Scientific Research Publishing

Open Journal of Ophthalmology, 2019, 9, 134-140
http://www.scirp.org/journal/ojoph
ISSN Online: 2165-7416
ISSN Print: 2165-7408

Retinopathy Revealing Cerebral Venous Thrombosis in Sickle Cell Disease in Niger

Nouhou Diori Adam[1*], Yacoubou Soumana[2], Saley Ali[3], Amadou Moussa Salia[3], Saley Idrissa[4]

[1]Ophthalmology at the University of Lomé, Lomé, Togo
[2]Niamey Clinique Lumiere, Lumiere, Niger
[3]The General Reference Hospital of Niamey, Niamey, Niger
[4]Ophthalmology at the University Hospital Omar Drissi of Fez, Morocco
Email: *adamslalou@gmail.com, ysoumana@yahoo.fr, drsaleyali@gmail.com, moussa.salia@yahoo.fr, idrissasaley787@gmail.com

How to cite this paper: Adam, N.D., Soumana, Y., Ali, S., Salia, A.M. and Idrissa, S. (2019) Retinopathy Revealing Cerebral Venous Thrombosis in Sickle Cell Disease in Niger. *Open Journal of Ophthalmology*, **9**, 134-140.
https://doi.org/10.4236/ojoph.2019.93014

Received: June 4, 2019
Accepted: August 23, 2019
Published: August 26, 2019

Copyright © 2019 by author(s) and Scientific Research Publishing Inc.
This work is licensed under the Creative Commons Attribution International License (CC BY 4.0).
http://creativecommons.org/licenses/by/4.0/

Open Access

Abstract

Cerebral venous thrombosis (CVT) in sickle cell disease has been rarely described in the literature. Some authors consider sickle cell disease as a risk factor for CVT. We report the case of a 20-year-old boy, known as sickle cell, followed at the National Reference Center for Sickle Cell Disease in Niger. Admitted in consultation for a decrease of acuity with the right eye with perception of a black spot evolving since about three (3) days, ophthalmological examination revealed sickle cell retinopathy associated with cerebral venous thrombosis on MRI. The rest of the balance showed normochromic anemia, ionic disturbance and a D-dimer at 1500 µg/l. Rehydration, strict rest, antibiotic therapy and analgesics were started with an internal medicine opinion for further treatment. There was an improvement in visual acuity of 6/10 in 48 hours with disappearance of black spot perception, after three weeks, an ad integrum recovery of retinal lesions to control imaging. Despite the ocular complications of sickle cell disease involving visual prognosis, cerebral venous thrombosis is another complication to be investigated in any sickle cell retinopathy.

Keywords

Sickle Cell Retinopathy, Cerebral Venous Thrombosis, Niger

1. Introduction

Sickle cell disease is the most common hereditary genetic pathology in the world. The gene frequency of the disease is 5% to 20% in Africa and 40% in Central Africa [1]. It is a real public health problem in Niger. The prevalence of

sickle cell trait according to the WHO in 2010 is about 25% in Niger [2]. Niamey being an area located in the sicklemic belt [3] [4].

Retinal involvement is the most common ophthalmological complication of the disease. Peripheral vascular occlusion resulting from sickling of erythrocytes by hemoglobin polymerization is responsible for sickle cell retinopathy [5].

A cerebral venous thrombosis is a rare form of cerebral attack in sickle cell patients which is the most difficult diagnosis [6]. Magnetic resonance imaging allows its diagnosis most often.

We report the case of cerebral venous thrombosis in an SS sickle cell with retinopathy, which has the particularity of the severity of both visual and vital prognosis.

2. Observation

This is a 20-year-old boy, known as Sickle Cell Disease, followed at the National Reference Center for Sickle Cell Disease in Niger. The interview did not find any particular ophthalmological history, metabolic, endocrine or neurological disease. There is a family history of death of her sister sick Sickle cell in a coma table. Admitted to a private clinic (CLINIQUE LUMIERE-NIAMEY) for a visual keenness to the right eye with a perception of a black spot evolving for about three (3) days.

Clinical examination evokes (**Table 1**) sickle cell retinopathy.

Table 1. Sociodemographic and clinical characteristics of the patient.

Sociodemographic data			
Age		20 years	
Sex		Male	
Race		Black	
Profession		student	
Clinical characteristics			
Ophthalmological examination		Right eye	Left eye
Visual acuity	without correction	Count the fingers at (1) meter	10/10
	With correction	Not ameliorable at pinhole	10/10
Slit lamp	Anterior segment	Normal	Normal
	Posterior segment	• Bleeding area in front of salmon-pink macula • Dilation and tortuosity of the vessels • Disappearance of the sharpness of the papillary margin • Hemorrhages in flames.	Normal
Intraocular pressure		19 mm hg	10 mm hg
General review	The rest of the general clinical examination is without peculiarities.		

Retinophotography found a retinal vein occlusion on pre-retinal hemorrhage (Figure 1) associated with fan-like peripheral retinal lesions achieving the classic "sea fan" (Figure 2) was demonstrated after retinophotography and coherence tomography optically objective retro hyaloid hemorrhage with presence of a clot (Figure 3). Angiography could not be performed.

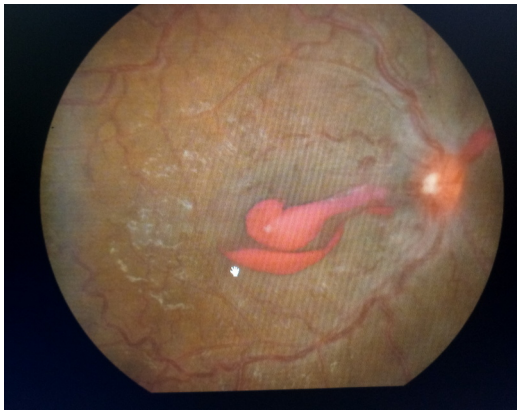

Figure 1. A retinal vein occlusion on pre-retinal hemorrhage right eye.

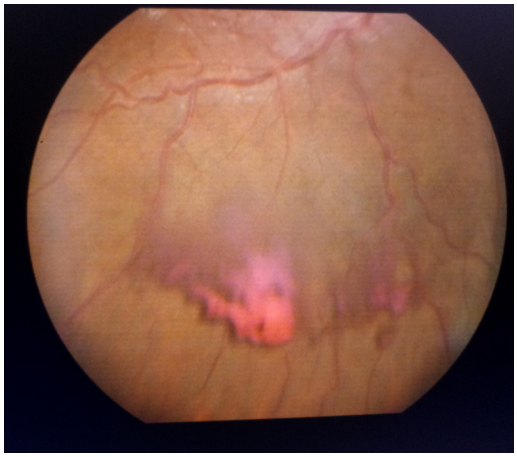

Figure 2. Peripheral retinal lesions achieving the classic "sea fan".

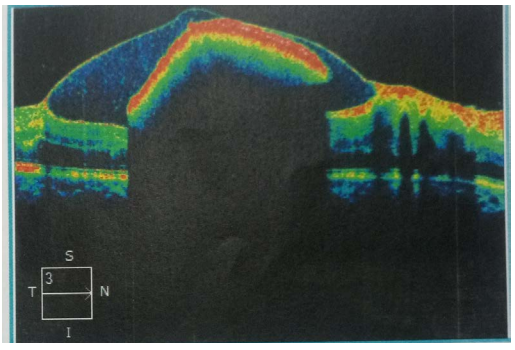

Figure 3. Retro hyaloid hemorrhage with presence of a clot.

A post-operative assessment showed normocytic norno chromium anemia at 6.5 a/l, hematocrit at 18%; a sodium level of 135 Méq; kaliemia: 4 Méq; TP/TCK are normal, D-dimer: 1500 ug/l.

An MRI revealed partial sagittal sinus thrombosis in the Torcular with left lateral fresh blood (Figure 4 and Figure 5).

Rehydration based on 3 liters Saline-Glucose-Ringer serum, combined with a strict rest in bed was started. Ceftriaxone 100 mg/kg/day, paracetamol 60 mg/kg/day were administered. An internal medicine opinion was requested for the continuation of the care.

The clinical course in 48 hours was marked by an improvement of the visual acuity which passed to 6/10 without correction with disappearance of the perception of the black spot. Ocular control imaging after after three weeks showed recovery ad integrum Figure 6 and Figure 7.

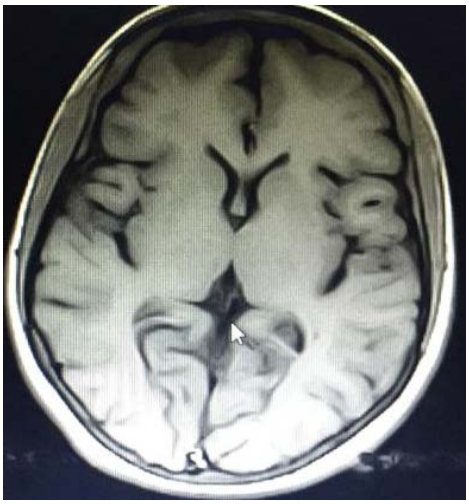

Figure 4. Partial sagittal sinus thrombosis at the level of the Torcular.

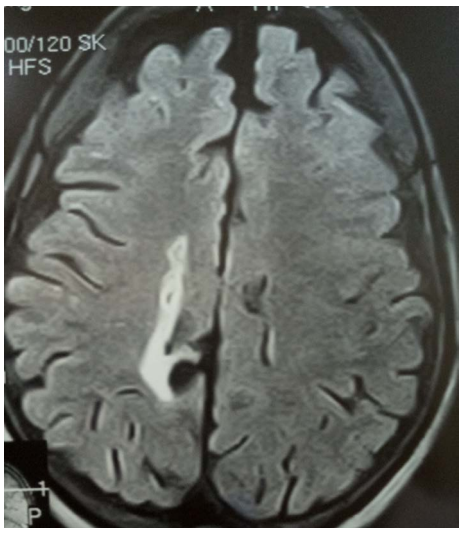

Figure 5. Lateralized fresh blood on the left.

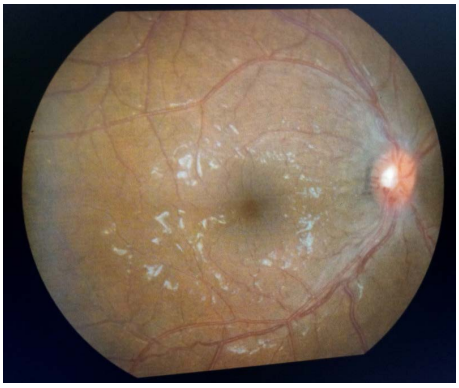

Figure 6. Control retinography after a few weeks.

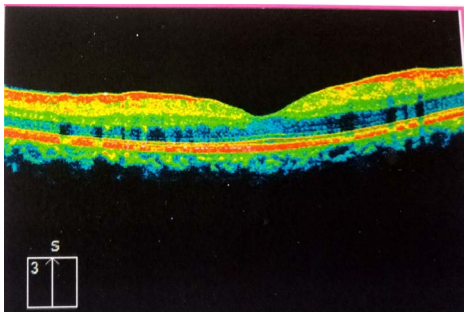

Figure 7. OCT control after a tree weeks.

3. Discussion

Sickle cell disease is the most common hereditary genetic disorder in the world and especially in Africa. In Niger, it presents a public health problem with a prevalence of hemoglobin S (HbS) carriage of 25% [7].

Ophthalmologic involvement with variable gravity according to age and type of sickle cell disease is progressing insidiously, for a long time without any functional signs, and exposes to sometimes serious complications [8].

The functional symptomatology is absent until an advanced stage of the disease, where the reduction of visual acuity then represents a serious complication. Sickle cell retinopathy affects young people and its diagnosis is based on a systematic examination of the fundus Bio microscopy and angiography [1] [9] [10]. The sudden drop in visual acuity was the revealing mode of sickle-cell retinopathy in our patient.

Cerebral venous thrombosis (CVT) is a rare disease affecting about 5 people per million per year with huge regional variations [11]. Cerebral venous thrombosis has a broad spectrum of clinical manifestations and modes of onset that can mimic many other neurological disorders and lead to frequent misdiagnosis or delayed diagnosis [12]. Some authors in Senegal have also found that sickle cell disease is a risk factor for vascular thrombosis but the mechanism could not be clarified [13]. It is a pathology very little described in the African literature. Lalya F. *et al*., there has been one case of death following a CVT in sickle cell pa-

tients in 2017 in Cotonou [6]. It should be noted that a family death in a coma context was reported by the parents of the patient of one of the sisters also sickle cell. This proves the seriousness of the disease.

4. Conclusion

In spite of ocular complications of sickle cell disease involving visual prognosis, cerebral venous thrombosis is another complication to look for in any sickle cell retinopathy. His clinical picture is polymorphic and nonspecific, medical imaging to improve his diagnosis.

Contributions of the Authors

All authors state that they have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflict of interest.

References

[1] Balo, K.P., Ségbana, K., Mensah, A., Mihluedo, H. and Bechetoille, A. (1996) Hémoglobinopathies et rétinopathies au CHU de Lomé. *Journal Français D'Ophtalmologie*, **19**, 497-504.

[2] Organisation Mondiale de la Santé (2010) Programme Maladies Non Transmissibles, y compris les affections buccodentaires et la Santé mentale. Rapport annuel OMS-Niger 20.

[3] Livingstone, F.B. (1967) Abnormal Hemoglobins in Human Populations. Aldine Publishing Company, Chicago, IL, 482.

[4] Sergeant, G.R., Higgs, D.R., Aldridge, B., Hayes, R.J. and Weatherall, D.J. (1981) Alpha Thalassemia and Homozygous Sickle Cell Disease. *Progress in Clinical and Biological Research*, **55**, 781-788.

[5] Binaghi, M. and Levy, C. (1993) Eil et hémoglobinopathies. Encyclopedie Medico-chirurgicale. Ophtalmologie, 21-452-G-20. 7 p.

[6] Lalya, F., d'Almeida, M., Yevedo, T. and Biaou, O. (2017) Thrombose veineuse cérébrale chez un enfant drépanocytaire. *Journal africain de pédiatrie et de génétique médicale*, No. 1, 57-60.

[7] Malam-Abdou, B., Mahamadou, S., Brah, S., *et al.* (2016) Hemoglobinopathies in Niger. A Review of 6532 Electrophoresis of the Biochemistry Laboratory of the Faculty of Medicine of Niamey. *Health Sciences and Diseases*, **7**, 93-97.

[8] Fanny, A., Coulibaly, F., Gbe, K., Meite, M., Adjorlolo, C., Konan-Toure, M.L., *et al.* (2005) Les bêtathalasso-drépanocytoses pourvoyeuses de rétinopathies ischémiques graves. *Journal Français D'Ophtalmologie*, **28**, 391-395. https://doi.org/10.1016/S0181-5512(05)81070-8

[9] Morel, C. (2001) Atteinte rétinienne des hémoglobinopathies. *Journal Français D'Ophtalmologie*, **24**, 987-992.

[10] Balo, K.P., Ségbana, K., Mensah, A., Djagnikpo, P., Mihluedo, H., Adjivon, K., *et al.* (1995) Étude des complications rétiniennes au cours des hémoglobinopathies: A propos de 32 cas. *Médecine Tropicale*, **55**, 450-453.

[11] Brousser, M.G. and Ferro, J.M. (2007) Cerebral Venous Thrombosis: An Update. *The Lancet Neurology*, **6**, 162-170. https://doi.org/10.1016/S1474-4422(07)70029-7

[12] Leys, D. and Cordonnier, C. (2008) Cérébral Venous Thrombosis: Update on Clinical Manifestations, Diagnosis and Management. *Annals of Indian Academy of Neurology*, **11**, 79-87.

[13] Fall, A.O., Proulle, V., Sall, A., Mbaye, A., Ba, P.S., Diao, M., *et al.* (2014) Risk Factors for Thrombosis in an African Population. *Clinical Medicine Insights*: *Blood Disorders*, **7**, 1-6. https://doi.org/10.4137/CMBD.S13401