Advanced Glaucoma in a Patient with Facial Port Wine Stains

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Abstract
This a typical case of nevus flammeus with hemifacial hypertrophy and intractable glaucoma.

Keywords: Glaucoma; Facial Port Wine Stains; Nevus Flammeus; Sturge-Weber Syndrome (SWS)

Introduction
Nevus flammeus also known as port-wine stain can be a finding of Sturge-Weber Syndrome (SWS), a neurocutaneous disorder; it is a congenital facial cutaneous vascular malformation that classically affects the ophthalmic (V1) and maxillary (V2) dermatomes of Trigeminal nerve. Intracranial leptomeningeal angiomatosis is the vascular anomaly, seen in SWS [1].

SWS can lead to the development of glaucoma. Improper irido-corneal angle, vascular malformations of the trabecular meshwork and increased pressure from tortuous episcleral vessels are suggested pathophysiological mechanisms [2-4].

Case Presentation and Discussion
A 72-year-old man presented to an ophthalmology clinic with complete vision loss in his right eye as a result of intractable glaucoma. His right eye vision was no light perception and the left eye 20/30 with an intraocular pressures of 35 and 18 mmHg in right and left eye (Goldman applanation tonometry) respectively. In slit lamp exam conjunctival injection and hyperemia with diffusely cloudy cornea noted, left eye slit lam exam was not remarkable. Fundus view was not available in right eye due to severe media opacity, in the left eye fundus was unremarkable and appropriate for age. His right eye had previously required multiple surgeries including trabeculectomies and cyclocryodestruction as well as medications such as topical Timolol, Dorzolamide and Brimonidine. On physical exam of his face, the patient had a large, purple-red discoloration affecting the right side of his face consistent with a nevus flammeus (Figure 1a).

The magnetic resonance imaging (MRI) of this patient revealed extensive hypertrophy of the soft-tissues on the right side of the face and head (Figure 1b).

The patient’s advanced stage of glaucoma caused irreversible damage to his vision along with opacification of the eye. If pain due to high intraocular pressure is severe, then methods of fluid drainage can be considered.

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*Figure 1a:* Large, purple-red discoloration affecting the right side of the patient, known as a port-wine stain.

*Figure 1b:* Magnetic resonance imaging (MRI) of his brain, showing extensive hypertrophy of the soft-tissues on the right side of the face and head.

Conclusion

Nevus flammeus also known as a port-wine stain may be finding of Sturge-Weber Syndrome (SWS), which may cause intractable glaucoma with severe vision loss and leptomeningeal angiomatosis which mandates central nervous system imaging.

Bibliography


