

CASE REPORT

Orbital compression syndrome in a Ugandan child with sickle cell disease: A case report

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Funding information

This paper received no funding from any organization or individual

Abstract

Orbital compression syndrome is a rare acute complication of sickle cell disease that may impair vision. Assessment by a multidisciplinary team incorporates detailed history and physical examination, fundoscopy, and appropriate imaging to exclude infections or neoplasms. Supportive treatment is adequate unless there is evidence of life-threatening space-occupying lesion warranting surgery.

KEYWORDS

orbital compression syndrome, sickle cell anemia, Uganda

1 | INTRODUCTION

Sickle cell disease (SCD) is the most common childhood hemoglobinopathy. Africa is the most affected continent with about 300,000 children born with SCD annually.¹ In Uganda, approximately 20,000 children are born with SCD every year. A recent survey showed that the national prevalence of SCD and sickle cell trait (SCT) in Uganda is 0.7% and 13.3% respectively, with some regions having prevalence of SCD and SCT as high as² (19.8%) and 1.5% respectively.³ The hallmark of SCD is recurrent hemolytic anemia and vaso-occlusion, which may manifest as acute crises and chronic complications. These SCD-related complications can affect almost all systems of the body.

Ocular complications include ischemia, retinopathies, secondary glaucoma, and rarely orbital bone infarction leading to orbital compression syndrome (OCS).⁴ A few cases of OCS have been reported in sub-Saharan African countries, including Sudan⁵ and a similar presentation in Nigeria.⁶ To our knowledge, this is the first case of OCS from Uganda in the literature despite the high burden of SCD in the country.

2 | CASE PRESENTATION

We report an 11-year-old boy, a known patient with SCD (HBSS type). He was referred from a peripheral hospital to

the pediatric emergency Unit of Mulago National Referral Hospital with a one-week history of bilateral eyelid swelling and progressive proptosis, more marked on the left eye with associated reduction in vision. The patient also had fever and severe headache but no history of trauma, neck pain, convulsions, or loss of consciousness. He had bilateral lower limb pains but with no joint pain or swelling. He was receiving prophylactic daily folic acid and monthly sulfadoxine/pyrimethamine (fansidar) but not on hydroxyurea. He had been transfused with two units of whole blood at the peripheral hospital prior to admission.

On examination, the child was sick looking with mild pallor of mucous membranes and mild icterus but no dehydration and lymphadenopathy. He was febrile with an axillary temperature of 38.0°C. The pulse rate was 100 beats per minute, respiratory rate at 32 breaths per minute, blood pressure of 112/66 mmHg, and oxygen saturation of 98% on room air. The left eyeball was protruding and hyperemic with a pus discharge, tearing with eyelid edema without pupillary reflexes elicited (Figure 1). The right eye was mildly swollen with mild hyperemia but no pus discharge or tearing. The right pupil was small and reacting poorly to light. There was a firm tender swelling measuring about 2 × 3 cm on the right parietal scalp. Per abdomen, there was a palpable spleen, about 4 cm below the costal margin in the left mid-clavicular line. Both lower limbs were tender below the knee joints but there were no swellings.

3 | LABORATORY INVESTIGATIONS AND IMAGING

The laboratory investigations included a complete blood count (CBC) prior to transfusion which showed a total white blood cell count of $18.9 \times 10^9/L$, hemoglobin of 8.2 g/dl, mean corpuscular volume of 75 fL, mean corpuscular hemoglobin concentration of 30.0 g/dl, red cell distribution width of 18.5%, and a platelet count of $194 \times 10^3/\mu l$. An electrolyte panel showed a mild hyponatremia ($Na^+ = 131.3$ mmol/L) but normal potassium (4.12 mmol/L), chloride (100.9 mmol/L), creatinine (0.36 mg/dl), and urea levels (8.1 mg/dl). Plasmodium

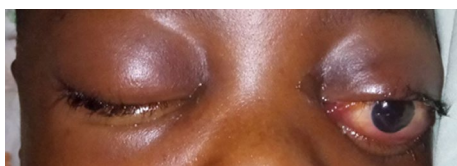


FIGURE 1 Face photograph showing proptosis and pus discharge in the left eye

hemoparasites were absent on smear and microscopy whereas blood culture and sensitivity were not performed due to financial constraints.

A contrasted axial computed tomography showed features of multiple extra-axial hypodense biconvex collections in the right frontal (measuring 2.13×3.6 cm) and left temporal regions suggestive of epidural hematomas (Figure 2). Similar lesions were also seen in the extra-conical superolateral aspects of both orbits, marked on the left orbit (Figure 3). There was left orbital proptosis, but the adjacent bones appeared normal (Figure 3). Other brain findings were normal with no space-occupying lesions and left frontal, ethmoidal, and sphenoidal sinusitis (Figure 2).

An ocular exam of the left eye by the ophthalmologists revealed proptosis, chemosis, and hyperemia with a mild epithelial defect of punctate staining pattern. The lens was normal, and the anterior chamber was deep and quiet with no signs of inflammation. Visual acuity was 6/60 and an intraocular pressure of 11 mmHg. The right eye was otherwise normal with a visual acuity of 6/24 and intraocular pressure of 16 mmHg.

4 | MANAGEMENT AND FOLLOW-UP

He was managed as a patient with SCD with orbital compression syndrome, vaso-occlusive crisis, and probable bacterial sepsis with intravenous antibiotics (ceftriaxone 2 g daily and ampicillin/cloxacillin 500 mg four times daily for 2 weeks), hydration (intravenous fluids), corticosteroids (daily prednisolone 20 mg for five days tapered in 2 weeks), and analgesia (oral morphine and paracetamol). A protective eye shield and tetracycline ointment were also provided for the left eye to prevent exposure keratopathy.

By day 7 of admission, there was marked clinical improvement. The pain and fever had subsided. A repeat CBC on the 7th day showed white blood cell count of $9.89 \times 10^9/L$, hemoglobin of 9.7 g/dl, mean corpuscular volume of 78.5 fL, mean corpuscular hemoglobin concentration of 31.4 g/dl, and red cell distribution width of 22.7%.

Following the recommendation of the neurosurgery and otolaryngology team, a right frontal craniotomy was performed for collection of pus from the ethmoidal and sphenoidal sinuses. A necrotic frontal bone was seen during the surgery, but there was no pus collection in the ethmoidal and sphenoidal sinuses as earlier anticipated. The child had recovered substantively by the end of the first post-operative week and was discharged home.

FIGURE 2 An axial CT showing a right frontal hypodense lesion (A). A left temporal hypodense lesion, a left ethmoid, and bilateral sphenoid hyperdense fullness (B)

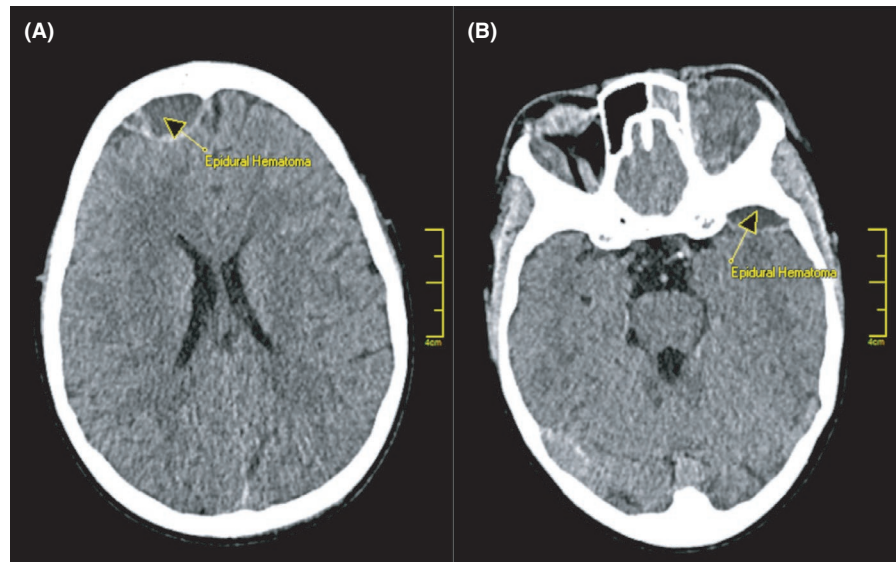
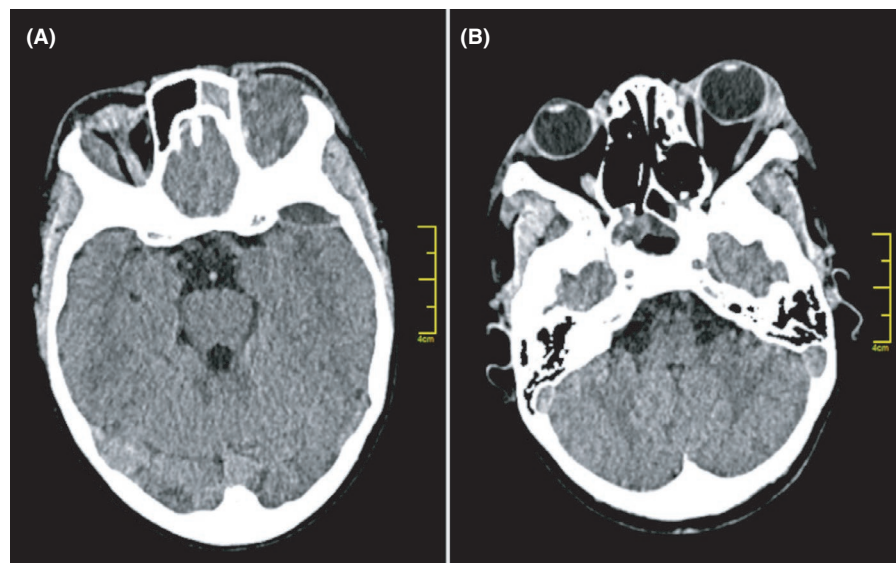


FIGURE 3 A-Axial CT scan showing hypodense lesions in bilateral orbits. The lesion appears larger in the left orbit. B- Proptosis of the left eye



5 | DISCUSSION

Ocular manifestations of SCD have been reported in the literature over the past decades.⁴ These include soft head syndrome,⁷ sub-periosteal orbital hematoma,⁸ orbital bone infarction,⁹ and orbital compression syndrome.^{10–12} Orbital compression syndrome (OCS) is a rare acute complication of SCD.¹³ There is no study reporting the prevalence in SCD, and most studies in the literature have been case reports and case series. The majority of patients are male, aged between 10 and 19 years, with a few isolated cases at 2 years,¹⁴ 8 years,^{5,12} and 22 years of age.¹⁵ In contrast with our case, the patient in Sudan was an 8-year-old female patient but both had HBSS genotype and similar symptoms at presentation with a vaso-occlusive crisis⁵

The presentation is similar to other ocular manifestations of SCD but the diagnosis is made in the presence

of eyelid edema, proptosis, periorbital pain, fever, and ocular or visual disturbances.^{9,10,16} Other examination findings may include hyperemia and eyeball movement limitations.¹⁶ Our patient also presented with several scalp bone swellings consistent with that reported by Alli and colleagues¹⁷ and in Sudan.⁵ Laboratory tests reveal leukocytosis with neutrophilia and raised acute inflammatory markers such as c-reactive protein.^{12,16} Brain imaging shows features of orbital bone infarction and sub-periosteal hemorrhage. Epidural hemorrhage and cephalohematoma have also been reported.¹¹

Treatment of OCS is mainly conservative including adequate hydration, analgesia, antibiotics, and corticosteroids.^{10,12} Methylprednisolone is the most widely used steroid in the literature, and patients showed remarkable clinical improvement as early as 48 h after administration.¹⁵ Blood transfusion can also be administered in cases of severe anemia. The prognosis is usually good with full

recovery of vision in those who initially presented with visual disturbances.¹⁸ The visual acuity in the left eye of our patient fully recovered to 6/24 after 4 weeks compared to 6/60 at admission. This is similar to the patient in Sudan who recovered and was discharged with full recovery of vision within one month.⁵ No fatality has been reported in all cases of OCS in the literature.

6 | CONCLUSIONS

Orbital compression syndrome is a rare, acute ocular manifestation of sickle cell disease. This case highlights the importance of a multidisciplinary team in the prompt diagnosis and management of orbital compression syndrome. Assessment should include a detailed history, ocular exam, and appropriate imaging investigations to exclude other possible differential diagnosis including space-occupying lesions, central nervous system infections, and ocular malignancies such as retinoblastoma. Most cases will resolve on conservative management with analgesics, hydration, empirical antibiotics, corticosteroids, and eye protection. Surgical intervention may be required in case of a large hematoma compressing the optic nerve to prevent visual loss.

ACKNOWLEDGEMENTS

We thank the medical and nursing team at Mulago Hospital Haematology-Oncology Ward (Jelliffe /16A), ENT ward, and the neurosurgery team at Mulago National Referral Hospital who participated in the care of this patient. We also thank the patient and the parent for allowing this case report to be published.

CONFLICT OF INTERESTS

The authors declare no conflict of interests.

AUTHOR CONTRIBUTIONS

RO, CN, SBK, VKM, RN, and DM evaluated and managed the patient. RO collected the patient's information and drafted the initial manuscript. All authors reviewed and approved the final version of the manuscript.

ETHICAL APPROVAL


A written consent and assent were obtained from the parent and the patient to publish these findings and images.

DATA AVAILABILITY STATEMENT

Not applicable. All the data associated with this case report have been described in the manuscript.

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REFERENCES

1. World Health Organization. Sickle cell disease in the African region: current situation and the way forward. 2006. Regional Committee for Africa. Published online 2006:6.
2. Ndeezi G, Kiyaga C, Hernandez AG, et al. Burden of sickle cell trait and disease in the Uganda Sickle Surveillance Study (US3): a cross-sectional study. *Lancet Glob Heal*. 2016;4(3):e195-e200. [https://doi.org/10.1016/s2214-109x\(15\)00288-0](https://doi.org/10.1016/s2214-109x(15)00288-0)
3. Mbiya BM, Kalombo DK, Mukendi YN, et al. Improvement of SCD morbimortality in children: experience in a remote area of an African country. *BMC Health Serv Res*. 2021;21(1):294. <https://doi.org/10.1186/s12913-021-06286-7>
4. Fadugbagbe AO, Gurgel RQ, Mendonça CQ, Cipolotti R, dos Santos AM, Cuevas LE. Ocular manifestations of sickle cell disease. *Ann Trop Paediatr*. 2010;30(1):19-26. 10.1179/146532810X12637745451870
5. Abdelmajeed O, Yassin A, Elamir RM. Multiple scalp infarctions and bilateral orbital compression in a child with sickle cell disease. *Sudan J Med Sci*. 2016;11(1):37-42.
6. Helen OO, Ajite KO, Oyelami OA, Asaleye CM, Adeoye AO. Bilateral orbital infarction and retinal detachment in a previously undiagnosed sickle cell hemoglobinopathy African child. *Niger Med J*. 2013;54(3):200.
7. Al-Ansari RY, Al Harbi M, Al-Jubair N, Abdalla L. Acute soft head syndrome (Subgaleal Haematoma) with periorbital oedema as a rare presentation in sickle cell disease. *Eur J Case Rep Intern Med*. 2020;7(10):1766.
8. Alghamdi A. Recurrent orbital bone sub-periosteal hematoma in sickle cell disease: a case study. *BMC Ophthalmol*. 2018;18(1):211.
9. Janssens C, Claeys L, Maes P, Boiy T, Wojciechowski M. Orbital wall infarction in child with sickle cell disease. *Acta Clin Belg*. 2015;70(6):451-452.
10. Sundu C, Dinç E, Sari A, Ünal S, Dursun Ö. Bilateral subperiosteal hematoma and orbital compression syndrome in sickle cell disease. *J Craniofac Surg*. 2017;28(8):e775-e776.
11. Ilhan N, Acipayam C, Aydogan F, et al. Orbital compression syndrome complicated by epidural hematoma and wide cephalohematoma in a patient with sickle cell disease. *J AAPOS*. 2014;18(2):189-191.
12. Schündeln M, Ringelstein A, Storbeck T, Kocadag K, Grasemann C. Orbital compression syndrome in a child with sickle cell disease. *J Pediatr*. 2014;164(3):671.
13. Ganesh A, William R, Mitra S, et al. Orbital involvement in sickle cell disease: a report of five cases and review literature. *Eye (Lond)*. 2001;15(Pt 6):774-780.
14. Ghafouri RH, Lee I, Freitag SK, Pira TN. Bilateral orbital bone infarction in sickle-cell disease. *Ophthalm Plast Reconstr Surg*. 2011;27(2):e26-e27.
15. Sokol J, Baron E, Lantos G, Kazim M. Orbital compression syndrome in sickle cell disease. *Ophthalm Plast Reconstr Surg*. 2008;24(3):181-184.
16. Douvoyiannis M, Fakioglu E, Litman N. Orbital compression syndrome presenting as orbital cellulitis in a child with sickle cell anemia. *Pediatr Emerg Care*. 2010;26(4):285-286.

17. Alli NA, Wainwright RD, Mackinnon D, Poyiadjis S, Naidu G. Skull bone infarctive crisis and deep vein thrombosis in homozygous sickle cell disease- case report and review of the literature. *Hematology*. 2007;12(2):169-174. <https://doi.org/10.1080/10245330601111912>
18. Procianoy F, Brandão Filho M, Cruz AA, Alencar VM. Subperiosteal hematoma and orbital compression syndrome following minor frontal trauma in sickle cell anemia: case report. *Arq Bras Oftalmol*. 2008;71(2):262-264.

How to cite this article: Olum R, Nabaggala C, Mwebe VK, Namazzi R, Munube D, Kitaka SB. Orbital compression syndrome in a Ugandan child with sickle cell disease: A case report. *Clin Case Rep*. 2021;9:e04766. <https://doi.org/10.1002/ccr3.4766>