

Spontaneous bilateral intra-orbital hematoma: A particular form of complications of sickle cell disease in children.

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January 23, 2022

Abstract

Spontaneous bilateral intra-orbital hematoma is a rare complication of sickle cell disease in children. Imaging examinations are of paramount importance in the diagnosis and condition the management in order to avoid complications that can compromise the visual function prognosis.

INTRODUCTION

Sickle cell disease is an inherited hemoglobinopathy characterized by vaso-occlusive crises. Intraorbital hematoma secondary to hematopoietic medullary infarction is a rare complication of sickle cell disease [1]. The visual prognosis is at stake in the absence of immediate treatment. We report an atypical form of intraorbital hematoma revealed by bilateral exophthalmos in a sickle cell child, then we will highlight the role of imaging in the diagnosis and management of intraorbital hematoma.

CASE REPORT

A 15 year old boy, known to have homozygous sickle cell disease, came for a consultation with headache and bilateral eye pain which had been evolving for two weeks in a context of apyrexia. In his background, there was no notion of craniofacial traumatism or recent surgery. On ophthalmological examination; bilateral diplopia, palpebral edema and bilateral exophthalmos painful, non-pulsatile and non-reducible were noted. The measured visual acuity was altered to 5/10 for the right eye and 7/10 for the left eye. The fundus examination was very limited and did not show any particularities. Other examinations, especially neurological, were unremarkable. The formal blood count showed a discrete anemia with a hemoglobin level of 9.3 g/dl; the neutrophilic polynuclear and platelet counts were within the normal range. The C-reactive protein level was discretely elevated at 7 mg/L. The hemostasis assessment was also normal. Oculo-cerebral CT scan with parenchymal window showed a bilateral intraorbital structure, spontaneously hyperdense, of hematic density, in intra and extra-conical situations, pushing forward and down the two ocular globes (figure 1). In the bone window, it showed a bilateral exophthalmos of grade 3 without abnormal bone structure (Figure 2). Ocular B-mode ultrasound was performed but was very limited because of the narrowing of the acoustic window due to the exophthalmos. In front of the anamnestic, clinical and scannographic context, the diagnosis of a

spontaneous bilateral intraorbital hematoma in a sickle cell patient was evoked. The patient was put on oral corticosteroids for five days. And in front of the ocular pain, a treatment containing analgesic was instituted. The evolution was favorable after one week, marked by the regression of the exophthalmos and a progressive recuperation of visual acuity and oculomotricity. The Oculocerebral CT scan at one week showed regression of the intraorbital hematoma (**Figure 3**). The recuperation of visual acuity and oculomotricity ad integrum was achieved after one month with total disappearance of the exophthalmos (**Figure 4**).The patient was then readmitted to the sickle cell management center.

DISCUSSION

In the orbit; the hematopoietic medullary space is smaller, It is therefore rare to find orbital bone infarcts in sickle cell patients [1]. About thirty cases are described in the literature with an average age of 14 years , half of which had an intraorbital hematoma [2]. The singleness of our case in Madagascar would therefore allow comparison with the literature and could provide additional data on the diagnosis and management of intraorbital hematoma in children with sickle cell disease. The bilateral form is rarely reported in the literature. In a series conducted by Soko, the bilateral form represents one third of reported cases [2]. This one makes our case a particular form of intraorbital hematoma. Exophthalmos is a reflection of the mass effect, it is the main mode of discovery of the intra-orbital hematoma. The first functional manifestations are ocular pain and localized edema. In the absence of urgent management, the visual prognosis is at risk in case of optic nerve compression [1,3]. Before the rapid onset of exophthalmos, especially since it is bilateral; the differential diagnosis is vast. Among these, we can cite orbital pseudotumor, orbital rhabdomyosarcoma, optic nerve glioma, orbital extension of bilateral retinoblastoma, bilateral orbital cellulitis. In all these possibilities, Imaging studies are of paramount importance in characterizing the nature of the compressive mass. In our case, the hematic density was spontaneously hyperdense on the CT scan, suggesting the diagnosis of a bilateral intra-orbital hematoma. The Magnetic resonance imaging, which would determine the hematic nature of the collection [4] not being accessible, the CT scan therefore remains an examination that retains its place although it is relatively non-specific. However, the absence of sinus pathology and the location of the collection on the lateral wall of the orbit are the arguments against an abscess [2] especially that the clinical and biological examination are sterile. Medical treatment seems to be sufficient in most cases, including treatment of the crisis and the use by some teams of corticosteroids to be used with caution in the context of sickle cell disease [1,2]. Surgery is reserved for cases of persistent compression of the optic nerve [2]. In our case, the treatment is purely medical, making the exophthalmos disappear after one month with a recovery of the visual acuity ad integrum.

CONCLUSION

Spontaneous bilateral intra-orbital hematoma secondary to bone infarction is a rare but serious complication of sickle cell disease. Imaging examinations hold an important place in the diagnosis and have an implication in the management to preserve the visual function. A regular follow-up of sickle cell disease with the implementation of a background treatment and the precaution of factors favoring vaso-occlusive attacks could avoid this complication which sometimes has a poor prognosis.

KEY CLINICAL MESSAGE

The report of this case would allow researchers to compare with the literature and could provide additional data on the diagnosis and management of intraorbital hematoma in children with sickle cell disease.

ACKNOWLEDGEMENTS

This work was carried out with the working group of SIMAD (Société d’Imagerie de Madagascar).The authors thank the patient who gave written permission for this report.

AUTHOR’S CONTRIBUTIONS

All the authors have read and approved the final draft of the manuscript. All authors contributed to this work and approved the final version.

HA: followed up the patient, collected the clinical data.

HR: Collected the clinical data, and drafted the report

KR: followed up the patient, collected the clinical data.

PHR: followed up the patient

EPGA, LHNONR: designed and critically revised the report.

JLR, HMDV and AA: validated the report.

CONFLICTS OF INTEREST

None declared

ETHICAL APPROVAL

The article does not contain any personal information that could identify the patient. The names and dates on the chest CT scan have been hidden. The authors have included only information necessary for scientific understanding.

FINDING STATEMENT

This research has not received any specific grant from any public, commercial or for-profit.

DATA AVAILABLE STATEMENT

Data available on request from the corresponding author.

CONSENTS STATEMENTS

Written consent was obtained from the patient for publication of this case report and accompanying images.

REFERENCES

- [1] Ganesh A, Al-Zuhaibi S, Pathare A, William R, Al-Senawi R, AlMujaini A, et al. Orbital infarction in sickle cell disease. *Am J Ophthalmol* 2008;146(4):595–601.
- [2] Sokol JA, Baron E, Lantos G, Kazim M. Orbital compression syndrome in sickle cell disease. *Ophthalmic Plast Reconstr Surg* 2008;24(3):181–4.
- [3] Talbot M, Meunier B, Lavoipierre V. L'hématome sous-périoste, une complication méconnue de la drépanocytose. *Rev med int. Volume 40, Supplément 1* ; 2019 : 159-60. <https://doi.org/10.1016/j.revmed.2019.03.207>
- [4] Tostivint L, Pop-Jora D, Grimprel E, Quinet B, Lesprit E. Orbital bone infarction in a child with homozygous sickle cell disease. *Arch Pediatr* 2012;19(6):612–5.

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Figure 1: Oculo-cerebral CT scan with parenchymal window: Bilateral intraorbital formation, spontaneously hyperdense, intra and extra-conical situation, hematic density.

Figure 2 : Oculo-cerebral CT scan with bone window: Exophthalmia stage 3 , with absence of abnormalities in bone structure.

Figure 3: Oculo-cerebral CT scan with parenchymal window: Regression of the intraorbital hematoma

Figure 4 : Oculo-cerebral CT scan with bone window: Total disappearance of exophthalmos







