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Congenital Glaucoma in Sturge-Weber Syndrome

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Abstract **Case Report**

The case study of Sturge-Weber syndrome highlights the intricate management required for congenital glaucoma. This rare neurocutaneous condition presents challenges due to its varied clinical manifestations, including facial cutaneous angiomas and potential eye involvement leading to glaucoma. The onset of glaucoma in Sturge-Weber syndrome can vary significantly, occurring from birth to adulthood, with different underlying mechanisms proposed by researchers. These mechanisms include dysgenesis of the anterior chamber and increased episcleral venous pressure, contributing to elevated intraocular pressure and subsequent glaucoma development. A multidisciplinary approach is essential to improve the patient's visual prognosis and quality of life, emphasizing the importance of personalized management of this rare disease.

Keywords: Glaucoma, struge weber, congenital, syndrome, angioma.

INTRODUCTION

Sturge-Weber syndrome is a rare congenital neurocutaneous condition characterized by three main symptoms: skin angiomas present on the territory of V1 branching in the face, meningeal brain hemangioma, and possible eye involvement, especially the development of glaucoma. The eye has increased pressure, causing glaucoma, and it is particularly significant as this may produce visual complications.

This disease is mainly diagnosed clinically. Unlike the ailment, the treatment of Sturge-Weber syndrome need multidisciplinary management such as medical neurologists and ophthalmologists.

OBSERVATION

We report the case of a 6-month-old infant, with no particular pathological history, who has presented since birth with a facial cutaneous angioma, with exophthalmos of the right eye and megalocornea.

Ophthalmological examination of the child under general anesthesia revealed: right conjunctival hyperemia; slight right corneal edema; right buphthalmia (corneal diameter 13 mm on the right and 10 mm on the left); right ocular hypertonia (intraocular pressure 20 mmHg on the right and 11 mmHg on the left); Papillary atrophy on the right; Papillary excavation of 0.1 on the left. General examination revealed an erythematous patch on the child's right hemiface in the ophthalmic (V1) and maxillary (V2) territory of the trigeminal nerve. This erythematous patch was non-infiltrated, with a smooth surface, and was noted by the mother from birth. This was a planar angioma of the face. B-mode ultrasound was normal. Magnetic resonance imaging (MRI) to look for cerebral involvement was normal.

The diagnosis of Sturge-Weber syndrome type 2 was made in view of the presence of an angioma affecting the face without cerebral involvement and the presence of congenital glaucoma.

Treatment consisted of surgical abstinence and medical management with local antihypertensive agents and beta-blockers with follow-up.

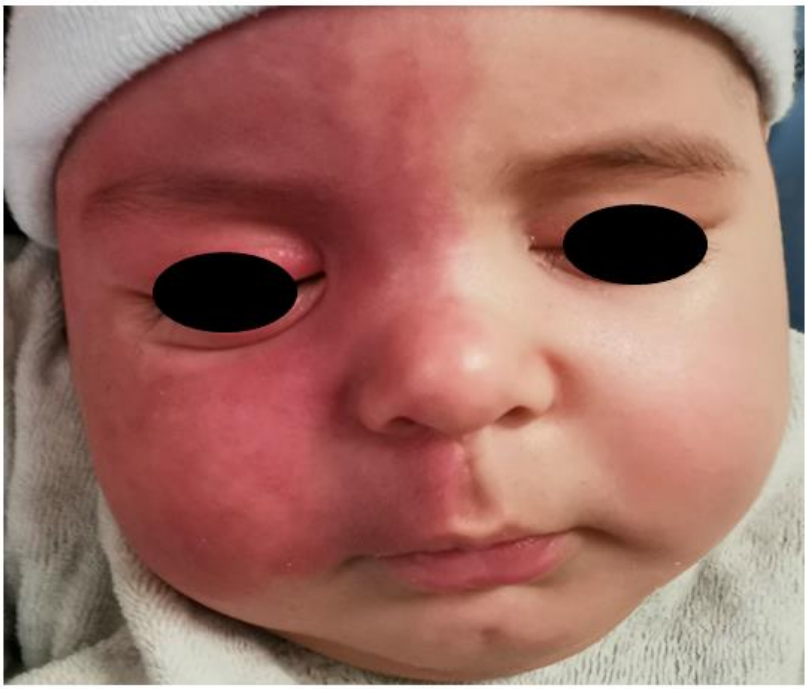

Figure 1: Image Showing Facial Angioma

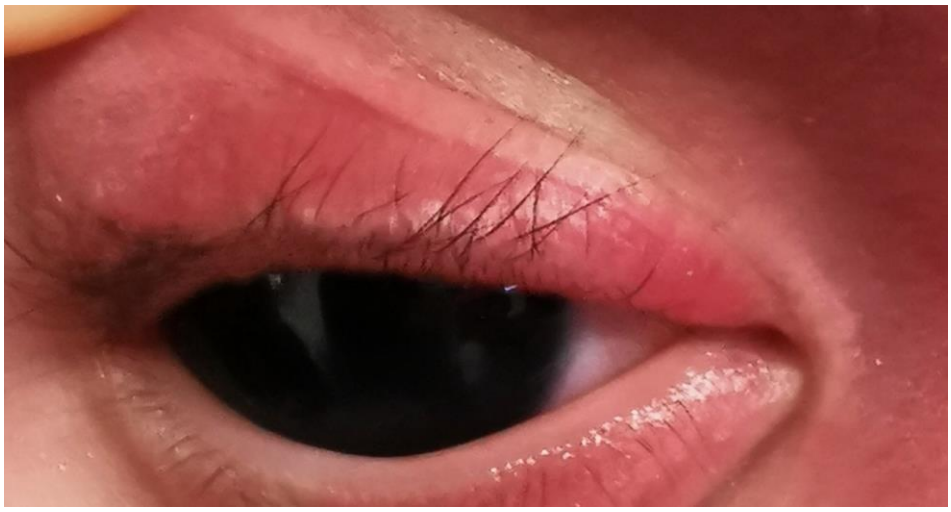

Figure 2: Image showing what megalocornee looks like

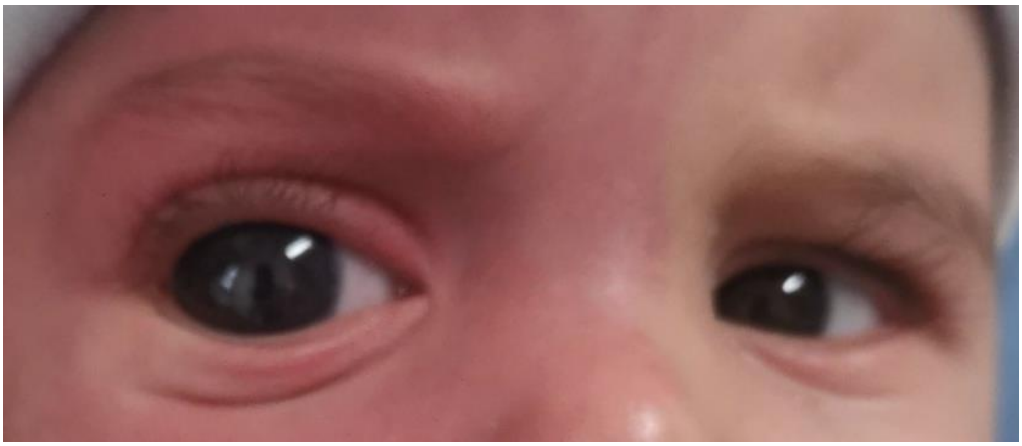

Figure 3: Image showing the difference between the right and left eye with the presence of the angioma and megalocorne on the right

DISCUSSION

The prevalence of this congenital disease is estimated at around 1 case per 10,000 live births worldwide. In France, the prevalence is lower, at just one case per 100,000 births. In Morocco, there are as yet no exact figures for these patients, but the condition affects both sexes equally and is found in a variety of ethnic groups [1]. The disease can sometimes present exceptional familial cases, such as identical twins, although it is generally rare [2].

Glaucoma is frequently observed in individuals with Sturge-Weber syndrome, affecting between 30 and 70% of patients. It can occur either unilaterally or bilaterally. In particular, patients with a facial plane angioma on the upper eyelid appear to be predisposed to this complication [3].

Several hypotheses have been put forward to explain the onset of glaucoma in Sturge-Weber syndrome. Dysgenesis of the anterior chamber and increased episcleral venous pressure are the two theories accepted by most authors [4].

The onset of glaucoma varies considerably, from birth to age 41. Research has shown that 60% of individuals develop glaucoma in early childhood, often characterized by buphthalmia and myopia, while 40% develop it later, in childhood or adulthood [5].

With regard to the origin of glaucoma in SWS, two main hypotheses have been put forward. On the one hand, defects in the angle of the anterior chamber of the eye lead to increased resistance to the outflow of aqueous humor, resulting in an increase in eye pressure, mainly in infants. On the other hand, in individuals with late-onset glaucoma, increased pressure in the episcleral veins is observed, attributed to arteriovenous shunts within the episcleral angioma [6].

Other lesions that may be observed include choroidal angioma and conjunctival and episcleral angioma. Other less frequent manifestations include iris heterochromia, retinal detachment, pigmentation of the sclera, epileptic nystagmus, and angioma of the orbit [7].

The management of glaucoma in Sturge-Weber syndrome presents a therapeutic challenge. Beta-blockers are the most commonly used drugs, although they are not approved for pediatric use. Prostaglandin analogues are less widely used. Latanoprost and travoprost are the most widely used, given the rarity of adverse effects associated with this class of drugs.

Surgical treatment remains the arsenal of treatment for congenital glaucoma during SWS and is associated with medical treatment, but the choice of technique remains a subject of debate among authors [8].

CONCLUSION

The management of congenital glaucoma associated with Sturge-Weber syndrome remains difficult, between the choice of first-line medical treatment or the combination of medical and surgical treatment. The visual prognosis remains poor, and the management of the syndrome is multidisciplinary.

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