STURGE-WEBER SYNDROME-INDUCED GLAUCOMA

On gonioscopy, the angle structures appear indistinct, with a high iris insertion. An anteriorly displaced iris root, under- developed scleral spur, and thickened uveal meshwork have been observed on histopathological specimens. SWS patients with early-onset glaucoma present with typical signs of congenital glaucoma, including buphthalmos, anisometropia, or amblyopia. Congenital angle abnormality, episcleral haemangiomas and elevated episcleral venous pressure can be responsible for early-onset glaucoma, whereas elevated episcleral venous pressure is the main mechanism underlying late-onset glaucoma

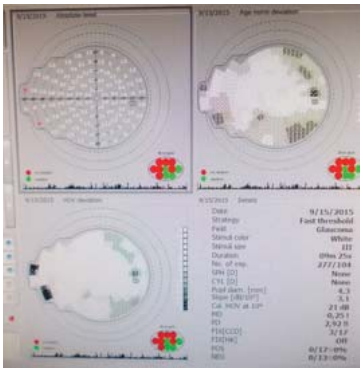

Figure 3.Visual field of the right eye showing early glaucomatous defects (superior and inferior scotomas as well as enlargement of the blind spot).

MANAGEMENT

Managing glaucoma in patients with SWS is complex because of early onset and resistance to conventional therapy. For early onset glaucoma with associated angle abnormalities, surgical intervention is usually required, with either goniotomy or trabeculotomy. However, the outcomes are generally worse than those observed with primary congenital glaucoma, and these cases often require additional surgery with trabeculectomy or a glaucoma drainage device. In SWS, the Ahmed-type valve has shown to induce long-term decrease of IOP by improving aqueous humor outflow. Other surgical approaches include filtering procedures such as trabeculectomy, posterior lip sclerotomy,and trabeculotomy–trabeculectomy, which bypass the alterated trabecular meshwork–Schlemm's canal complex, creating an alternative outflow passage for the aqueous humor, independent to the distal episcleral veins. However, filtering procedures have been associated with severe complications such as expulsive choroidal haemorrhage, bleeding, prolonged flat anterior chamber, and high risk of bleb failure. Intra-operative use of antimitotic agents such as mitomycin does not improve the outcome.Therefore, combined procedures such as trabeculotomy–trabeculectomy have been suggested as the first-line approach in infants and children. Another treatment option is cyclophotocoagulation (CPC) which can be performed in patients with refractory glaucoma or in patients who are likely to have much higher risk of intra-operative or post-operative complications (choroidal expulsive haemorrhage or choroidal detachment) after glaucoma filtration surgery. Alternative surgical procedures include non-penetrating sclerectomy, valve drainage implants, and ciliodestructive procedures in adults. Non-penetrating sclerotomy has been shown to have an efficacy similar to trabeculectomy in controlling SWS-related glaucoma, with a lower rate of complications. However, in SWS patients, the presence of episcleral hemangioma and angle malformations make this procedure extremely difficult and increase the rate of failure.

For late onset glaucoma, topical medications are initiated first. If topical medication does not produce an adequate response, glaucoma filtration surgery remains the procedure of choice for late-onset cases because it bypasses the episcleral venous system.

The presenting IOPs in this patient was 19.5 mmHg in the right eye and 11mmHg in the left eye. Glaucomatous changes (vessel baring, nasal displacement of vessels, superior and inferior rim notching) where seen in the right eye only. Lumigan nocte was started for the right eye only. After 2 months, the IOP in the right eye was 16.5mmHg and 11mmHg in the left eye. Since I was not happy with the IOP reduction, Timolol 0.5% bid was added OD. After a month, the IOP was 12mmHg in the right eye and 11mmHg in the left eye. This IOP target was acceptable.

CLINICAL PEARLS

- In SWS, the haematomatous haemangiomas can be found on the face, eyelid (increases risk of glaucoma in 30-70% of patients with SWS), choroid, and brain. These lesions are not malignant, but will cause local problems leading to glaucoma and seizures. Of the systemic haematosis, SWS has the highest incidence of glaucoma. Approximately 60% of patients present with glaucoma at birth and 40% manifest glaucoma later in life. The incidence of glaucoma increases when the PWS involves the eyelid. It presents most often ipsilateral to the PWS but can also manifest bilaterally. The classical clinical signs of SWS are present, consisting of unilateral facial PWS along the first branch of the trigeminal nerve, hemiatrophy, progressive seizures, contralateral hemiparesis, mental deficiencies, hemianopia, and ipsilateral glaucoma.
- Congenital glaucoma patients with SWS have the following associated with glaucoma: photophobia, epiphora, blepharospasm, Haab striae, large corneas, corneal clouding, dilated and tortuous episcleral vessels.
- Glaucoma associated with SWS is generally more difficult to manage than other forms of glaucoma, with a lower success rate and an increased risk of surgical complications. The prominent role played by increased episcleral venous pressure may be responsible.
- The current dermatological management of PWS is treatment with a pulsed dye laser. Usually, early intervention and multiple treatments are required to lighten the skin discoloration and enhance cosmetic outcomes.
- Leptomeningeal capillary-venous malformation on the same side as the PWS occurs in 10 to 20 percent of newborns with a typical PWS. However, the risk of intracranial lesions increases with the size and bilateral presence of the PWS. When the leptomeningeal lesions are bilateral, patients usually have more severe neurological manifestations and a worse prognosis.
- The most common neurologic complication of SWS is epilepsy, which occurs in 75 percent and 95 percent of patients with unilateral or bilateral brain involvement, respectively. Seizures usually occur within the first year of life but may appear at any age. These may manifest with any acute illness or may accompany the hemiparesis that occurs contralateral to the site of CNS involvement. Seizures may be focal motor, complex partial, or atonic. Permanent hemiplegia and homonymous hemianopia may develop as a consequence of epilepsy. Seizures may be controlled with anticonvulsant therapy in 40 percent of patients.
- Developmental delay, learning disabilities, and severe headaches commonly occur in children with SWS. These children also experience social and behavioural difficulties. Mental retardation may occur in up to 60 percent of all patients with SWS.
- Patients with SWS have a lower life expectancy than does the general population.

CONCLUSION

Sturge Weber Syndrome is a rare, sporadic disorder involving vasculature in the facial skin, CNS, and eye. The main ocular manifestation is glaucoma, in which developmental anomaly of the anterior chamber angle and elevated episcleral venous pressure are underlying mechanisms.Treatment of glaucoma requires an understanding of underlying pathology and mechanisms to guide appropriate pharmacological and surgical intervention.

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