Bilateral Port Wine Stain with Involvement of Chest Dermatomes and Unilateral Buphthalmos: A Case Report

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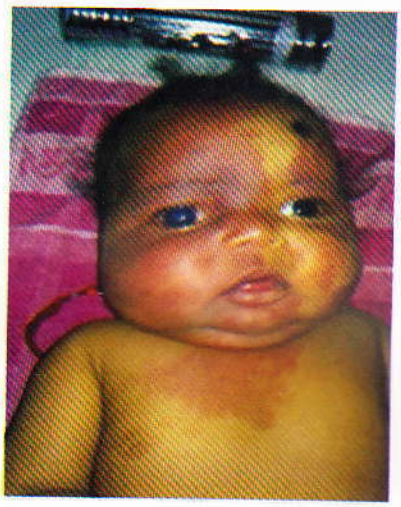
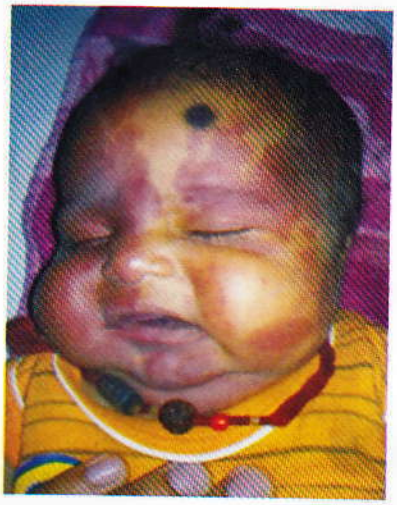
Introduction

Sturge Weber Syndrome (SWS) is a rare Oculo cutaneous disorder, which manifests with facial capillary haemangioma, congenital glaucoma and lepto-meningeal abnormalities which in turn leads to central nervous system (CNS) afflictions like seizures, behavioural and developmental disorders. The CNS involvement is considered a hallmark of SWS.[1] SWS has been reported more commonly with unilateral port wine stains but bilateral presentations with associated chest dermatome involvement is rare.[2] We report a patient one month old with bilateral port wine stain, with buphthalmos and involvement of chest dermatomes, with no evidence of CNS involvement.

Case Report

A two month old male child presented with bilateral facial port wine stains and also in right upper chest area. The infant was a normal, full term, uneventful and unsupervised home delivery in a rural household. The right eye of the patient has a central corneal opacity, which historically has increased in size in the last two months.





The right eye had a horizontal corneal diameter of 14 mm while the vertical diameter is 13 mm; a hand held slit lamp shows the opacity to be stroma deep and presence of haab's stria, the anterior chamber was deep and the iris and pupil appeared normal, fundus details were not visible because of the opacity. The left eye had a horizontal corneal diameter of 12mm, a vertical corneal diameter of 11mm, the cornea was clear and the anterior chamber deep, the iris and pupil had no abnormalities and the optic nerve head as seen by a direct ophthalmoscope appeared normal. The intraocular pressure (IOP) of the right eye by a hand held Perkins tonometer was 22mm and 16 mm in the left eye. The port wine stain was present in the distribution area of the ophthalmic and maxillary division of the trigeminal nerve on both sides of the face; stains were also present on the chest dermatomes on the right side. A CT-scan head was done which was normal. The child was followed up for more than one year, and all developmental milestones were normal.

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A differential diagnosis of Sturge Weber syndrome and other neurocutaneous syndromes were considered. The IOP was lowered using topical beta blockers initially for a week, and later a Trabeculectomy with Mitomycin –C was done in the right eye.

Discussion

Sturge Weber syndrome is a neuro-cutaneous syndrome with variable presentations, with common manifestations like congenital glaucoma, facial and lepto-meningeal angiomas without any definite genetic predisposition. It is found in 1 in 50,000 live births.[3,4] Buphthalmos has been commonly found in patients with bilateral port wine stains and with CNS involvement[5] but in our case it is bilateral port wine stains with no CNS involvement. Onset of seizures below one year of age is more common with bilateral port wine stains and extra facial locations of port wine stains e.g. chest or torso[6] while in our case there has been no evidence of seizure, this indicates that despite presence of large angiomas, the Vascular-steal-phenomenon[7] has not been able to have its effect. The fact that the milestones were normal, also suggests that there has been no CNS involvement. The presence of corneal haziness with raised IOP indicates that glaucoma could be because of maldevelopment of structures of angle of anterior chamber than due to raised episcleral pressure. This view is also supported by the fact that glaucoma is unilateral despite facial angiomas being present of both sides of the face.

Conclusion

Sturge Weber syndrome may not present clinically in its typical form, but may have extensive port wine stains with congenital glaucoma, and no central nervous system involvement.

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In seahorses, it's the male who gives birth to the young.