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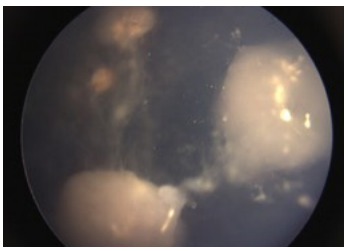
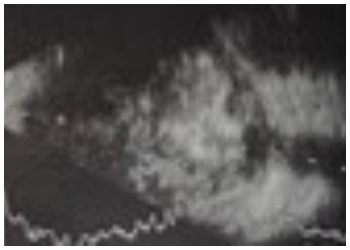
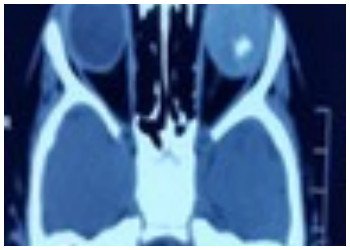
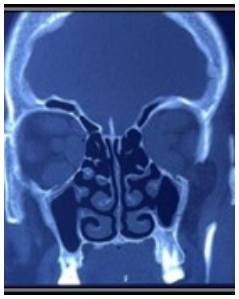
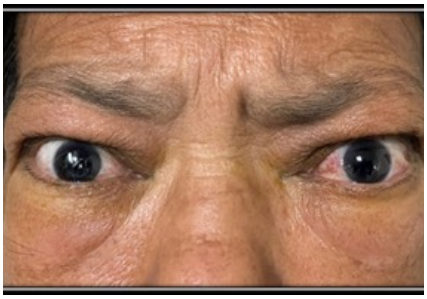
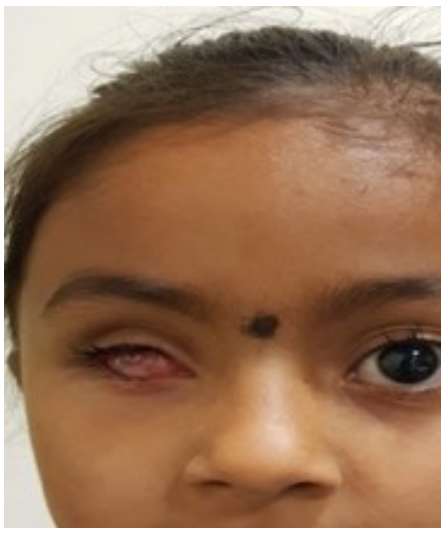
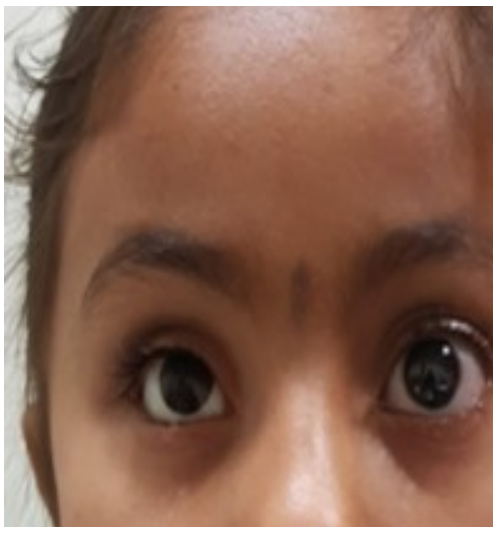
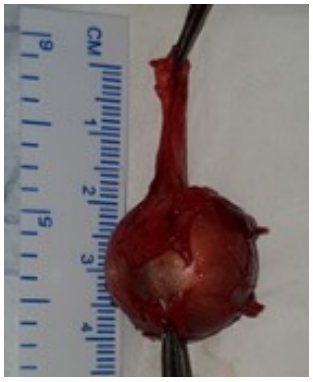
OPSSA NEWSLETTER

OCULOPLASTICS SOCIETY OF SOUTH ASIA

OPSSA

OFFICIAL NEWSLETTER OF
OCULOPLASTIC SOCIETY OF SOUTH ASIA

EDITOR'S NOTE

Dr. Purnima Rajkarnikar Sthapit
Editor-in –Chief, OPSSA Newsletter
Head of Department of Orbit, Oculoplastic, Ocular oncology and Prosthesis
Tilganga Institute of Ophthalmology, Nepal.

Dear colleagues and friends,

It is my great pleasure to present to you this first ever official newsletter of **OPSSA (Oculoplastic Society of South Asia)**! It will be published biannually from now onwards.

Since this is the first issue, I am highlighting on the introduction of OPSSA as an organization and what are its mission and vision. As a general secretary of OPSSA, Prof Golam Haider has highlighted it's milestones along with the pictures in his "Secretary's Note".

There is also a special interview of our founder president of OPSSA, **Prof Ashok Grover**, where he talks about his own journey in the field of oculoplasty as well as why it was necessary to establish a separate South Asian association of Oculoplastic surgeons when we already have national and international organizations serving similar purposes.

Next is a special article by **Prof Rohit Saiju** on his journey of being the pioneer in the field of Oculoplasty and Orbit and establishing Nepalese Society of Oculoplastic Surgeons.

As you are aware that OPSSA has eight member countries. I tried to accommodate a case report from members from different countries. In this issue we have **Dr Syeed** from Bangladesh taking about the management of "Giant Kissing Nevus". We have **Dr Murtaza** from Pakistan describing a "Rare case of Noonan syndrome". **Dr Malita** from Nepal writes about her " Challenge in managing a case of Dysthyroid Optic Neuropathy". Finally we have **Dr Akshay** from India writing about .....

From next issue onwards, we are planning to collect articles based on a single disease entity in each edition. Hence, I request you all to provide articles, case reports or case series on Ptosis for the next issue.

We appreciate your engagement and contribution in OPSSA newsletter and encourage your feedback on this first issue.

Happy reading!

Editorial

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Dr. Purnima Rajkarnikar Sthapit

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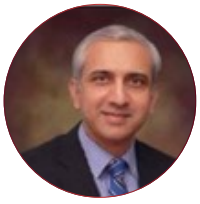

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CONTENTS

INTRODUCTION OF OPSSA

The Purpose

– To bring out the latest & the best in the current practice of Oculoplastics, Orbital, Lacrimal & aesthetics in the periocular area.

– Special focus on the aspects that concern is the most in the South Asian Region.

The name of the society shall be Oculoplastic Society of South Asia, in short form society will be known as OPSSA.

It is non-profitable, non-political, non-government voluntary organization. It aims to develop the academic, medical, scientific, socioeconomic, cultural, and technological capability building of the society and ensuring good governance through advocacy. This Society Shall function considering the state policy and overall public interest and it shall not arrange, support or take part in any such activities which disregard the state rule and create social disorder.

Scope of Operation:

The scope of this society will primarily be all over Bangladesh and others countries in south Asia, with the permission from the competent authority.

Objectives of the Society:

All the objectives mentioned here under will be implemented after obtaining necessary permission from the government concern authority. The objects contrary of the provision of section 20 of the Act 1860 shall be treated as ineffective:

a) To enrich skill, knowledge and attitude of the Oculoplastic Surgeons in this region.

b) To arrange various continuing medical educational programs, seminars, conferences, hands on workshop for Ophthalmologists, doctors, oncologist, ocularist, eye patients and community leaders to bring about an overall improvement in the attitude, knowledge of common people regarding orbital diseases, eyelid diseases, ophthalmic tumors, prevention of blindness, oculo-facial aesthetic issues and their remedies.

c) To establish partnership and close collaboration with other organizations with similar aims and objectives with others national, regional and international organizations/ societies in order to make a concern global effort for the improvement of this branch of ophthalmic discipline.

d) To eliminate blindness due to Ophthalmic Trauma, oculoplastic and ophthalmic oncology related diseases.

e) To develop a carrier planning of the ophthalmologists those who are willing to build their carrier in this subspecialty.

f) To provide guidelines to the hospitals/institutions for the Management of Ophthalmic Trauma, Ophthalmic Tumors, orbit, Socket, Lacrimal apparatus and eyelid related disorders in their service.

g) To establish & maintain an office, library, reading room, recreational & other facilities.

h) To encourage research in this subspecialty by, organizing workshops and establishing grants fellowships, scholarships, prizes, and awards.

i) To enjoy authority or to enter lease, buying, selling constructing transferring and disposing any movable or immovable property to earn resources or capital for development of the society and its interests.

j) To highlights the problems and prospects of profession through media, journals and publications.

k) To open and operate bank accounts and to sign and endorse any per on cheque negotiable instruments, bills of exchange or any accounts for the society.

PESIDENT'S MESSAGE

Dr. (Prof) Ashok Kumar Grover
MD, MNAMS, FRCS (Glas), FICO, FAICO
Awarded Padma Shri by the President of India
Chairman Vision Eye Centres
Chairman, Department of Ophthalmology
Sir Ganga Ram Hospital.

President, Oculoplastics Society of South Asia (OPSSA)
President, Ocular Trauma Society of India
Councillor at Large, Asia Pacific Academy of Ophth (APAO)
Board member, Afro Asian Council of Ophth (AACO)

Oculoplastic Society of South Asia (OPSSA) has been created with the objective of bringing the Oculoplastic surgeons from the region on one platform.

We share so much in common that we could learn a great deal from the experiences of each other.

The society aims to raise the standard of the specialties of Oculoplastic, Orbit, Lacrimal and Facial aesthetics in the region and to bring about greater cohesiveness and cooperation amongst those practicing the specialty in the region. This can be achieved by collaborative efforts in improving education, services and research in the specialty.

The newsletter of the society is a giant step in the direction of achieving our objectives.

I am so glad that Dr Purnima has made this tremendous effort to create a wonderful inaugural newsletter to mark the OPSSA meeting at the NESOS conference.

I express my sincere thanks to Dr Purnima and the NESOS and convey my best wishes.

Look forward to meeting all of you in Kathmandu.

SECRETARY'S NOTE

Prof Dr. Golam Haider
MBBS, FCPS, MS (Fellowship in oculoplasty, India)
Head of department of orbit, oculoplasty and oncology services.
Bangladesh Eye Hospital and Institute
General secretary, Bangladesh Oculoplastic Surgeon Society (BOSS)
Oculoplastic Surgeon Society of South Asia (OPSSA)
Ex. Council Member, (APSOPRS)
Dhaka, Bangladesh
APAO achivement awardee (2017)

This is our great pleasure that OPSSA newsletter is coming to light in 2024 and it is obviously encouraging to highlight OPSSA activities. OPSSA is established as a ad hock committee on 11th April 2018 during the SAO conference held in Kathmandu, Nepal.

Our aims and objectives: To bring out the latest & the best in the current practice of Oculoplastics, Orbital, Lacrimal & Periocular Aesthetics with special focus on the aspects that concern the most in the South Asian Region. We have done several meetings in the last 5 years-

- First meeting: 11th April 2018, SAO conference 2018
- Second meeting: OPAI OPSSA session at Gowahati, Asam, 2019
- Third meeting: 8 th March, 2019 at APAO Thailand
- Fourth meeting on 5 th December, 2021
- Fifth meeting:24 th February 2023 at APAO Kualalampur, Malaysia
- Sixth meeting: 7 th May 2023, at WSOPRAS, Dubai
- Online meeting: 6 th August 2023
- Last meeting: 23 rd February 2024 at APAP Bali, Indonesia

In the last meeting we had decided that membership from respective OPSSA countries to be stepped up by online payment. Lifetime membership fee remain 100USD until October 2024. Thereafter the fees will be raised to 200USD. Physical Biannual OPSSA conference will be held first in Kathmandu, Nepal in October 24-25th 2024 during the NESOS conference. It will be followed by Pakistan in 2026, Bangladesh in 2028 and then in India in 2030. So therefore our request to all the oculoplasty society of native countries to encourage to be a member of OPSSA to increase their OPSSA activities by their academic contribution.

OPSSA also organized three webinars which are as follows-
 11 th July, 2020- "Oculoplastics 2020: The future is here now"
 18 th October, 2020- "Through Generations in Oculoplasty"
 3 rd January, 2021- "Eye cancer in children- a comprehensive approach"

GALLERY

First meeting – OPSSA forming committee at SAO conference in 2018 in Nepal.

Executive meeting of OPSSA at OPAI, Assam in 2019

OPSSA session at OPAI Confrence, Assam 2019

OPSSA Executive meeting during WSOPRAS 2023, at Dubai

OPSSA members with Dr Robert Goldberg and Dr Bita Esmaeli, USA

Executive meeting of OPSSA at APAO 2024 in Bali

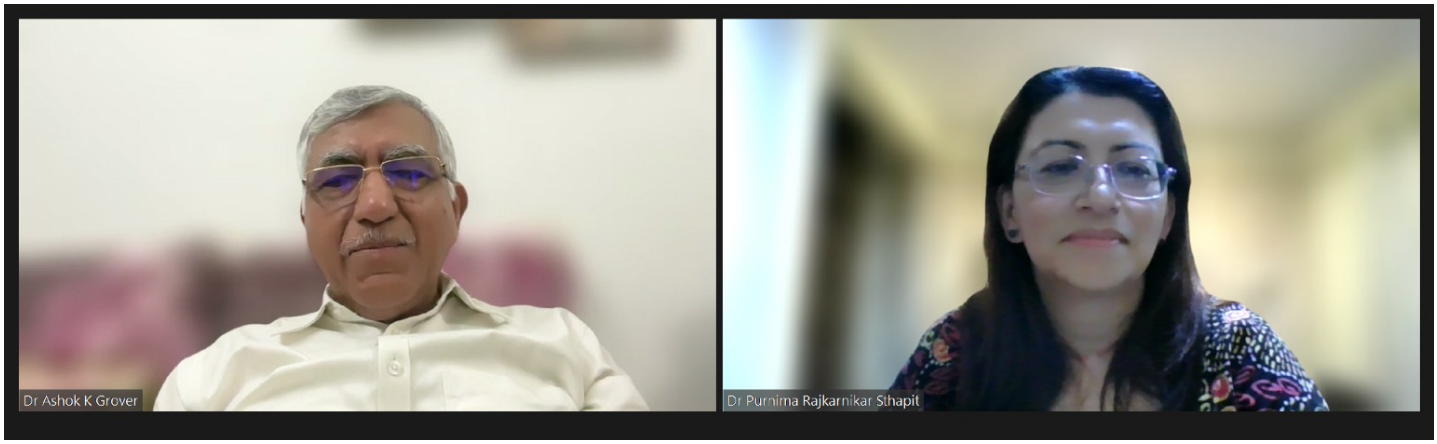
OPSSA members attending WSOPRAS 2023, Dubai

GALLERY

OPSSA Members attending APAO, Bali 2024

Third meeting of Executive meeting of OPSSA at APAO 2019in Bangkok

THE INSPIRING JOURNEY OF DR. ASHOK KUMAR GROVER



What drew you to ophthalmology? Any experiences or individuals that inspired you?

Ophthalmology was an appealing choice for me due to its potential to help the blind. I was inspired by the work done at medical camps and the excellent teaching I received during my undergrad years at Maulana Azad Medical College. One of our senior residents, who had trained at RP Center, was particularly influential in guiding me toward this field. I was fortunate to join the first three-year residency batch at the All India Institute of Medical Sciences, starting right after my internship.

Oculoplastic surgery was not as popular then as cataract or retina. Why did you choose it?

At AIIMS, I worked under Dr. Dayal, a pioneer of oculoplastics in India, during my senior residency. This opportunity, along with my interest in the field, led me to specialize in oculoplastics. When I joined Maulana Azad Medical College as faculty, there was no oculoplastic service, so I initiated it. Over the years, the clinic grew significantly, and it was a rewarding experience starting something from scratch in a major medical college in the capital.

What challenges have you faced in your career?

Starting out, there were no seniors to guide us, no videos or advanced equipment. We had to improvise—using silicone blocks as implants and repurposing needles for procedures. The journey involved reading surgeries from books repeatedly and figuring out techniques ourselves. Documenting work, making videos, and editing them was also a struggle, but it was a fulfilling learning process.

Do you still have those early VHS recordings?

Some of them, yes, but the quality is outdated now. I converted a few, like my initial transnasal wiring and DCR, but they've since been replaced with better versions.

What guiding principles have shaped your approach to patient care?

Hard work and dedication are key. You can't achieve anything without fully committing to it. I also believe strongly in empathy—connecting with patients is vital for their well-being. Being a good teacher is important too; teaching helps you organize your thoughts and learn better.

How do you view the newer generations of ophthalmologists?

Young people today are intelligent and have a better work-life balance than we did. They plan more efficiently, using modern resources to learn faster. The world is evolving, and so are they. We have much to learn from them, especially in balancing life and work.

In the same note, like some ophthalmologists, do you still do cataract surgeries, not cataract and refractive?

Yes, I still do cataract and glaucoma surgeries, though I've stepped back from refractive surgeries as younger professionals take over. While oculoplastics is my passion, cataract surgery has always been important for financial sustainability.

What inspired the formation of organizations like OPSA and WSOPRS?

The idea to form organizations like OPSA (Oculoplastic Association of South Asia) and WSOPRS (World Society of Oculoplastic and Reconstructive Surgery) came from the shared challenges faced by countries in the region, such as heavy patient loads, limited resources, and a shortage of specialized training opportunities. We realized that by uniting under a common platform, we could exchange knowledge, improve training, and elevate the standards of care in oculoplastics.

When we started the Oculoplastic Association of India (OPAI) in 1989, we were just a small group of around 25-30 members. Today, the specialty has grown tremendously, with nearly 1000 members and many medical centers now offering oculoplastic services. This growth encouraged us to expand our vision globally, which led to the formation of WSOPRS. While we've faced obstacles, like political and bureaucratic barriers to academic exchanges, we remain committed to overcoming these challenges to provide the best possible care and training across borders.

The goal has always been to build a community where collaboration and shared learning benefit everyone, especially regions with fewer resources. Despite the hurdles, it's been an incredibly rewarding journey, and the growth of these organizations shows just how important collective effort is in advancing our field.

MY EXPERIENCE OF ESTABLISHING NEPALESE SOCIETY FOR OCULOPLASTIC SURGEONS - NESOS: CHALLENGES AND OPPORTUNITIES

Prof Dr Rohit Saiju, MD
Visiting faculty- Tilganga Institute of Ophthalmology, Kathmandu
Managing Director- Drishti the Vision, Jamal, Kathmandu
Head of Oculoplasty and Orbit- Kathmandu Eye Care Center, Kathmandu

In the area of ophthalmology, the specialization of oculoplastic surgery encompasses a wide range of procedures, from correcting congenital defects to repairing traumatic injuries and addressing age-related changes in the eyelids. These interventions are not merely cosmetic but are essential for maintaining vision, preventing complications, and improving quality of life. In developing countries like Nepal, where healthcare infrastructure is often limited, the need for specialized eye care is profound and establishing an Oculoplastic Society can be a transformative step toward improving eye health outcomes.

Developing countries face a high prevalence of ocular disorders due to various factors, including limited access to healthcare, inadequate education about eye health, and a high incidence of trauma. In Nepal, conditions such as eyelid malpositions, orbital tumors, and lacrimal system disorders are prevalent but often untreated due to a lack of specialized services. An Oculoplastic Society would address these needs by providing a platform for specialists to collaborate, share knowledge, and advance treatment protocols.

I believed that the establishment of a Society would facilitate the training of local ophthalmologists in this special subspecialty. Currently, there is still a shortage of trained professionals in this field in many parts of Nepal. Through workshops, seminars, and hands-on training sessions organized by the society, local practitioners would gain the skills necessary to perform complex procedures, thereby reducing the need for patients to travel to big cities and abroad for treatment. The society would play a critical role in raising awareness about oculoplastic conditions and the importance of timely intervention. By advocating for better eye care policies and funding, society can help secure resources for underserved communities. Public education campaigns can reduce the stigma associated with certain conditions and encourage individuals to seek medical help before conditions worsen like Retinoblastoma. With all these facts and after attending the OPAI 2004 in Assam India and APSOPRS 2008 in Seoul Korea we had initiated a thought to make a similar organization in the country. In the 2014 NOSCON in Pokhara we, a handful of oculoplastic surgeons discussed this matter and made a preliminary draft to form NESOSS officially. I was very much thankful to my colleagues who supported me in executing the thought ahead.

Advocacy and Awareness: Experiences on Challenges-
Establishing this Society in Nepal involves several practical and logistical challenges, yet the experiences from similar initiatives in other regions offer valuable insights. One of the first steps in forming a society was to build a network of professionals committed to oculoplastic surgery. This involves reaching out to ophthalmologists, surgeons, and medical institutions interested in the specialty. Securing funding and resources for the society's activities was a significant challenge. Financial support is needed for setting up the organization, organizing training programs, and conducting conferences. This was addressed by seeking grants from international health

organizations, collaborating with non-governmental organizations, and building partnerships with medical equipment manufacturers and pharmaceuticals, where INGOs like Fred Hollows Foundation, Himalayan Cataract Project, and Tilganga Institute of Ophthalmology supported its activities from the beginning of its days. Developing countries across Asia, Africa, and Latin America face similar challenges in eye care, making the establishment of such societies a valuable model. By setting up specialized societies, developing countries can improve health outcomes related to ocular disorders. This can lead to a reduction in the burden of untreated conditions, lower rates of complications, and enhanced overall well-being for patients. We believe that specialized societies can promote equity in healthcare by ensuring that advanced treatments are not limited to urban centers but are accessible in rural and underserved areas and bring up global collaboration, allowing for the sharing of knowledge and resources across borders.

The establishment of an Oculoplastic Society in Nepal is a critical step towards addressing the significant gap in specialized eye care in developing countries. By focusing on academic - training, and collaboration, we believe such a society can transform the landscape of eye health, improving outcomes for countless individuals. The experiences and challenges encountered in setting up such a society provide valuable lessons for similar initiatives globally. As the world moves towards greater healthcare equity, the establishment of specialized The establishment of an Oculoplastic Society in Nepal is a critical step towards addressing the significant gap in specialized eye care in developing countries. By focusing on academic - training, and collaboration, we believe such a society can transform the landscape of eye health, improving outcomes for countless individuals. The experiences and challenges encountered in setting up such a society provide valuable lessons for similar initiatives globally. As the world moves towards greater healthcare equity, the establishment of specialized societies like those for oculoplastic surgery will play a pivotal role in ensuring that advanced medical care reaches all corners of the country and region.

My experience in founding such an organization was both challenging and rewarding, and it involved several critical steps that contributed to its success. The journey began with identifying a clear need within the professional community. I observed a lack of cohesive representation and support for professionals in our field, which hindered both personal development and industry advancement. Recognizing this gap, I envisioned a society that could offer a platform for networking, professional growth, and advocacy.

The first step was to conduct thorough research. I gathered information about My experience in founding such an organization was both challenging and rewarding existing societies in similar fields, both locally and internationally. This research helped me understand their structures, benefits, and challenges. I also reached out to key stakeholders—industry leaders, potential members, and experts—to gauge interest and gather insights. Their feedback was invaluable in shaping the society's mission and objectives. With a solid understanding of the landscape, I assembled a core team of committed individuals who shared the vision of creating a professional society. This team played a crucial role in developing the society's foundational elements, including its mission statement, goals, and organizational structure. We crafted a clear mission that emphasized professional development, industry advocacy, and community engagement, for this purpose, my colleagues Dr. Basanta Sharma and Dr. Naresh Joshi supported in building and strengthening the society from the beginning days.

Next, we focused on the legal and administrative aspects. We navigated the legal requirements for establishing a nonprofit organization, which involved drafting bylaws, registering the society, and obtaining necessary licenses. This process was complex, requiring careful attention to detail and adherence to regulations. We also sought legal counsel to ensure compliance and to address any potential issues.

Building a membership base was another critical step. We launched a targeted outreach campaign to professionals in our field, highlighting the benefits of joining the society. This included organizing informational webinars and attending conferences. Our efforts paid off, as

we were able to attract a potential group of members who were enthusiastic about contributing to the society's growth.

To provide value to our members, we developed a range of programs and services. These included professional networking events and national conferences. From 2010 to 2017 we organized three national conferences with one nationwide symposium. The beauty of the subject and the attractive skill of delivering lectures by renowned experts at these conferences have attracted many fresh graduates to pursue this as a future career. One of the most rewarding aspects of this experience was witnessing the positive impact of society on its members and the broader industry. The society became a hub for knowledge exchange, innovation, and collaboration. Members reported increased career opportunities, enhanced professional skills, and a stronger sense of community.

In conclusion, while the Oculoplastic Society in Nepal, NESOS faces several challenges, including resource disparity, limited training, and economic constraints, there are substantial opportunities for growth and advancement. By addressing the challenges through targeted strategies and leveraging emerging opportunities, the field of NESOS activities in Nepal can continue to evolve, improving patient outcomes and expanding access to this specialized care.

NESOS Founding Executive team 2014 2016

GIANT KISSING NAEVUS: AN OCULOPLASTIC CHALLENGE

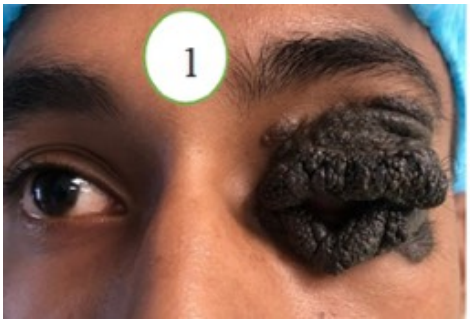
Syeed Mehbub Ul Kadir, Md. Golam Haider
1. Assistant Professor and Head, Oculoplastic Services, Sheikh Fazilatunnesa Mujib Eye Hospital & Training Institute, Bangladesh
2. Professor and Director-Medical Education, Bangladesh Eye Hospital and Institute, Bangladesh

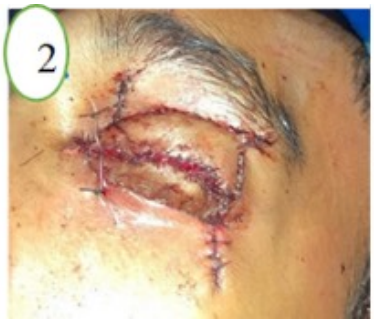
Abstract:
A kissing naevus is usually congenital and found in approximately 1–3% of neonates. These lesions are present at birth and gradually affect both upper and lower eyelids and extend to the eyelid margin. There is a risk of developing malignant melanoma, which ranges from 5–40%. The larger the size of the naevus, the higher the chance of developing malignant change. We present a case of a congenital melanocytic giant kissing nevus involving the entire upper and lower eyelids in a fourteen-year-old male boy who was surgically correcting the lesion. Surgical reconstruction after excision is challenging and multiple sessions are required to get the optimum result.

Introduction:
A kissing nevus is a compound variety of congenital pigmented naevus that affects both the upper and lower eyelids equally and involves the eyelid margins. Congenital naevus is a rare melanocytic lesion and has a risk of developing malignant melanoma that is proportional to the size of the lesion, specifically giant congenital naevus, if it involves over 5% of the body surface or the lesion size is more than 20 cm in adolescents. Here, we attempt a case report on a giant melanocytic naevus of the eyelids which is challenging to manage in surgical aspect.

Case Report:
A 14-year-old male boy presented with a diffuse, large painless enlarging pigmented lesion, involving both upper and lower eyelids of the left eye [Fig. 1] which is starting from his birth which is increasing in size over the time. The upper lid lesion was about (Wide *height) 3 cm × 2 cm, which extends horizontally from lateral to medial canthus, and vertically up to eyebrow in lateral aspect. The upper eyelid crease could not be demarcated. The skin over the lesion was, coarse, multi lobulated and pigmented with fine hair present over the lesion, large hairs are present in few areas. The lower eyelid mass was about 3 cm × 2 cm which was dark black colored pigmentation and hair bearing. The visual axis was covered due the lesion which caused the mechanical ptosis of the left upper eyelid. The other variety of the ptosis could not be evaluated due to giant melanocytic lesion. The best corrected visual acuity (VA) in the right eye was 6/6 and left eye was counting fingers due to stimulus deprived amblyopia. Anterior segment and fundus of the right eye was normal. Slit lamp evaluation and fundus examination of left eye could not be performed preoperatively due to diffuse giant variety of melanocytic lesion. CT scan of the eye and adnexa revealed a heterogenous mass in the upper and lower eyelid in the left eye with extension to the median canthus and lateral canthus but not extending the lesion to the left orbit. The complete excision of the entire full thickness lesions involving the anterior lamella of the both eyelids from the medial to lateral canthus was performed, and the excised tissue was sent for histopathological analysis, which revealed a compound melanocytic nevus involving both

lids. The anterior lamella of each eyelid was reconstructed by the full thickness skin graft [Fig. 2] which was harvested from antero-medial aspect of the right arm. We used 6–0 Vicryl suture throughout the reconstructive surgery and 4-0 silk suture was used for tarsorrhaphy. The tarsorrhaphy was released after 6 weeks of the primary surgery. The surgery removed the obstruction of the visual axis and improved the aesthetic appearance of the patient [Fig. 3]. Eyelash reconstruction is still challenging for this patient and may need more session of the reconstructive surgery.





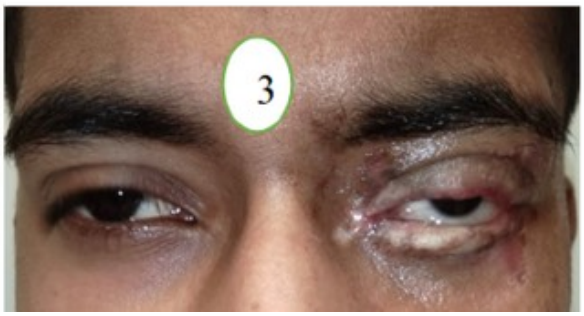


Figure 1: The diffuse, giant, melanocytic, pigmented naevus involving the upper and lower eyelids, including the eyelid margin.
Figure 2: status of the immediate postoperative image of the reconstruction of the anterior lamella of both the upper and lower eyelids with full-thickness skin graft; the lacrimal drainage system is demarcated and fixated with silicone rod separately.
Figure 3: The patient's eyelid condition status, with good aesthetic and functional outcomes after releasing the tarsorrhaphy. Still, ptosis is present, but it may be improved over time.

Discussion:
Congenital melanocytic naevus usually appears at birth and is reported in about 1% of all newly born babies. Based on their architecture, benign melanocytic nevi are divided into three categories: junctional, compound, and intradermal.
Eyelid nevi can be flat, elevated, dome-shaped, or pedunculated. Flat lesions are often junctional nevi, dome-shaped lesions are often intradermal or compound nevi, and pedunculated lesions are usually intradermal nevi. Nevi are typically tan with deep brown pigmentation and are well-circumscribed, not associated with ulceration.

Fuchs A described the eyelids' congenital divided or kissing nevus in 1919. Collenza D published a case report on two patients with kissing nevi in 1937. Subsequently, a study by Callahan A, another study by Harrison A, and Okun M all are reported additional cases in various literature. In 1969, Ehlers N reported a case series on 10 cases of the melanocytic kissing nevus.

Our study patient, a 14-year-old male boy, presented with a diffuse, large, painless, enlarging pigmented kissing naevus involving both upper and lower eyelids, including the left eye's lid margin.

The risk of small melanocytic nevi to transforming malignant condition is still not clear, but the larger lesions of more than 4 cm have a chance of 4.6% for malignant transformation over a long time.

Fuchs A managed two patients through simple excision; two required full-thickness skin grafts to repair the defect, two were treated with cryotherapy, and the remainder received no treatment.

It is always better to excision the lesion as early as possible because a large lesion requires more extensive excision and a difficult reconstructive procedure.12If the melanocytic naevus involving the subcutaneous tissue and deep dermis, the treatment consists of full-thickness excision followed by repair with a skin graft.

Jacob SM et al. described the management of congenital melanocytic nevus or panda naevus or kissing naevus of the eyelid of a 25-year-old female patient who underwent surgical excision with a full-thickness skin graft for the residual defect.14 In our study, we found that the traditional approach to treating a large kissing nevus on both the upper and lower eyelids involves completely removing the lesion in a vertical and horizontal direction until reaching the normal tissue depth. To reconstruct the front layer of the eyelids, a full-thickness skin graft is used. Additionally, we repair the levator muscle and adjust the upper eyelid crease during the surgery.

Current treatment options for Giant congenital naevus include surgical resection of the lesion, but there is no effective medical management for this type of lesion. Trametinib was recently used for a case of a school-going child with a giant congenital melanocytic naevus who was shown an AKAP9-BRAF fusion, which resulted in a good outcome as well as a dramatic improvement in the extent of the melanocytic naevus.

Conclusion:

Complete surgical excision is the main modality for managing the congenital compound melanocytic kissing naevus. Multiple session of the reconstructive surgery is needed to get better functional and cosmetic results.

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OCULAR FEATURES OF A RARE CASE OF NOONAN SYNDROME IN PAKISTANI POPULATION

Dr Murtaza Sameen
Associate consultant
Orbit and Oculoplastics
Al Shifa Trust Eye Hospital,
Rawalpindi, Pakistan

Abstract

A heterogenous congenital disorder, Noonan syndrome(NS), presents with typical features like triangular face, short stature and cardiac defects. It typically presents as an autosomal dominant trait. Noonan syndrome is one of the RASopathies due to involvement of RAS-MAP-Kinase pathway. Diagnosis is based on clinical features that include, typical facial features (triangular face, hypertelorism, ptosis), skeletal abnormalities(scoliosis), short stature, mild developmental delay, presence of cardiac defects, lymphatic dysplasia and a family history of NS. Here we report a case of 12 years old boy with bilateral upper eyelid ptosis. On detailed examination, it turned out to be Noonan Syndrome.

Introduction

Noonan Syndrome is a congenital disorder with prevelance of 1:1000 to 1:2500. Mostly occurs as an autosomal dominant triat. NS is familial in less than 50% of cases. In 2001, the first gene to be connected with Noonan syndrome is PTPN11, while 20 other genes have been discovered, related to this heterogenous clinical condition. Out of 3 RASopathies, Noonan syndrome is considered to be among one of them.

Short stature is one of the main feature of this syndrome. The syndrome includes number of features : Dysmorphic facial features, heart defect, short stature, chest deformity, developmental delay, cryptochidism, delayed puberty, ptosis, hyertelorism, hand contractures, hearing problem.
The aim of discussing this case is to inform and acquaint ophthalmological community and health care professionals about the signs and symptoms of this rare syndrome.

Case Report

A 12 years old boy presented to Armed Forces Institute of Ophthalmology with drooping of both upper eyelids since birth.It was noticed by parents due to his abnormal head posture. There was no history of trauma, redness, photophobia, ocular allergies or ocular medicines. He has 1 sister of 4 years of age, who also have same symptoms since birth. While personal history and socioeconomic history was non-contributory.
On general physical examination, a short stature boy with lean built and contracted fingers and vertebral problems was standing comfortably and was well oriented with time, place and person. Visual acuity was 6/6 OU. An increased intercanthal distance was noticed between two eyes along with drooping of both upper eyelids (hypertelorism and ptosis) and poor levator function (3mm) while rest of anterior and posterior segment examination was within normal limits. He also had triangular face with low set ears and small jaw along with vertebral abnormality like scoliosis. Bilateral hand contractures (Clinodactyly, Brachydactyly and Blunt fingers) were also present in our patient(Fig 1 A-F, Fig 2). No cardiomyopathy like ASD (atrial septal defect) was noticed in this subject. He was suffering from deafness, for which he was referred to E.N.T specialist.

Bilateral Upper eyelid ptosis was corrected with frontalis sling procedure under GA, to prevent amblyopia and correct his head posture (Fig 3).

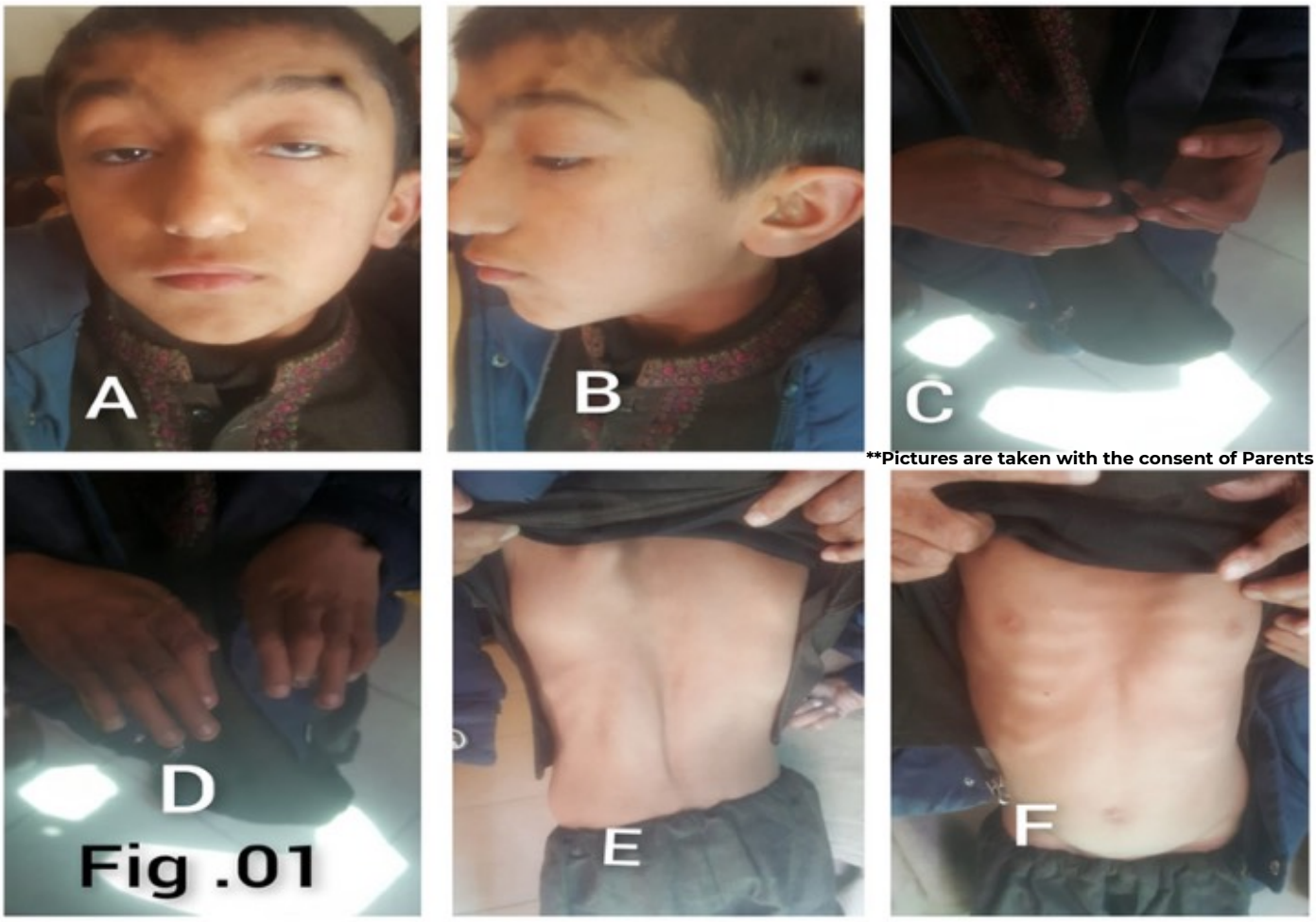

Fig: 1 A) Bilateral Ptosis with hypertelorism
B)Low set ears
C & D)Hand Contractures (Clinodactyly, Brachydactyly and Blunt fingures)
E) Scoliosis
F) Pectus Excavatum

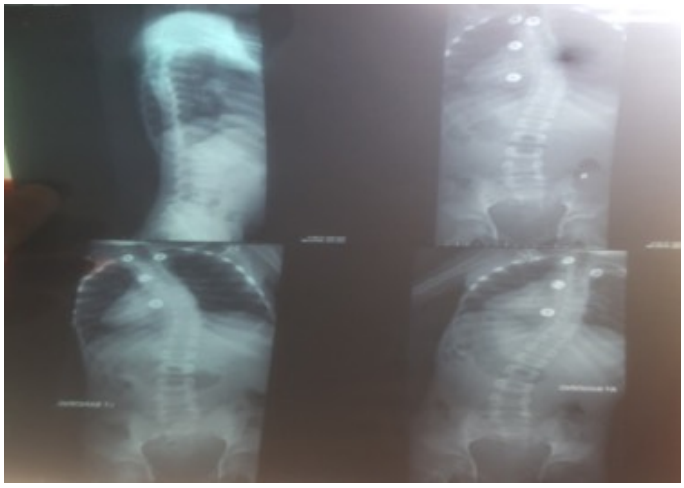

Fig 2: Xray Chest showing Scoliosis

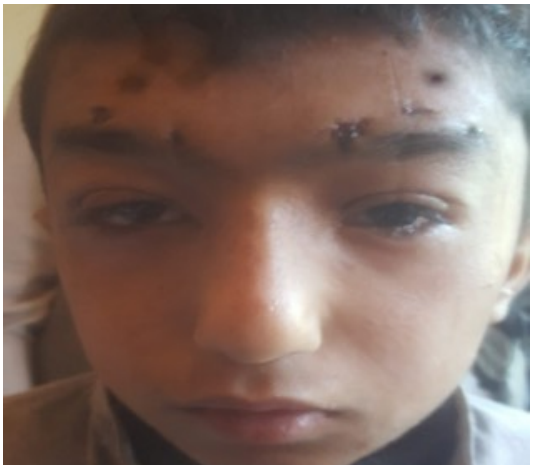

Fig 3: After bilateral Frontalis Sling Procedure

Discussion

In 1963, Noonan described many features that were also common in Lentigines syndrome, so the name Noonan was labeled. Same pleotropic gene has been observed in both syndromes (Noonan and Lentigines).

It has been observed that a patient with Noonan syndrome requires multidisciplinary team approach to treat and manage this rare syndrome. We also sent our patient to cardiologist, dermatologist, endocrinologist, E.N.T specialist, orthopaedic surgeon and paediatrician.

Patient was operated for bilateral ptosis correction with frontalis sling procedure under GA in our case report to make his chin up posture in more comfortable posture and prevent him from amblyopia.

Mendez and Optiz in their study confirm that ocular manifestations are the commonest and consistent features almost 95%, occurring in Noonan Syndrome.

Marin et al, in their study also suggest that ocular features accounts for larger clinical features in Noonan syndrome patients.

The patient we reported here was a young male with ocular and systemic features of Noonan Syndrome.

In summary, NS is a rare disorder with multiple ocular features that should be diagnosed and treated early to prevent vision threatening complications, therefore long term follow up and multidisciplinary team approach is required.

Increased awareness of Noonan syndrome among ophthalmologists and other health care professionals could help parents to seek specialist advise and proper management.

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THE CHALLENGE – DYSTHYROID OPTIC NEUROPATHY

Dr. Malita Amatya
Consultant Oculoplastic Surgeon
Tilganga Institute of Ophthalmology
Kathmandu, Nepal

Introduction

Dysthyroid optic neuropathy (DON) is an optic nerve dysfunction and one of the severe complications of thyroid eye disease (TED). Predisposing causes includes compressive, ischemic, optic nerve stretch neuropathy or mixed etiology, Untreated and if long standing it may result in permanent visual loss. (1,2, 3, 4) Because of its potential reversibility with prompt intervention medically, surgically or in combination, accurate and prompt diagnosis and appropriate management are important.

Several mechanisms have been proposed for the pathogenesis of DON. The most accepted mechanism is compression of optic nerve (CON) by enlarged EOMs at the orbital apex which disturbs axoplasmic flow.(5) The second is from optic nerve stretch from severe proptosis with disruption of the axonal function and blood flow of the optic nerve. This occurs much less frequently than the compressive mechanism.(6, 7) The third mechanism is inflammation which causes optic neuritis.

Intravenous corticosteroids (CS) are the first therapy in active TED and for cases of progressive compressive DON. (1) Intravenous methylprednisolone (IVMP) pulse therapy has greater efficacy and less adverse events compare to oral prednisolone. EUGOGO suggested IVMP as the first line treatment in DON. (8) A variety of immunosuppressive and immunomodulating agents have been studied in patients with active TED and DON. Early orbital decompression should be considered if insufficient or no improvement in optic nerve function is achieved within a few weeks following medical treatment. Complete visual recovery may occur in about 40% of patients with DON after treatment but far more so with surgical decompression. (8)

Financial Interest: Nil
Conflict of Interest : Nil

Case

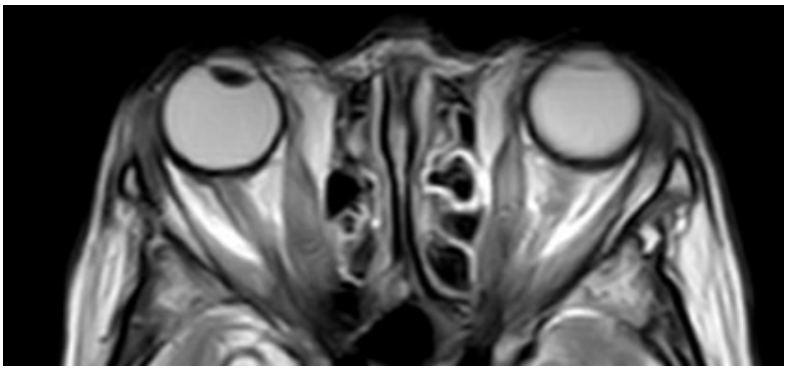
•A 63 years female presented to Oculoplastic clinic with complain of periorbital puffiness both eyes since 3 months duration, gradually increasing. It is
associated with both eyes redness and dull aching pain. She is known case of Grave's disease under medication (Tablet Carbimazole 5- 10 mg TDS) since 10 years. She is non smoker, non diabetic and non hypertensive. On examination her BCVA OD was 6/12 , OS 6/9. Her Pupil was round, regular and reacting to both direct and consensual light reflex. There was immature cataract on OD and pseudophakia on OS.

Initial clinical Activity Score = 5/7Spontaneous retrobulbar pain
1. Spontaneous retrobulbar pain - presentPain on upward or downward gaze
2. Pain on upward or downward gaze - absentRedness of eyelids
3. Redness of eyelids -absentwelling of eyelids
4. Swelling of eyelids -presentConjunctival congestion
5. Conjunctival congestion -presentConjunctival chemosis
6. Conjunctival chemosis -presentSwelling of Caruncle/ Plica
7. Swelling of Caruncle/ Plica –present

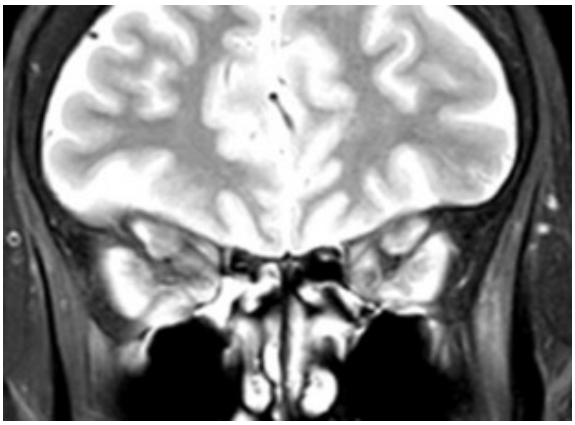
Her hertel exophthalmometry was 16mm on OU with base 95mm. IOP on Applanation tonometry was 30 mm Hg on primary gaze and 32 on upgaze OU. Diplopia was present more on OD maximum on upgaze. Her posterior segment examination including optic disc was normal. Her color vision was normal on OU.
Baseline investigation including blood pressure, Complete Blood count, Renal function tests and Serum electrolytes, ECG, Chest X-ray were sent and the reports were normal. Thyroid function tests were deranged.
She was diagnosed as Active Moderate to Severe Thyroid Eye Disease so she was given Eye drop Bimatoprost 0.01% HS, Eye drop Carboxymethyl cellulose 0.5% QID, Endocrinologist consultation was sent for IVMP pulse therapy regimen - IVMP 500 mg on 3 consecutive days as a loading dose, followed by 500 mg every 3 weeks x 5 cycles. She improved significantly in symptoms and signs in one month. Her CAS score decreased to 2 from 5.

After 4th dose of IVMP her vision was slightly decreased to 6/36 on OD and no change in vision on OS. CAS again increased to 3 and Tab Azathioprin 50 mg BD was added and followed her in every month. After two months of Azathioprin her vision was further decreased in OD to CF1m and CAS became 4 and her colour vision was deranged so tab Azathioprin was stopped and Tab Mycophenolate Mofetil 500mg BD was given and followed in a month, she still did not improve. She developed OD RAPD. She also developed hyperemic, pale and blurring of optic disc on OD. Plain MRI orbit showed bulky all recti muscles with apical crowding causing compression of OU optic nerves, mild proptosis. MRI T2-weighted and fat suppressed images using TRIM (Turbo-Inversion – Recovery- Magnitude) and STIR (Short-Tau Inversion Recovery) showed marked extraocular muscle/orbital fat interstitial edema and inflammation.
She was sent for radiotherapy 20 Gy (2 Gy/day for 10 days).There was no improvement even after 2 months of Radiotherapy, ongoing tab Myacophenolate and oral corticosteroid so she underwent endoscopic orbital decompression on OD. She also underwent OD cat surgery with IOl implantation after 6 weeks of endoscopic orbital decompression.

After orbital decompression surgery patient was gradually improved, her diplopia was gone, RAPD became negative, her vision got back to 6/12 after 4 months of surgery and patient was significantly comfortable.



Plain MRI orbit T2 weighted , Mid Axial view –
Bulky recti muscles with apical crowding causing compression of BL optic nerves



Plain MRI orbit T2 weighted , Posterior coronal view

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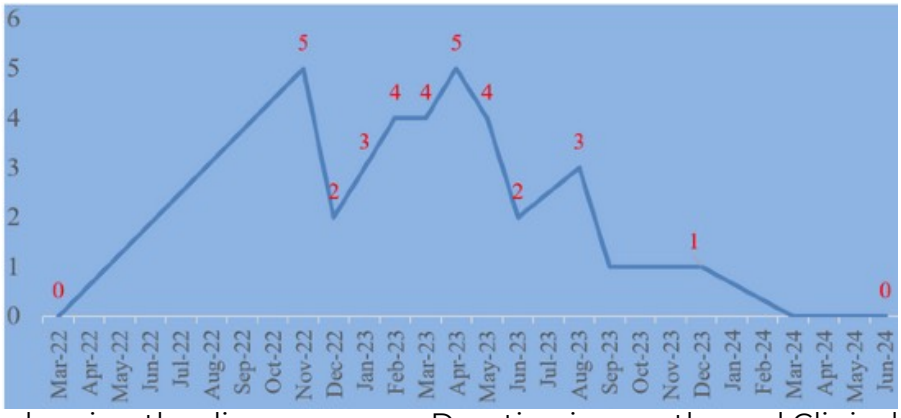
Here is the review table for detailed management and course of disease in this patient

Date	CAS Score	Intervention	Severeity	RE BCVA	LE BCVA	
Mar-22	0	LE Phaco + PCIOL	None	6/12	6/18	
Nov-22	5	IVMP 500 mg 0,1,2	Moderate	6/18	6/9	diplopia
Dec-22	2	IVMP500mg	Moderate	6/18	6/9	
Jan-23	3	IVMP 500 mg+ Azoran 50mg BD	Moderate	6/36	6/9	
Feb-23	4	IVMP 500 mg+ Azoran 50mg BD	Moderate	CFlm	6/9	
Mar-23	4	IVMP 500 mg+ Azoran 50mg BD	Severe	CFlm	6/12	deranged color vision
Apr-23	5	IVMP 1gm + Mycofenolate Mofetil 500mg BD	DON	CFCF	6/24	RE RAPD+ MRI Bulki recti muscles with apical Crowding
May-23	4	Mycofenolate Mofetil 500mg BD+ Radiotherapy	DON	PLPR	CF3m	RE RAPD+
Jun-23	2	Mycofenolate Mofetil 500mg BD+ Oral steroids	DON	HM+	CF3m	
Aug-23	3	Mycofenolate Mofetil 500mg BD+ Endoscopic orbital decompression BE	Moderate	HM+	6/60	diplopia
Sep-23	1	Mycofenolate Mofetil 500mg BD + RE Phaco +PCIOL	Moderate	CFCF	6/36	RAPD negative
Dec-23	1	Mycofenolate Mofetil 500mg BD	Mild	CFlm	6/24	No Diplopia
Mar-24	0	Mycofenolate Mofetil 500mg BD	Mild	5/60	6/18	
Jun-24	0	Mycofenolate Mofetil 500mg BD	Mild	6/12	6/18	

Table showing the course of the disease



Ryandal curve showing the disease course. Duration in months and Clinical Activity Score

Discussion

DON is a sight threatening complication which occurs in approximately 3-7% of patients with Graves orbitopathy. (9) Diagnosis of DON is made following clinical, ophthalmological and radiological examination. Decreased visual acuity, impaired color vision, visual field defects found in clinical evaluation as well as optic disc swelling/palor and RAPD revealed during ophthamological examination are indication of DON. (10)

First line treatment for DON recommended by the 2021 European Group on Graves Orbitopathy (EUGOGO) guidelines consist of high-dose intravenous methylprednesolone (IVMP) pulse therapy with 0.5-1.0g given for 3 consecutive days or on every second day which may be repeated for another week. If the response is poor or absent, orbital decompression must be performed promptly within 1-2 weeks.(11).

Immunomodulators like Azathioprin, Mycophenolate mefetil are also included into first-line treatment for mederate-severe and active GO in combination with IVMP, as it was found to result in significantly greater improvement in CAS and orbital signs and symptoms compared to IVMP alone (11,2, 13). The 2021 EUGOGO guidelines also recommend a combination of orbital rediotherapy with glucocorticoids as second-line treatment for moderate-to-severe and active Graves orbitopathy especially in patients with reduced eye muscle motility.

It is imperative to understand that the prognosis of DON largely depends upon whether the treatment was received and the stage or time at which the intervention was initiated. A timely intervention in DON saves vision, with full recovery in about 70% of cases.(14,15)

Conclusion

DON is an infrequent yet devastating consequence of Thyroid Eye Disease. Prompt recognition, confirmation with appropriate investigations, institution of medical followed by surgical treatment with postoperative rehabilitation help restore vision and patient satisfaction.

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DR AKSHAY NAIR

Dr Akshay Nair