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CASE REPORT

Congenital Glaucoma: Thinking about Sturge-Weber-Krabbe Syndrome

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ABSTRACT
Congenital glaucoma is called "primary" if it is due to an isolated anomaly of the development of the iridocorneal angle. It is secondary when associated with other lesions, ocular or systemic. Phacomotoses present 50% to 70% of the etiologies of childhood Glaucoma, including Sturge Weber Krabbe syndrome (SWKS). We report the case of an infant admitted for a clinical picture made of angioma with seizures associated with glaucoma evoking an SSWK to remind practitioners of the clinical, radiological and especially evolutionary aspects of this disease entity. Childhood Glaucoma is a rare disease that occurs from birth to adolescence. This usually results in vision loss. About 8% of cases of blindness have been attributed to this disease in the pediatric population.

KEYWORDS: Congenital Glaucoma; Sturge-Weber-Krabbe Syndrome

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INTRODUCTION

Congenital Childhood Glaucoma is classified as "primary" if it is due to an isolated anomaly in the development of the iridocorneal angle and is secondary when associated with other ocular or systemic lesions.
Phacomatoses are characterized by hamartomas, which Sturge-Weber syndrome has a frequency of about 1 in 50.000 births.

CLINICAL OBSERVATION

An infant of 04 months, without particular antecedent notably no notion of epilepsy, or consanguinity with a good psychomotor development. He was admit for partial convulsion of the right hemi-body during the examination, we found a conscious infant with a cutaneous angioma occupying the right half of the face (Figure 1).
A cerebral CT showing diffuse atrophy of the right cerebral hemisphere, seat of fine calcification under low cortical, a leptomeningeal enhancement of the entire hemisphere in relation to a pial angioma, hypertrophy of the two choroid plexuses predominant on the right, on the left a less important leptomeningeal enhancement of the temporo-occipital (Figure 2,3).
An ophthalmological examination was requested in front of the clinical and radiological data showing glaucoma of the right eye.
The diagnosis of Sturge-Weber syndrome is retained in front of the clinical and radiological picture, and the child is put under antiepileptic, a treatment for glaucoma and followed in consultation.

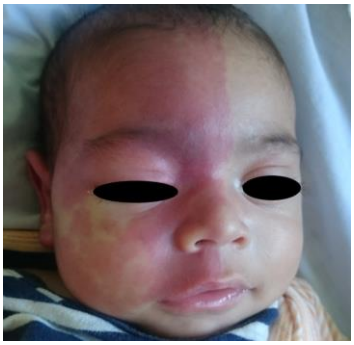

Figure n° 1: A cutaneous angioma occupying the right half of the face.

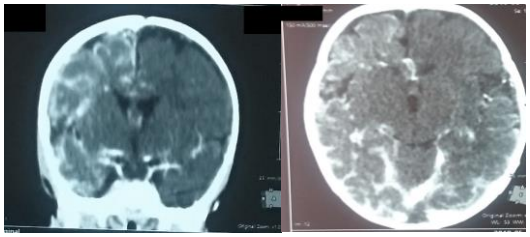

Figure n° 2 & 3: A cerebral CT showing diffuse atrophy of the right cerebral hemisphere, seat of fine calcification under low cortical, a leptomeningeal enhancement of the entire hemisphere in relation to a pial angioma.

DISCUSSION

Sturge-Weber Krabbe syndrome is a neurocutaneous and ocular phacomatosis with very rare vascular malformative substratum with a prevalence estimated between 1/20000 and 1/50 000 births [2], associating a cutaneous angioma of the trigeminal pathway, ophthalmic and a neurological affect [3].

The cutaneous angioma can be either absent, unilateral extended beyond the territory of the trigeminal nerve or bilateral, if the plane angioma reaches the forehead, the risk of neurological and ophthalmological complications is higher [4].

The main ocular manifestation is glaucoma, wich may be precocious or late, it is present with a prevalence estimated between 30 and 60%. The risk of having glaucoma increases with facial angioma that interest the eyelids [2,5].

The neurological damage should be sought in any patient with a hemiface plane angioma. Pial angioma is most often homolateral to cutaneous angioma [2]. In our case, the infant has convulsive seizures of the right hemi-body, a cerebral CT showing diffuse atrophy of the entire right cerebral hemisphere with pial angioma. Currently, MRI is the best test to look for brain damage, even before the clinic, this examination must be performed in the neonatal period and must be repeated after one year if it is normal in the neonatal period [2,6].

Epilepsy is the main neurological complication of SWKS that appears in the first two years of life.

In our case, the combination of cutaneous involvement and epileptic seizures indicated an ophthalmic examination and brain imaging.

The risk of developing glaucoma is important in the first 10 years of life, but some patients maintain normal intraocular pressure until adulthood, hence the importance of close monitoring. It is recommended to have an eye exam every three months, during the first year of life. Sometimes anesthesia is needed to measure the intraocular pressure. After the first year, the exams are spaced 6 months apart, to become annual [2].

Therapeutically, skin involvement require the pulsed laser.wich can induce or aggravate glaucoma by increasing intraocular pressure by decreasing the episcleral venous network [2].

The usual antiepileptic treatment is generally sufficient, sometimes with refractory epilepsy, a surgical treatment is to be considered, with in the most unfavorable cases a hemispherectomy is discussed [2,4].

The initial treatment of glaucoma is based on beta-blockers and carbonic anhydrase inhibitors, which act as aqueous suppressors. On the other hand, the use of prostaglandin analogs is contraindicated, since cases of cilio-choroidal effusion have been reported in patients with Sturge Weber syndrome.

In the long term, medical treatments will become insufficient in most cases, with recourse to surgical treatment: trabeculectomy and goniotomy with a success rate of 70%.

A technique combining trabeculoplasty and trabulectomy is also used in Sturge Weber syndrome. Other approaches include placement of an aqueous drainage device, filtration surgery and destruction of the ciliary body.

CONCLUSION

SWKS is rare but must be quickly identified because it requires early and multidisciplinary management. As soon as the diagnosis is suspected, an ophthalmological examination and a brain MRI must be performed. Ophthalmological monitoring is more important since various complications involving visual prognosis can occur throughout childhood, specially in the first year of life when the child does not know how to complain about a decrease in visual acuity. Glaucoma is an ophthalmologic complication that is difficult to treat.

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AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript, and provided approval for this final revised version.

COMPETING INTERESTS

The authors declare no competing interests with this case.

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None.

PATIENTS CONSENT

Written informed consents were obtained from the parents for the publication of this case report.

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