Content available at: https://www.ipinnovative.com/open-access-journals

Indian Journal of Clinical and Experimental Ophthalmology

Journal homepage: www.ijceo.org

Original Research Article

Comprehensive analysis of anterior and posterior segment ocular manifestations in sickle cell disease: A cross-sectional study in western Odisha

Ipsita Khuntia[1]*, Sharmistha Behera[2], Sasmita Sahu[2], Jeneeta Baa[2], Mahesh Sahoo[2], Prangnya Nanda[2]

[1]*Dept. of Ophthalmology, Postgraduate Institute of Medical Education & Research, Bhubaneswar, Odisha, India*
[2]*Dept. of Ophthalmology, Veer Surendra Sai Institute of Medical Sciences And Research (VIMSAR), Burla, Odisha, India*

ARTICLE INFO

Article history:
Received 03-08-2024
Accepted 09-11-2024
Available online 21-02-2025

Keywords:
Sickle cell disease
Ocular manifestations
Proliferative sickle retinopathy
Genetic counseling

ABSTRACT

Background: This study investigates the anterior and posterior segment ocular manifestations in sickle cell disease (SCD) patients in Western Odisha.
Materials and Methods: Conducted at Veer Surendra Sai Institute of Medical Sciences and Research (VSSIMSAR) from November 2018 to October 2020, the study involved 103 patients (204 eyes). The prevalence, clinical profiles, and demographic associations of ocular manifestations were assessed using advanced clinical and imaging techniques.
Results: The study highlighted a high prevalence of ocular morbidity in SCD patients, including cataract, glaucoma, and various forms of retinopathy. Anterior segment findings revealed conjunctival vessel abnormalities and iris atrophy. Posterior segment findings included retinal vessel tortuosity and proliferative sickle retinopathy (PSR). The analysis underscored clinical diversity, with significant genotype-specific and age-related associations observed among the patients.
Conclusion: Routine ocular screenings and early interventions are crucial to mitigate the risks of severe vision impairment in SCD patients. The use of advanced imaging techniques aids in precise diagnosis, thereby improving clinical outcomes. Public health initiatives that promote awareness and genetic counseling are essential for the effective management of SCD-related ocular manifestations. This study provides critical insights into the ocular health of SCD patients, guiding future research and healthcare strategies to address these complexities comprehensively. By emphasizing the importance of early detection and intervention, the findings aim to reduce the burden of ocular complications in this patient population.

1. Introduction

Sickle cell disease (SCD) represents a significant global health burden, particularly prevalent in regions like Western Odisha, where the disease affects approximately 9.1% of the population.[1] This genetic disorder results from a single amino acid substitution in the hemoglobin molecule, leading to chronic hemolysis and recurrent vaso-occlusive crises.[2] These crises cause pain and organ damage, and the vascular pathology of SCD extends to the ocular system, impacting both anterior and posterior segments of the eye, potentially culminating in severe visual impairment and blindness.

In recent years, advancements in healthcare have extended the life expectancy of SCD patients. As a result, the incidence of long-term complications, including ocular manifestations, has increased.[3] These complications range from proliferative retinopathy, vitreous hemorrhage, and retinal detachment, to less common manifestations such as conjunctival vessel abnormalities and iris atrophy. The severity and diversity of these ocular

* *Corresponding author*.
E-mail address: ipsita.khuntia3@gmail.com (I. Khuntia).

https://doi.org/10.18231/j.ijceo.2025.023

complications necessitate a comprehensive understanding of their epidemiology and clinical management.[4]

Ocular manifestations in SCD can significantly affect patients' quality of life. The anterior segment complications, such as conjunctival corkscrew vessels, icterus, and cataract formation, are often more visible and can be easier to diagnose early.[5] In contrast, posterior segment complications, including retinal vessel tortuosity, non-proliferative sickle retinopathy (NPSR), and proliferative sickle retinopathy (PSR), are more insidious and can lead to more severe visual impairment if not detected and treated promptly.[6] Early diagnosis and intervention are crucial in preventing these complications from progressing to irreversible stages.

This cross-sectional study conducted at Veer Surendra Sai Institute of Medical Sciences and Research (VSSIMSAR) in Western Odisha aims to provide a detailed analysis of anterior and posterior segment ocular manifestations in SCD patients.[7] By examining the prevalence, types, and clinical profiles of these manifestations within our tertiary healthcare center, we seek to underscore the critical importance of early diagnosis, preventive measures, and timely intervention.

Furthermore, this study explores the influence of demographic factors such as age, sex, and genotype on the frequency and severity of ocular manifestations. Understanding these associations can help tailor more effective diagnostic and therapeutic strategies. The study also evaluates the current diagnostic and therapeutic practices available locally, aiming to identify gaps in care and opportunities for improvement.[8]

By elucidating the clinico-epidemiological associations of ocular complications in SCD, this research aims to guide healthcare providers in optimizing patient care and enhancing public health strategies.[9] These findings are expected to provide a foundation for developing targeted screening programs and individualized treatment plans that can better manage and mitigate the morbidity associated with SCD-related ocular diseases.

Ultimately, our findings aspire to mitigate the morbidity associated with SCD-related ocular diseases, thereby improving the quality of life for affected individuals in our region and beyond.[10,11] By highlighting the importance of routine ocular screenings and the use of advanced imaging techniques, this study advocates for proactive measures to prevent severe vision impairment and blindness in SCD patients. Public health efforts promoting awareness, genetic counseling, and patient education are essential components of a comprehensive approach to managing the ocular complications of SCD effectively.

2. Materials and Methods

2.1. Study design

This study employed a hospital-based cross-sectional design to investigate ocular manifestations in sickle cell disease (SCD) patients at Veer Surendra Sai Institute of Medical Sciences and Research (VSSIMSAR), Burla, Sambalpur, Odisha, from November 2018 to October 2020.

2.2. Study location and population

The study focused on SCD patients attending the Ophthalmology Outpatient Department and Sickle Cell Unit at VSSIMSAR, including referrals from Medicine and Pediatric departments. The majority of patients managed at the Sickle Cell Unit were from western Odisha.

2.3. Sample size and sampling technique

A total of 103 subjects were enrolled, exceeding the calculated minimum sample size of 87. All eligible patients diagnosed with SCD, aged 6 years and above, were included. Sampling included patients attending routine follow-up and those referred during the study period.

2.4. Inclusion criteria

1. Sickle cell patients attending the Ophthalmology outpatient department.
2. Patients reporting for routine follow-up at the Sickle Cell Unit.
3. Patients referred from the Medicine department.
4. Patients referred from the Pediatric department (aged >5 years).

2.5. Exclusion criteria

1. Patients who refused to provide informed consent
2. Patients less than 6 years of age
3. Eyes with a history of serious injuries or extensive surgery
4. Patients with concurrent diabetic retinopathy, hypertensive retinopathy, talc emboli, Eales disease, sarcoidosis, infectious diseases (e.g., dengue), ocular ischemic syndromes, familial exudative vitreoretinopathy, chronic myelogenous leukemia, or scleral buckle.

Data collection procedure: Data collection included:

1. Structured interviews with patients or guardians using pretested questionnaires
2. Detailed demographic and clinical history including genotype, hemoglobin levels, and treatment duration
3. Comprehensive ocular examinations:
4. Visual acuity assessment using Snellen and Landolt's C charts

5. Anterior segment examination with slit lamp biomicroscopy and tonometry
6. Fundoscopy with direct and indirect ophthalmoscopy, and in selected cases, OCT and B-scan imaging
7. Assessment of ocular alignment, motility, and pupillary reflexes

2.6. Ethical considerations

1. Ethical approval obtained from the VSSIMSAR Ethical Committee
2. Informed written consent obtained from all participants or guardians
3. Confidentiality of participant information strictly maintained

2.7. Data management & analysis

1. Data recorded and managed using Microsoft Excel
2. Statistical analysis performed using SPSS:
3. Calculation of mean, standard deviation, and prevalence rates
4. Comparison of parametric and non-parametric data using appropriate statistical tests.

3. Results

Table 1: Demographic and clinical characteristics of study participants

Characteristic	Frequency (n=103)	Percentage (%)
Mean Age (years)	32.49 (range 6-65)	-
Gender		
Female	65	63.1
Male	38	36.8
Genotype		
HbSS	60	58.2
HbSC	43	41.7

Table 1 outlines the demographic characteristics of the study participants, including age distribution, gender, and genotype distribution.

Table 2: Prevalence of anterior segment ocular manifestations

Anterior Segment Signs	Frequency (n=103)	Percentage (%)
Conjunctival Corkscrew Vessels	43	41.74
Icterus	38	36.9
Iris Atrophy	1	0.97
Cataract	3	2.9

Table 3 presents the prevalence of various anterior segment ocular manifestations observed in the study population.

Table 3 shows the distribution of anterior segment signs among different genotypes, highlighting the higher prevalence of conjunctival corkscrew vessels in theHbSC genotype.

Table 4 illustrates the significant age associations with anterior segment signs, indicating higher prevalence of these signs in certain age groups.

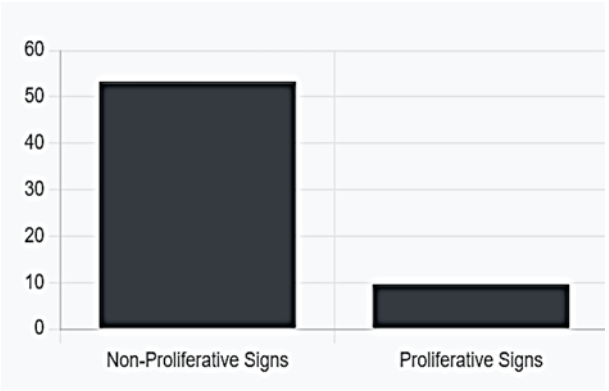

Figure 1: Prevalence of posterior segment ocular manifestations

Graph 1 depicts the prevalence of posterior segment ocular manifestations, showing that non-proliferative signs are more common than proliferative signs.

Table 5 shows the genotype-specific distribution of posterior segment signs, indicating a higher prevalence of non-proliferative signs in both genotypes and a higher prevalence of proliferative signs in the HbSS genotype.

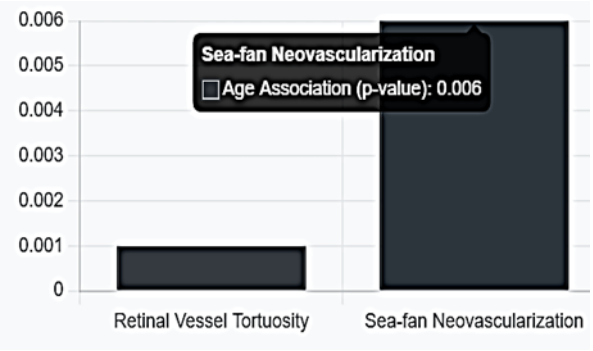

Figure 2: Age association with posterior segment signs

Figure 2 illustrates the significant age associations with posterior segment signs, particularly retinal vessel tortuosity and sea-fan neovascularization, with higher prevalence in older age groups.

Table 6 provides an overview of visual acuity among the study participants and identifies the primary causes of vision loss in the patient population.

This comprehensive analysis underscores the high prevalence of both anterior and posterior segment ocular manifestations in SCD patients, with significant genotype-

Table 3: Genotype-specific distribution of anterior segment signs

Anterior Segment Signs	HbSC (n=43)	Percentage (%)	HbSS (n=60)	Percentage (%)
Conjunctival Corkscrew Vessels	23	53.5	20	33.3
Icterus	17	39.5	21	35
Iris Atrophy	0	0	1	1.67
Cataract	1	2.3	2	3.33

Table 4: Age association with anterior segment signs

Age Group (years)	Conjunctival Corkscrew Vessels (p = 0.005)	Icterus (p = 0.024)	Cataract (p = 0.024)
6-20	12 (28%)	8 (21%)	0
21-40	25 (58%)	21 (55%)	1
41-65	6 (14%)	9 (24%)	2

Table 5: Genotype-specific distribution of posterior segment signs

Posterior Segment Signs	HbSC (n=43)	Percentage (%)	HbSS (n=60)	Percentage (%)
Non-Proliferative Signs	22	51.1	33	55
Proliferative Signs	2	4.65	8	13.3

Table 6: Visual acuity and causes of vision loss

Visual Acuity	Frequency (n=103)	Percentage (%)
Normal Vision	55	53.3
Mild to Moderate Vision Impairment	45	43.7
Severe Vision Impairment	3	2.9
Causes of Vision Loss		
Cataract	2	1.94
Sea-fan Neovascularization	1	0.97
Vitreous Hemorrhage	1	0.97

specific differences and age associations for certain ocular signs. Understanding these manifestations is crucial for comprehensive management and early intervention in SCD patients. The use of routine ocular screenings and advanced imaging techniques is recommended to mitigate the risks of severe vision impairment. Public health initiatives promoting awareness and genetic counseling are essential for effective management.

4. Discussion

Our study conducted a detailed examination of anterior and posterior segment ocular manifestations in sickle cell disease (SCD) patients in Western Odisha, highlighting the significant prevalence and clinical implications of these ocular complications. The findings underscore the importance of regular ophthalmic screening and targeted management strategies to prevent severe vision impairment in this patient population.

Conjunctival vessel abnormalities, observed in 41.74% of patients, were one of the most common anterior segment manifestations. These abnormalities, characterized by corkscrew-shaped vessels, were found to have a slightly higher prevalence in individuals with the HbSC genotype. This finding aligns with previous studies suggesting that the HbSC genotype may be associated with a more benign clinical course yet can still present significant ocular complications.[12] Jaundice, noted in 20.3% of patients, correlated with markers of disease severity, including hemolysis and liver dysfunction. The presence of jaundice in the conjunctiva serves as a visible indicator of systemic disease activity and highlights the interconnectedness of ocular and systemic health in SCD patients.

Posterior segment complications were also prevalent, with increased retinal vessel tortuosity observed in 30.09% of patients. This manifestation was significantly associated with the HbSS genotype and female gender, suggesting that these demographic factors may influence the severity of retinal changes in SCD.[13] The presence of proliferative sickle retinopathy (PSR), including sea-fan neovascularization (4.8%) and vitreous hemorrhage (2.91%), underscores the risk of severe vision complications. These findings are particularly concerning as PSR can lead to retinal detachment and irreversible vision loss if not detected and managed promptly.

Variability in the prevalence of PSR across different populations highlights the role of genetic and environmental factors in disease severity. For instance, studies from different geographic regions have reported varying rates of

PSR, suggesting that local environmental factors, healthcare access, and genetic background may all contribute to the observed differences.[14] This variability underscores the need for region-specific studies to tailor screening and treatment protocols effectively.

The high prevalence of both anterior and posterior segment ocular manifestations in our cohort underscores the need for regular ophthalmic screening in SCD patients. Early detection of conditions such as PSR is crucial for timely intervention, which can prevent progression to more severe complications like retinal detachment and blindness. Additionally, the use of advanced imaging techniques, such as optical coherence tomography (OCT) and fluorescein angiography, can aid in the precise diagnosis and monitoring of ocular changes in SCD patients.[15]

Our study's single-center design and relatively small sample size are notable limitations. These factors may limit the generalizability of our findings to the broader SCD population. Future studies should aim to include larger, multicenter cohorts to validate our results and explore additional risk factors for ocular manifestations in SCD. Moreover, longitudinal studies are needed to understand the progression of ocular changes over time and the long-term efficacy of different management strategies.[4]

Our study provides comprehensive insights into the ocular manifestations of SCD patients in Western Odisha. The high prevalence of both anterior and posterior segment complications highlights the need for targeted screening and management strategies to improve visual outcomes in this population. Regular ophthalmic evaluations, coupled with advanced imaging techniques, can facilitate early detection and intervention, thereby reducing the risk of severe vision impairment. Additionally, public health efforts should focus on raising awareness about the ocular complications of SCD and promoting genetic counseling to help manage the disease more effectively. Our findings underscore the critical importance of integrating ophthalmic care into the routine management of SCD patients to enhance their quality of life and overall health outcomes.

5. Conclusion

This study underscores the critical importance of ocular health management in addressing the national concern posed by SCD. The high prevalence of ocular morbidity, including conditions such as cataract, glaucoma, and various forms of retinopathy, highlights the urgent need for routine ocular examinations and follow-up check-ups. Early detection through these measures is pivotal in preventing visual impairment and loss, particularly stemming from vaso-occlusive crises affecting the eyes. The severity of ocular manifestations, notably proliferative sickle retinopathy (PSR), correlates closely with the systemic severity of SCD, emphasizing the necessity for targeted screening programs. Advances in ocular imaging have revolutionized diagnostic precision, enabling early intervention and management of sickle cell retinopathy, thereby mitigating blindness risks. Public health initiatives, including community education on consanguineous marriages and widespread awareness about early ocular symptoms, are essential in reducing the prevalence of SCD and its associated ocular manifestations. By integrating these strategies into healthcare policies and promoting regular screening procedures, we can effectively mitigate a substantial portion of ocular morbidities in SCD patients in Western Odisha and similar regions.

In conclusion, this study underscores that proactive ocular health management is crucial in safeguarding SCD patients from visual impairment. Implementing comprehensive screening protocols and advancing public awareness are pivotal steps towards alleviating the burden of ocular morbidity in the context of sickle cell disease.

6. Source of Funding

None.

7. Conflict of Interest

None.

References

1. Serjeant GR. The natural history of sickle cell disease. *Cold Spring Harb Perspect Med*. 2013;3(10):a011783.
2. Stuart MJ, Nagel RL. Sickle-cell disease. *Lancet*. 2004;364(9442):1343–60.
3. Connes P, Coates TD. Autonomic nervous system dysfunction: implication in sickle cell disease. *Curr Opin Hematol*. 2016;23(3):224–30.
4. Kato GJ, Piel FB, Reid CD, Gaston MH, Ohene-Frempong K, Krishnamurti L, et al. Sickle cell disease. *Nat Rev Dis Primers*. 2018;4:18010.
5. Osafo-Kwaako A, Kimani K, Ilako D, Akafo S, Ekem I, Rodrigues O, et al. Ocular manifestations of sickle cell disease at the Korle-bu Hospital, Accra, Ghana. *Eur J Ophthalmol*. 2011;21(4):484–9.
6. Pahl DA, Green NS, Bhatia M, Chen R. New Ways to Detect Pediatric Sickle Cell Retinopathy: A comprehensive review. *J Pediatr Hematol Oncol*. 2017;39(8):618–25.
7. Ware RE, Montalembert MD, Tshilolo L, Abboud MR. Sickle cell disease. *Lancet*. 2017;390(10091):311–23.
8. al Salem M. Benign ocular manifestations of sickle cell anemia in Arabs. *Indian J Ophthalmol*. 1991;39(1):9–11.
9. Al-Salem M, Ismail L. Ocular manifestations of sickle cell anaemia in Arab children. *Ann Trop Paediatr*. 1990;10(2):199–202.
10. AlRyalat SA, Nawaiseh M, Aladwan B, Roto A, Alessa Z, Al-Omar A. Ocular Manifestations of Sickle Cell Disease: Signs, Symptoms and Complications. *Ophthalmic Epidemiol*. 2020;27(4):259–64.
11. Elendu C, Amaechi DC, Alakwe-Ojimba CE, Elendu TC, Elendu RC, Ayabazu CP, et al. Understanding Sickle cell disease: Causes, symptoms, and treatment options. *Medicine (Baltimore)*. 2023;102(38):e35237.
12. Shukla P, Verma H, Patel S, Patra PK, Bhaskar L. Ocular manifestations of sickle cell disease and genetic susceptibility for refractive errors. *Taiwan J Ophthalmol*. 2017;7(2):89–93.
13. Elagouz M, Jyothi S, Gupta B, Sivaprasad S. Sickle cell disease and the eye: old and new concepts. *Surv Ophthalmol*. 2010;55(4):359–77.
14. Fadugbagbe AO, Gurgel RQ, Mendonça CQ, Cipolotti R, Santos A, Cuevas LE, et al. Ocular manifestations of sickle cell disease. *Ann*

Trop Paediatr. 2010;30(1):19–26.
15. Clarkson JG. The ocular manifestations of sickle-cell disease: a prevalence and natural history study. *Trans Am Ophthalmol Soc*. 1992;90:481–504.

Author's biography

Ipsita Khuntia, Assistant Professor

Sharmistha Behera, Professor

Sasmita Sahu, Associate Professor

Jeneeta Baa, Associate Professor

Mahesh Sahoo, PG Trainee

Prangnya Nanda, PG Trainee

Cite this article: Khuntia I, Behera S, Sahu S, Baa J, Sahoo M, Nanda P. Comprehensive analysis of anterior and posterior segment ocular manifestations in sickle cell disease: A cross-sectional study in western Odisha. *Indian J Clin Exp Ophthalmol* 2025;11(1):132-137.