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RESEARCH ARTICLE

A CASE REPORT OF VOGTS KOYANAGI-HARADA SYNDROME

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Abstract

Introduction: Vogt-Koyanagi-Harada syndrome (VKH) is a rare multi-systemic disease that affects tissues containing melanin, such as the eye, inner ear, meninges, and skin. It is characterized by chronic bilater al panuveitis associated with auditory, neurological, and cutaneous manifestations.

Material and Methodology : A 19 years old female presented to the out patient department of the ophthalmology department of Maharani Laxmi Bai Medical College, Jhansi with the following complaints for Drooping of upper eyelid in left eye since childhood and Retraction of left upper eyelid on jaw movement .

Management and Treatment : The patient was treated with high-dose systemic corticosteroids, consisting of intravenous methylprednisolone followed by oral prednisolone with a gradual taper. Topical corticostero ids and cycloplegic agents were administered for anterior segment inflammation. Long-term immunosuppressive therapy was initiated as required to achieve sustained control of inflammation and prevent disease recurrence. Regular follow-up with visual acuity assessment, fundus examination, and optical coherence tomography (OCT) was performed to monitor treatment response.(1,2)

Conclusion:Vogt–Koyanagi–Harada (VKH) disease is a multi-system autoimmune disorder that can lead to significant visual morbidity if not recognised and treated promptly. Early diagnosis, prompt initiation of high-dose corticosteroid therapy, and appropriate immunosuppressive treatment are crucial for achieving favourable visual and anatomical outcomes and preventing chronic recurrent inflammation.(3,4)

Introduction:-

Vogt-Koyanagi-Harada (VKH) syndrome is a rare multi-systemic disease that affects tissues containing melanin, including the eye, inner ear, meninges, and skin, characterized by chronic bilateral panuveitis associated with auditory, neurological, and cutaneous manifestations [5].The eponym derives from three 20th-century physicians: Alfred Vogt, who reported bilateral iridocyclitis in 1906; Yoshizo Koyanagi, who described bilateral serous retinal detachments with CSF pleocytosis in 1929; and Einosuke Harada, who characterized acute diffuse choroiditis as a hallmark feature of the disease [6].VKH occurs more commonly in patients with a genetic predisposition, including those from Asian, Middle Eastern, Hispanic, and Native American populations, with several HLA associations identified, including HLA-DR4, HLA-DR53, and HLA-DQ4 [7]. Pathophysiologically, VKH manifests due to loss of immune tolerance to melanocytes within the meninges, eyes, skin, hair, and ears, involving a T lymphocyte-

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mediated autoimmune process directed against melanocyte tyrosinase-related proteins [8].Patients typically present with sudden visual loss, ocular pain, and photophobia, followed weeks to months later by cutaneous signs such as vitiligo, poliosis, and alopecia. Four clinical stages have been described: prodromal, uveitis, chronic, and recurrent [9].The key to successful management is early and aggressive systemic treatment, as delayed therapy carries a more guarded visual prognosis and a greater risk of chronic inflammation. Current evidence supports initiating high-dose corticosteroids combined with mycophenolate mofetil (MMF) within 2–3 weeks of symptom onset to prevent chronic disease progression and achieve optimal visual outcomes [10].

Clinical Features and stages:-

Clinical features:-

VKH is characterized by bilateral granulomatous panuveitis, exudative retinal detachments, meningeal signs, hearing loss, and pigment loss (vitiligo, poliosis, and alopecia).

Clinical Stages:-

Four clinical stages have been described: the prodromal stage, uveitic stage, chronic stage, and recurrent stage. The prodrome typically lasts 7–10 days and is characterized by flu-like symptoms. Patients usually initially present to an ophthalmologist for sudden loss of vision, ocular pain, and photophobia, while hearing disturbances and dizziness may also be present. After weeks or months, most patients notice cutaneous signs such as hair loss, poliosis, and vitiligo.

Causes:-

Vogt-Koyanagi-Harada (VKH) syndrome is an autoimmune disorder primarily targeting melanin-containing tissues. Although the exact aetiology remains incompletely understood, three major contributing factors have been identified: autoimmune mechanisms, genetic predisposition, and environmental triggers.

Autoimmune Mechanism:-

The primary pathogenesis is a T-cell-mediated autoimmune response directed towards melanocyte or melanocyte-associated antigens, causing inflammation of the choroidal layer and diffuse inflammatory conditions throughout the eye [11]. More specifically, VKH involves T CD4+ Th1 lymphocyte-mediated aggression to melanocytes, targeting pigmented structures such as the eyes, ear, skin, meninges, and hair [12]. Alongside T lymphocytes, auto-reactive B cells also play a central role in disease development and propagation, with tyrosinase family proteins identified as important disease-specific antigens [13].

Genetic Predisposition:-

Genetic factors, including VKH-specific risk factors such as HLA-DR4 and general risk factors for immune-mediated diseases including IL-23R, are involved in disease development [14]. HLA-DRB1*0405 in particular plays an important role in pathogenesis, rendering carriers more susceptible to the disease [15].

Environmental Triggers:-

Environmental triggering factors are also involved in VKH development alongside immune dysfunction and genetic susceptibility, including cutaneous injury, head trauma, and certain medications such as interferon therapy [14].

Pathophysiology:-

VKH manifests due to loss of immune tolerance to melanocytes within the meninges, eyes, skin, hair, and ears. There is evidence to suggest it involves a T lymphocyte-mediated autoimmune process directed against melanocyte tyrosinase-related proteins, which are present in the uvea, retina, and leptomeninges.

Epidemiology and Genetics:-

VKH occurs more commonly in patients with a genetic predisposition, including those from Asian, Middle Eastern, Hispanic, and Native American populations. Several HLA associations have been identified, including HLA-DR4, HLA-DR53, and HLA-DQ4.

Material and Methodology:-

This is a case report of a 19 years old female patient who came to Maharani Laxmi Bai Medical College, Jhansiwith the following complaints for, Drooping of upper eyelid in left eye since childhood and Retraction of left upper eyelid on jaw movement case of Vogt–Koyanagi–Harada (VKH) syndrome. Detailed ocular and systemic examination were done to rule out any other abnormalities, which included history of the presenting illness, visual

acuity using Snellen's chart, , complete ocular examination using slit lamp, intra-ocular pressure using non-contact tonometer. Blood was sent for routine laboratory investigations. Patient wasadmitted and following blood investigations were sent

Case Report:-

A 19 years old female presented to the out patient department of the ophthalmology department of MLBMC Jhansi with the following complaints for, Drooping of upper eyelid in left eye since childhood and Retraction of left upper eyelid on jaw movement. According to the patient, she was apparently asymptomatic during childhood after when she developed drooping of left upper eyelid in which was acute in onset painless and gradually progressive in nature. There was no history of-Diminution of vision, Pain and Watering. No history of diabetes mellitus/ hypertyension/asthma, no history of trauma in either eye , no history of any chronic drug use, no history of any drug allergy. No relevant family history. Patient wasadmitted and following blood investigations were sent .

Ocular Examination

	Right Eye	Left Eye
Visual Acuity	FC at 1m	6/6
Best corrected Visual Acuity	FC at 1m	6/6
Perception to light	Present	Present
Perception to Rays	Present in all Quadrants	Present in all Quadrants
Orbital Margins	Intact on Palpation	Intact on Palpation
Ocular Movements	Within normal limits	Within normal limits
Eyelid/Eyebrows	Within normal limits	Within normal limits
Conjunctiva /Sclera	Within normal limit	Within normal limit
Cornea	Clear , stain negative	Clear , stain negative
Anterior Chamber	VH III	VH III
Iris	Normal color and pattern	Normal color and pattern
Pupil	Round /Regular/Reactive	Round /Regular/Reactive
Lens	Greyish Black Reflex	Greyish Black Reflex
Final Glow	Good	Good
IOP		

Fundus Examination

	Right Eye	Left Eye
Media	Clear	Clear
Optic Disc	DISC MARGIN CLEAR , WITH CDR not appreciated , DISC SIZE WNL WITH HYPERAEMIC DISC	Disc margin clear with CDR 0.3-0.4
Macula	FR present	FR present
Blood Vessels	Within normal limits	Within normal limits
Background and Periphery	MULTIPLE POCKETS OF SUB-RETINAL FLUID AT MACULA AND PERIPAPPILARY REGION s/o MULTIPLE EXUDATIVE RD , DIFFUSE RETINAL OEDEMA	Within normal limits

Od Os

Fig 1

Provisional Diagnosis:-

On The Basis Of History And Ocular Examinations, The Provisional Diagnosis Is Vogt-Koyanagi-Harada Syndrome (Vkh) .

Treatment:-

The key to successful therapy for VKH disease is early and aggressive systemic treatment. Patients treated later in the course of the disorder have a more guarded prognosis for recovery of visual acuity and a greater risk for chronic inflammation.

1. Early high-dose systemic corticosteroids are the cornerstone of treatment. Prednisolone 1–2 mg/kg/day or intravenous methylprednisolone pulse therapy (1 g/day for 3 days) should be started as early as possible, followed by a slow taper over at least 6 months.
2. Combined corticosteroid and immunosuppressive therapy is now recommended as first-line treatment in acute VKH to reduce chronic recurrence and sunset-glow fundus.

3. **Common steroid-sparing immunosuppressants include:**
   - Mycophenolate mofetil
   - Methotrexate
   - Azathioprine
   - Cyclosporine
   - Tacrolimus
4. Refractory cases may require biologics such as Adalimumab or Rituximab.

Patient was admitted and following treatment was done :

E/D Predfort1d 3t/d
E/D Homide 1d 3t/d
I.V Methylprednisolone 1000mg for 3 days
After starting MPS , vision improved
After 1st dose : from FC at 1 M to FC at 3 M
Subsequently after 3rd dose : 6/18 (at the time of discharge)
On fundus examination there was reduction in sub-retinal fluid

OD
Fig2

She was discharged on
Tab Wysolone 40 mg OD for 7 days
and B/E OCT macula and Disc was advised.

Conclusion:-

Vogt–Koyanagi–Harada disease is a rare autoimmune multi-system disorder that can lead to significant visual morbidity if not recognised and treated promptly. This case highlights the importance of early diagnosis based on characteristic clinical findings and multimodal imaging. Timely initiation of high-dose systemic corticosteroids, with appropriate immunomodulatory therapy when indicated, resulted in favourable anatomical and visual outcomes. Regular follow-up is essential to monitor disease activity, prevent recurrences, and minimise long-term complications. This report emphasises that early aggressive therapy remains the cornerstone for preserving vision and improving prognosis in patients with VKH disease.Early diagnosis and prompt initiation of high-dose corticosteroids combined with immunosuppressive therapy remain the current standard of care for VKH. Studies by Rao (2006), Herbort et al. (2021), and Urzua et al. (2022) have shown that combination therapy significantly decreases chronic recurrence, sunset-glow fundus, and vision-threatening complications while improving long-term visual outcomes.

Disclosure:-

All the photographs used in this journal have been published after obtaining informed consent and permissions from the patient. Financial support and sponsorship: Nil Conflicts of interest: Nil

References:-

1. Herbort CP Jr, Tugal-Tutkun I, Abu-El-Asrar A, Gupta A, Takeuchi M, Fardeau C, et al. Precise, simplified diagnostic criteria and optimised management of initial-onset Vogt–Koyanagi–Harada disease: an updated review. Eye (Lond). 2022;36(1):29–43.
2. Lavezzo MM, Sakata VM, Morita C, Rodriguez EEC, Abdallah SF, da Silva FTG, et al. Vogt-Koyanagi-Harada disease: review of a rare autoimmune disease targeting antigens of melanocytes. Orphanet J Rare Dis. 2016;11:29.
3. Herbort CP Jr, Abu El-Asrar AM, Takeuchi M, et al. Vogt–Koyanagi–Harada disease: recent advances in pathophysiology, diagnosis and management. Eye (Lond). 2021;35(2):478–492.
4. Abu El-Asrar AM, Al Tamimi M, Hemachandran S, Al-Mezaine HS, Kangave D. Prognostic factors for clinical outcomes in Vogt–Koyanagi–Harada disease treated with high-dose corticosteroids and immunomodulatory therapy. Immunol Inflamm. 2018;26(4):562–573.
5. [5] ScienceDirect Topics — Vogt-Koyanagi Syndrome Overview

6. StatPearls — Vogt-Koyanagi-Harada Syndrome (NCBI Bookshelf)
7. Medscape — VKH Disease: Background, Aetiology, Epidemiology
8. PMC — VKH: Brain Eye and Ear Syndrome (Shariff & Alzuabi, 2024)
9. Medscape — VKH Disease Clinical Presentation
10. DrOracle — Treatment of VKH Disease (2026)
11. Rathinam & Vijayalakshmi — ScienceDirect: Vogt–Koyanagi–Harada Syndrome: Perspectives for Immunogenetics, Multimodal Imaging, and Therapeutic Options (2016)
12. Greco et al. — PubMed: Vogt-Koyanagi-Harada Syndrome: A New Perspective for Healthcare Professionals (2020)
13. Nascimento et al. — PMC: New Perspectives on the Immunopathogenesis and Treatment of Uveitis Associated With VKH Disease (2021)
14. Yang et al. — PubMed: Vogt-Koyanagi-Harada Disease: Novel Insights into Pathophysiology, Diagnosis and Treatment (2016)
15. Damico et al. — PubMed: Diagnosis and Classification of Vogt-Koyanagi-Harada Disease (2014)