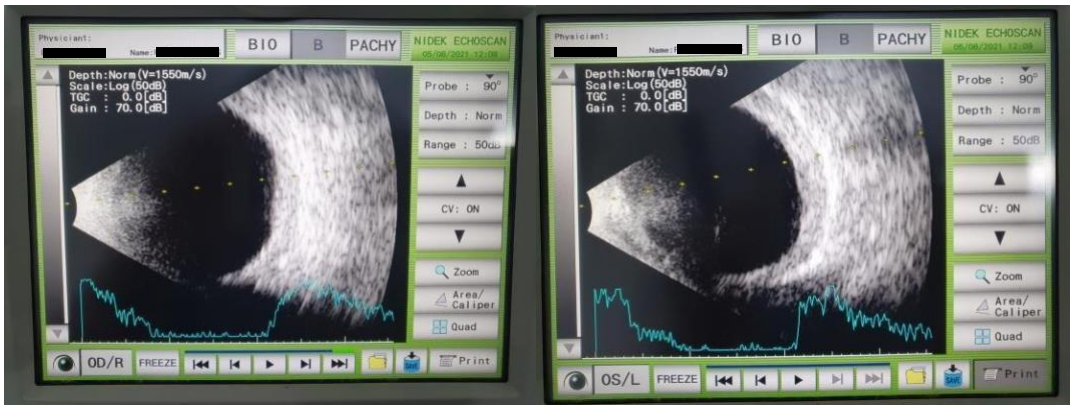

Figure 4. Ocular ultrasound

Figure 5. Biometry result

On laboratory tests, overall was within normal limits. Chest x-ray showed normal cardiac and lung. Echocardiography showed mild mitral regurgitation (MR), tricuspid regurgitation (TR), and pulmonary insufficiency (PI).

A multidisciplinary team was involved in treating our patient, including a pediatrician, otolaryngologist, nutritionist, cardiologist, medical rehabilitation, respirologist, and clinical pathologist. Of all the results, we observed signs and symptoms that may lead to MPS such as growth restriction, facial coarse, dysostosis multiplex, upper airway obstruction, and structural cardiac valvular dysfunction. The genetic test showed the activity of arylsulfatase B decreased at 1.9 μmol/L/H (normal range $\geq$8.8 μmol/L/H) which supports the possibility of MPS type VI. Hence, we concluded that this patient had secondary glaucoma due to MPS VI.

With the medications that have been prescribed before, yet the patient's IOP still had not been under control, instead, he began experiencing complaints such as watery eyes, pain, glare, and headache about 1.5 months ago. Therefore, we decided to perform a trabeculectomy alternately with a releasable suture in both eyes.

On 9th September 2021, the surgery was performed. The patient was laid on the operating table, and then disinfection and subconjunctival anesthesia were performed. Limbus fixation was performed at 12 o'clock. Conjunctival peritomy was performed at approximately 6-8 mm at 12 o'clock. Any bleeding was cauterized. Mitomycin C is placed as a subconjunctival compress for 2-3 minutes. Grooving measuring 4 x 2 mm is made and flaps with crescents on the grooving. The side port was installed at 10 o'clock. A sclerostomy was performed through the COA with a stab knife, then a hole was made with the Kelly Puncher, then an iridectomy was performed with a puncture. The sclera was sutured with a releasable suture using Nylon 10.0. Mattress suture to close the conjunctiva was performed using Vycril 8.0. Intracameral

antibiotic (Levofloxacin) and subconjunctival dexamethasone-gentamycin were administered. Antibiotic ointment was applied and the surgery was completed.

On 1st postoperative day, the patient complained about minimum pain and gritty sensation in the right eye. On ophthalmological examination of the right eye, visual acuity was 1/300, and intraocular pressure was 11.3 mmHg. Ocular movement was good in all directions. Conjunctival injection (+), intact suture, and high bleb were found. The cornea was cloudy with a releasable suture (+). Anterior chamber depth was VH II-III. Iris was brown, crypts (+), with iridodonesis minimal. Pupil was round and light reflex (+). The lens was difficult to assess (Figure 6). On the left eye, visual acuity was 1/60, and intraocular pressure was 23.1 mmHg. Other examinations remain the same.

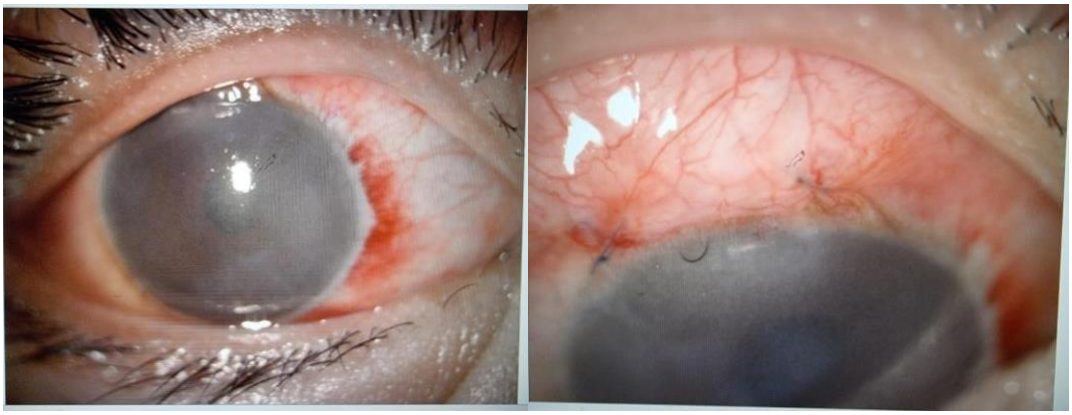

Figure 6. Anterior segment on the right eye day 1 post-trabeculectomy.

The patient was allowed to be discharged and treated as an outpatient, with a scheduled follow-up 1 week post-operative. Post-surgical management includes eye hygiene, amoxicillin 3x250 mg, paracetamol 3x250 mg, methylprednisolone 2x8 mg, prednisone acetate eye drop every 3 hours OD, levofloxacin eye drop every 2 hours OD, timol eye drop 0.25% 2x1 gtt OS, latanoprost eye drop 1x1 gtt OS, and eye vitamin.

On the 7th postoperative day, there was minimum pain on the right eye. On ophthalmological examination of the right eye, visual acuity was 1/300, and intraocular pressure was 17.0 mmHg. Ocular movement was good in all directions. Minimal conjunctival injection (+), intact suture, and high bleb were found. The cornea was cloudy with releasable suture (+). Anterior chamber depth was VH II-III. Iris was brown, crypts (+), with iridodonesis minimal. Pupil was round and light reflex (+). The lens was difficult to assess (Figure 7). On the left eye, visual acuity was 1/60 and intraocular pressure was 21.7 mmHg. Ocular movement was good in all directions. The cornea was cloudy. Anterior chamber depth was VH I-II. Iris was brown, crypts (+), with iridodonesis minimal. Pupil was round and light reflex (+). The lens was difficult to assess. Palpebra and conjunctiva were both normal. The steroid and antibiotic were being tapered off.

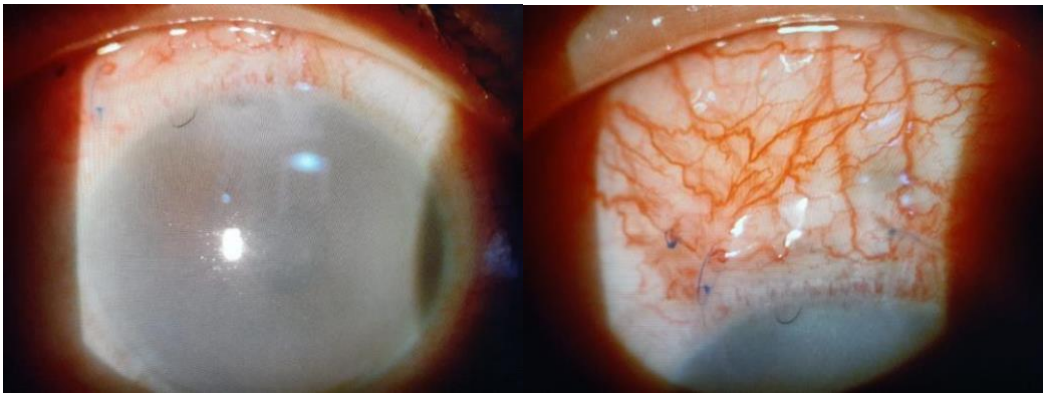

Figure 7. Anterior segment on the right eye on day 7 post-trabeculectomy.

On the 14th postoperative day, the patient complained of watery eye. On ophthalmological examination of the right eye, visual acuity was 1/60, and intraocular pressure was 33.0 mmHg. Ocular movement was good in all directions. Minimal conjunctival injection (+), intact suture, and high bleb were found. The cornea was cloudy with a releasable suture (+). Anterior chamber depth was VH II-III. Iris was brown, crypts (+), with iridodonesis minimal. Pupil was round and light reflex (+). The lens was difficult to assess (Figure 8). On the left eye, visual acuity was 1/60, and intraocular pressure was 24.0 mmHg. Ocular movement was good in all directions. The cornea was cloudy. Anterior chamber depth was VH I-II. Iris was brown, crypts (+), with iridodonesis minimal. Pupil was round and light reflex (+). The lens was difficult to assess. Palpebra and conjunctiva were both normal. We decided to release the releasable suture and did a slight digital pressure on the bleb under topical anesthesia. We then measured his IOP right after and 30 minutes after these procedures. We noticed that there was a reduction in IOP. Moreover, we shifted the steroid to a lower potent group while the timol and latanoprost therapy was continued.

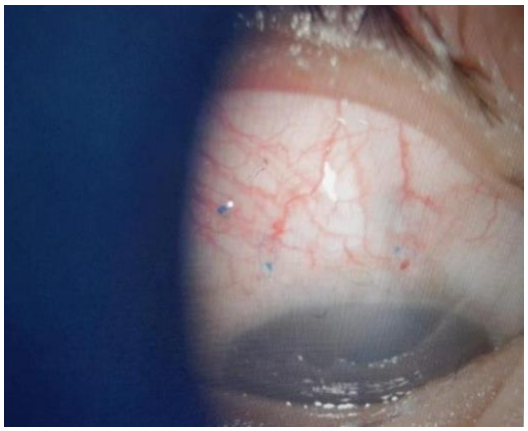

Figure 8. Anterior segment on the right eye on day 12 post-trabeculectomy.

The visual acuity remained the same as before the surgery, but the patient stated that he felt more relief. There was also a deepening of the anterior chamber in his right eye. The next follow-

up was scheduled for every 2 weeks. Unfortunately, the patient did not make any visit afterward for further follow-up.

DISCUSSION

Secondary glaucoma is glaucoma brought on by other ocular abnormalities (acquired or congenital) or linked to systemic illnesses or syndromes. Patients should be referred to a pediatrician for assessment if they have diseases that are known to be linked to systemic abnormalities. For inherited disorders, genetic counseling and family screening are recommended.[3,5]

Similar to adults, uveitis, infection, ocular trauma, and corticosteroids can all lead to secondary glaucoma in children. The age of the child, the extent of the IOP elevation, and the degree of vision loss all influence the presenting signs and symptoms. Patients with Marfan syndrome, homocystinuria, Weill-Marchesani syndrome, and microspherophakia may experience lens-related problems that result in angle-closure glaucoma.[6,7]

A diverse collection of lysosomal storage diseases called mucopolysaccharidosis (MPS) are brought on by the buildup of glycosaminoglycans (GAGs). This substance causes a multi-organ illness with extensive systemic effects that is characterized by a phenotypically diverse condition (Figure 9). In MPS, ophthalmologic abnormalities include retinopathy, optic neuropathy, glaucoma, and corneal opacities. The most prevalent ocular characteristic of MPS, particularly types I, IVA, and VI, is corneal opacification. The use of enzyme replacement therapy (ERT) and hematopoietic stem cell transplantation (HSCT) has significantly raised survival rates in recent years; nonetheless, the best results are obtained when treatment is started during the first 16 months of life. Ophthalmologists are therefore crucial in the accurate diagnosis and treatment of eye disorders by identifying them early on.[8,9]

Disease	Corneal Opacification	Retinopathy	Glaucoma	Optic nerve anomalies
MPS I-H (Hurler)	+	++	+	+
MPS I-HS (Hurler-Scheie)	++	++	++	++
MPS I-S (Scheie)	+++	++	++	++
MPS II (Hunter)	+	++	+	++
MPS III (Sanfilippo)	+	+++	+	+
MPS IV (Morquio)	+	++	+	+
MPS VI (Maroteaux-Lamy)	+++	Unknown	++	++
MPS VII (Sly)	++	Unknown	++	++
MPS IX (Natowicz)	Unknown	Unknown	Unknown	Unknown

MPS—mucopolysaccharidosis; + mild; ++ moderate; +++ severe

Figure 9. Manifestation of MPS type.[10]

Mucopolysaccharidosis I is an autosomal recessive disease caused by α-L-iduronidase deficiency. Heparan sulfate (HS) and dermatan sulfate (DS) build up as a result of an enzyme shortage. Hurler, Hurler-Scheie, and Scheie are the three phenotypic diverse subtypes; Hurler is the most severe variety, while Scheie is the mildest. All three genotypes exhibit ocular symptoms.[8]

The majority of MPS I patients have corneal opacity, which can appear within the first year of life. The opacity, which is frequently characterized as a diffuse ground glass look, is brought on by a disruption of the normal collagen alignment in the corneal stroma, even though GAGs are deposited in all layers of the cornea. GAG deposits thicken the cornea in addition to opacifying the cornea, obstructing vision, and making glaucoma diagnosis and treatment challenging.[8]

IOP readings can be useful in the detection and tracking of glaucoma. IOP measurements may be impacted by changes in corneal thickness, and precise vision of the optic nerve and cornea-sclera angle may be impeded by corneal opacity. According to a cross-sectional study conducted on children with MPS I, IOP readings should be adjusted to prevent needless glaucoma treatments, such as medication or surgery. GAG buildup in the trabecular meshwork can result in both open-angle and closed-angle glaucoma.[8] Refractive errors, particularly hypermetropia, are also common in MPS I because of inelastic reduced scleral axial length and inflexible corneal curvature, both of which are associated with GAG storage. GAG accumulation in the extraocular muscles and decreased corneal opacity are the main causes of anomalies in ocular motility, particularly exotropia. Incidence of retinopathy was discovered in all three MPS I disorders by another investigation. Due to the presence of corneal opacification on top of optic neuropathy and retinopathy, visual abnormalities may go unnoticed.[4,8]

Within the MPS family, mucopolysaccharidosis II (Hunter syndrome) is the only X-linked illness. DS and HS build up when there is a deficiency in iduronate-2 sulfatase, an enzyme that catalyzes the removal of sulfate groups. Hunter syndrome affects between one in 100,000 and one in 170,000 live male newborns. Compared to men, women experience milder clinical symptoms. Many times, phenotypic traits are not understood at birth, but they start to show up between the ages of two and four. Even though ocular symptoms are not the main characteristic of MPS II, posterior chamber abnormalities are more common than anterior chamber problems.[4,8]

In MPS II, corneal opacities are uncommon. The most frequent visual symptoms are exophthalmos and hypertelorism, which can result in long-term problems linked to corneal overexposure. A moderate frequency of retinopathy and anomalies of the optic nerve was also

seen in Hunter's patients. Chronic disc elevation without elevated intracranial pressure is caused by increased pressure on the optic nerve resulting from scleral GAG deposition and thickening, notwithstanding the rarity of glaucoma. The degree of the disease determines the extent of retinopathy; yet, electroretinograms have been helpful in many MPS II individuals exhibiting nyctalopia.[8]

Sanfilippo Syndrome, also known as mucopolysaccharidosis III, is an autosomal recessive condition that has four subtypes: A, B, C, and D. These subtypes are characterized by defects in heparan-N-sulfatase, N-acetylglucosaminidase, α-glucosaminide acetyltransferase, and N-acetylglucosamine-6-sulfatase, respectively. Retinopathy is the most common eye ailment associated with Sanfilippo syndrome. Retinal dysfunction, retinal degeneration, and progressive photoreceptor loss are brought on by the accumulation of heparan sulfate in retinal pigment epithelial cells and the photoreceptor matrix. In clinical settings, patients frequently report impaired vision and nyctalopia. MPS III is typically not linked to corneal opacities.[8]

MPS IVA and MPS IVB are two subtypes of mucopolysaccharidosis IV (Morquio syndrome), an autosomal recessive illness. Refractive errors and widespread corneal opacification are common ophthalmological symptoms. Patients may observe corneal opacity that gets worse with age and can still feel photosensitivity even though the degree of corneal clouding is not as great as it is in MPS I and MPS VI.[8]

Mucopolysaccharidosis VI (Maroteaux-Lamy syndrome) is an autosomal recessive disease. It is known to be caused by a deficiency of N-acetylgalactosamine-4-sulfatase (arylsulfatase B), which leads to the accumulation of DS and chondroitin-4-sulfate (C4S). Corneal opacity with corneal thickening often occurs. Ocular motility issues are another prevalent issue that can result in amblyopia and strabismus. Retinopathy is not linked to MPS VI.[8] Recently, patients with MPS VI may also have a scleral thickening, which is most likely a result of scleral GAG deposits.[11]

Patients typically have a characteristic phenotype early in life, such as respiratory disorders and facial dysmorphias. During growing, additional characteristics such as small stature, heart valve disease, sleep apnea, skeletal degeneration, medullar and peripheral compression, and sensory impairment may manifest. Unlike individuals with other forms of MPS, those with MPS VI typically do not have cognitive impairment. There is a treatment called enzyme replacement therapy (ERT) that can potentially alter the course of the disease if started early.[8,11]

Due to the low frequency and high childhood mortality, the majority of studies on these disorders have concentrated on their fatal symptoms. Glaucoma affects 2.1% to 12.5% of people

with all MPS, with MPS VI having a particularly high frequency of the condition.[11] Nonetheless, these patients' life spans increased and their quality of life improved as a result of the advent of innovative medicines.[12]

Corneal penetrating keratoplasty (PK) is frequently used to treat corneal opacity due to the deposition of GAG.[13] To a limited extent, PK facilitates early vision recovery; nevertheless, most patients experience low visual acuities, which are often related to optic neuropathy and ocular hypertension (OHT).[11]

Optic neuropathy most likely has multiple causes. OHT and glaucoma are prevalent in these individuals. OHT could result from trabecular meshwork infiltration and the inadequate drainage of the aqueous humor as a result. However, because of the elevated corneal thickness brought on by GAG deposition, OHT could also be an artefactual measurement. On the other hand, optic neuropathy and atrophy may also be caused by orbital and retro-orbital causes. The advancement of optic neuropathy may be significantly influenced by the rise in intracranial pressure and the posterior compression of the optic nerve.[8,11]

Figure 10. Corneal clouding in patients with MPS VI.[8]

Mucopolysaccharidosis VII (Sly syndrome) is an autosomal recessive illness caused by β-galactosidase deficiency. Corneal clouding is the most prevalent eye condition, however, it's typically not as bad as MPS I or VI. No abnormalities of the optic nerve were reported. It is still unknown how often retinopathy and glaucoma occur.[8]

With very little information available, mucopolysaccharidosis IX, also known as Natowicz syndrome, is the rarest type of the MPS family. As of right now, no instances of MPS IX have been reported with ocular symptoms. This could also be brought on by the low disease prevalence and the unavailability of data.[8]

Depending on the phenotype, children with MPS have varying prognoses: some may live until their second decade, while others may not make it past 50 or 60 years of age. For patients with severe MPS I, the optimal course of treatment is early (before 2 years of age) hematopoietic stem cell transplantation (HSTC) using compatible bone marrow or cord blood cells. A pediatrician oversees the multidisciplinary management team, which also includes regular evaluations by a pediatric ophthalmologist and input from several other specialties.[9]

Refractive errors are widespread, particularly hypermetropia and astigmatism, however not all patients benefit from standard glasses due to coexisting ocular conditions. Refraction with a retinoscope or autorefractor is strongly advised following cycloplegia with cyclopentolate and phenylephrine. Accommodative fixation targets and stereopsis assessment should be employed in strabismus situations. The symptoms of photophobia may be alleviated using photochromatic lenses.[8]

Assessing the degree of corneal opacification over time and looking for surface alterations or vascularization can be done with slit lamp examination and photos. It might not be possible to apply more exact and objective techniques utilizing Pentacam and iris cameras in every therapeutic situation. At this time, corneal transplantation is the only available treatment for corneal opacities. Patients with MPS have had deep anterior lamella keratoplasty (DALK) and penetrating keratoplasty (PK). Whereas DALK simply replaces the corneal epithelium and stroma, PK is a full-thickness corneal transplant that involves the removal of the corneal endothelium, stroma, and epithelium.[2,8,13]

It is common for patients who have had HSCT to experience dry eye syndrome. Topical lubricants are advised in these cases of keratoconjunctivitis sicca, along with corneal exposure reduction and topical steroids or cyclosporine for more severe cases. Moreover, topical lubricants are advised in cases of pseudoexophthalmos.[8]

Depending on the patient's age and level of intellect, various methods such as finger counting, Humphrey field analyzer, and Goldmann visual field perimetry are employed for visual field evaluation. Methods beyond standard gonioscopy and slit lamp examination are employed to enhance anterior chamber and optic nerve visualization to conduct a comprehensive glaucoma evaluation. In order to help with the diagnosis and treatment of glaucoma, advanced segment optical coherence tomography (OCT) and ultrasound biomicroscopy offer precise pictures of the anatomy behind a potentially clouded cornea. The increased corneal stiffness in MPS may cause a spurious increase in IOP. OCT facilitates the imaging of the optic nerve, photoreceptor layer, retinal nerve fiber layer (RNFL), corneal layers, and retinal thickness. When measuring intraocular pressure (IOP), Goldmann applanation

tonometry and ocular response analyzers may be more accurate because they rely less on corneal characteristics. The microendoscope is another instrument that aids in resolving the issue of clouded corneas during trabecular surgery.[8]

A thorough fundus examination is necessary for posterior chamber disease to detect retinal and optic nerve pathologies. Even though corneal opacification makes it difficult to manually visualize the retina, fundus photography can nonetheless yield better-than-expected photographs. A-scan ultrasonography helps determine axial length, while echography aids in the examination of the vitreous and retina.[8]

The precise processes that cause corneal opacities are still unknown. A well-known notion proposes that type 1 collagen is not properly organized. Diffuse corneal opacification and decreased corneal transparency are caused by a deficiency in the decorin gene. Decorin is a dermatan proteoglycan that controls collagen fibrogenesis. Patients with MPS I and age-matched controls with healthy corneas were compared for variations in corneal collagen expression. Furthermore, MPS I corneas exhibit elevated smooth muscle actin expression, a sign of stromal cell conversion to myofibroblasts. As demonstrated by corneal opacities and injuries, this conversion is linked to increased corneal collagen synthesis. It is yet unknown if collagen organization is impacted or if myofibroblast conversion has a direct impact on GAG deposition. Understanding how GAGs impact corneal transparency has also been made easier by typical age-related changes in the cornea. Compared to younger subjects, elderly subjects had a 30% higher overall GAG. Reduced antioxidant enzymes in aging eyes are thought to be the cause of these age-related changes in GAG, raising the risk of oxidative stress and slowing the healing of corneal injury.[8]

One of the biggest problems in the realm of glaucoma treatment is managing childhood glaucoma. For the majority of glaucomas, medication is the primary line of treatment; however, sustained efficacy is less common in newborns and infants, particularly in PCG (Primary Congenital Glaucoma). In their lifetime, the majority of youngsters with glaucoma will require surgery. Since there is no cure, lifelong monitoring is necessary to guarantee IOP management and identify any consequences. It is advisable to use protective eyewear, particularly for monocular individuals.[3]

Topical application of medication increases the risk of possibly fatal systemic side effects in children. It is best to avoid using beta-blockers in premature or newborn babies, children with asthma, or children with other heart conditions, such as arrhythmias. First-line treatment options for β blockers are timolol 0.1% and timolol maleate 0.25% because of their superior risk profile and effectiveness. While oral acetazolamide is more effective than dorzolamide in

decreasing intraocular pressure (IOP), its use in children is restricted due to severe systemic adverse effects, including altered hyperactive behavior, failure to thrive, and bedwetting. The first medication authorized for usage in children was latanoprost. Brimonidine can cause drowsiness, unconsciousness, and apnea in newborns. It can also penetrate the blood-brain barrier. The literature suggests against using it on children younger than six years old or under 20 kg in weight. Because parasympathomimetic drugs can enhance aqueous outflow and decrease anterior synechiae development, they are helpful in the postoperative therapy of PCG following angle surgery.[2,14]

Surgery is the primary therapeutic option for pediatric glaucoma. The chosen procedure of treatment is mostly determined by the type of glaucoma; additional factors that may be relevant include the age at which the condition first manifests, the degree of optic nerve damage, corneal clarity, coexisting eye diseases, surgical experience, and surgical history.[3]

The success rate following several goniotomies typically ranges from 70% to 90% with medium-term follow-up, indicating that goniotomy is an effective procedure. Trabeculotomy is more commonly used than goniotomy; nonetheless, it is a more intrusive procedure that may result in conjunctival scarring. For trabeculotomy to be successful, Schlemm's canal (SC) localization must be precise.[3]

Failure of the surgical angle is one of the primary indications for Mitomycin C-treated trabeculectomy. When a very low target pressure is needed, the glaucoma is primarily secondary, the presentation is either very early or very late, and the surgeon lacks the competence to conduct angle surgery, this operation is the first choice. Due to their powerful wound-healing response, children are more likely to have a failed outcome from this technically more challenging surgery. Subconjunctival 5-fluorouracil (5-FU) (0.1–2 ml 5-FU 50 mg/ml) and a steroid, like betamethasone, can be temporarily administered next to the bleb if there is noticeable conjunctival inflammation near the drainage site under anesthesia (EUA).[3,14,15,16]

In order to help control long-term IOP, glaucoma drainage devices, or GDDs, are an essential component of the therapeutic arsenal for pediatric glaucoma. Children with aphakic or pseudophakic uveitis, glaucoma following cataract surgery, cataracts that need to be removed right away, and extremely serious conditions that start at birth are among the indications.[3]

Every child with glaucoma within the vulnerable age range has to have their amblyopia evaluated regularly. When the cornea is clear, refraction should be included in routine exams along with the prescription for glasses as needed. For amblyopia, occlusion therapy ought to be attempted on all children who may benefit from improved vision.[3]

In this patient, signs and symptoms were found that were consistent with secondary glaucoma associated with systemic disorders, namely Mucopolysaccharidosis. The level of visual impairment depends on the severity and/or combination of existing eye disorders.[15] Visual disturbances in Mucopolysaccharidoses type VI are common and can be caused by corneal opacities, optic neuropathy, or cerebral visual impairment. The majority of patients with MPS are hypermetropic due to altered corneal refraction and reduced axial length.[17]

Physical examination revealed short stature, short and stubby fingers and toes, and facial coarse features. Visual acuity was reduced in the right and left eyes. The cornea appeared cloudy and the anterior chamber angle was shallow in both eyes. The lens and posterior segment are difficult to evaluate due to the cloudy cornea. The description is consistent with MPS.[14] Corneal opacity is a characteristic of several MPS disorders (MPS I, MPS IV, MPS VI, and MPS VII), and can appear in infancy. A patient with mild corneal opacities may be asymptomatic, but photophobia and decreased vision occur as the opacities worsen.[17] For the majority of patients, corneal opacity can be resolved with PK, improving vision at least right after surgery. Nevertheless, following successful PK, MPS VI patients typically have extremely low visual acuities.[11,18]

Intraocular pressure in this patient may appear within normal limits, but it should be noted that the patient has received previous anti-glaucoma therapy. Corneal clouding affected the measurement of intraocular pressure (IOP) by altering corneal thickness and stiffening the cornea, in addition to obstructing the inspection of the lens and posterior segment (vitreous and retina).[11,12] According to Ashworth et al., MPS type VI has ophthalmologic characteristics and is predisposed to glaucoma in 50% of cases and corneal clouding in 95% of cases.[19]

Gonioscopy, perimetry, OCT, fundoscopy, CCT, and UBM examinations are difficult to perform due to the deposition of GAG in the cornea which leads to poor refractive media. The ultrasound results showed an echolucent image in both eyes. Chest x-ray examination concluded that there were no visible abnormalities in the heart and lungs. On a simple laboratory examination, no significant abnormalities were found. On echocardiography examination, mild MR, mild TR, and mild PI were found. One important characteristic of MPS VI is the increasing deterioration in cardiorespiratory function. All individuals had cardiac involvement to varying degrees of severity; the most pertinent findings were mitral and aortic thickening/dysplasia.[11] Aortic valve disease is seen in 43% of cases, mitral valve disease in 96% of cases, and tricuspid valve disease in 71% of cases, according to Azak and Golda. As a result, cardiac examinations, which should include an electrocardiogram, echocardiogram, and blood pressure measurement to evaluate any abnormalities in cardiac rhythm or conduction, as well as any changes in the

structure or function of the heart, are advised to be performed every one to two years.[19,20] Additionally, patients may exhibit spinal stenosis and hydrocephalus.[21]

The treatment for this patient was trabeculectomy for both eyes. In eyes that have undergone multiple procedures, it is important to make the next surgery definitive. Some of the advantages of trabeculectomy are postoperative IOP titration with removable sutures, lower mean IOP can be achieved compared with GDD, less reliance on drugs for IOP control compared with GDD, fewer postoperative surgical revisions compared with GDD, significantly clear cloudy corneas, and avoids potential corneal surgery.[3] Unfortunately, the patient's visual acuity remains the same as before surgery. Spartalis et al., stated that in addition to helping with the early diagnosis and treatment of posterior segment ophthalmologic disorders, keratoplasty can enhance vision.[18]

The MPS is still a significant medical issue that requires early detection and timely treatment. When patients exhibit significant clinical symptoms (respiratory symptoms, mental retardation, eyes and ears problem), along with an abnormal appearance (short stature, coarse face, short neck, short nasal bridge, wide nose, swollen eyelids, shortened forearm, genu valgum, and coarse face), it is time to suspect MPS. Enzyme assays are required for these people to diagnose themselves.

CONCLUSION

Any suspicion of glaucoma in children should always be treated seriously and immediately to minimize visual impairment. The goal of the initial assessment is to make a diagnosis of glaucoma and determine its type. Ocular management of Mucopolysaccharidosis is challenging given the complexity of the condition in conjunction with the limited available studies. Despite their challenging management, these patients should have their visual acuity maximized to give them the best quality of life possible. Future eye management is expected to prevent eye abnormalities, such as corneal opacities.

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