CASE REPORT

Ipsilateral Glaucoma in Sturge-Weber Syndrome

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Abstract

A case of Sturge-Weber syndrome in a young Chinese woman is presented. The classic facial cutaneous angioma extends to involve other parts of her body. Ipsilateral glaucoma previously undiagnosed was detected. Associated field defects include enlargement of the blind spot and a general depression of the field.

Abrégé

L'histoire d'une jeune chinoise souffrante du syndrôme Sturge-Weber fait le sujet de ce rapport. L'angiome facial typique se reproduit ailleurs sur son corps. Un glaucome monoculaire insoupçonné, du même coté que l'angiome a été révélé ainsi qu'une contraction du champ visuel et un aggrandissement de la papille de Mariotte.

A twenty-three year old Chinese female university student presented at the University of Waterloo School of Optometry clinic for a routine eye examination complaining of decreased acuity in her right eye. She reported that the vision of her left eye had been poor since birth. Her previous spherical correction had been provided one year earlier. She claimed and appeared to be in good general health and was taking no medications. A family and personal history, elicited with difficulty, was unremarkable.

Diagnostic Data

The patient's best corrected acuity was 6/6(20/20) in her right eye with -4.25/-0.75 x 165 but her left eye could not be improved to better than CF at 2/3 metre although laser interferometry suggested a potential 6/120 (20/400). A dramatic "port wine" angioma (nevus flammeus) covered most of the left trigeminal region with sparing of the lower maxillary region (Fig. 1). The lesion extended to involve the scalp, ear, retroauricular region, shoulder, upper chest, arm and hand (Fig. 2). She reported additional islands existing elsewhere on her body and the involvement of the left oral mucosa was readily apparent.

Other than a left Marcus Gunn pupil there were no overt neurological signs or symptoms.

The external structures and anterior segment of the right eye were normal; ophthalmoscopy revealed a healthy fundus with a 0.5 physiologically cupped disc vertically and horizontally.

Telangiectatic vessels were noted in the left conjunctiva and although no choroidal hemangiomata were seen the left disc was 0.9 pathologically cupped (Fig. 3). The size of the cornea and globe did not vary from the right eye.

Intraocular pressure was measured at R.18 mmHg L. 45 mmHg. (Goldmann) Gonioscopy (Fig. 4) revealed mesodermal tissue extending over the trabeculum in an otherwise normal open angle.

Goldmann perimetry (Fig. 5) indicated an overall depression of all isopters and a very enlarged blindspot rather than other classical glaucomatous defects.

Treatment Plan

The patient was diagnosed as Sturge-Weber Syndrome with ipsilateral glaucoma but without overt CNS or visceral signs, and referred for ophthalmological management. The initial diagnosis was confirmed and pilocarpine 2% q.i.d. was prescribed for the left eye. After four weeks this was increased to pilocarpine 4% q.i.d. which reduced IOP to 31 mmHg (Goldmann). Subsequently timolol 0.5% b.i.d. has been added to the treatment regimen resulting in adequate medical control of IOP level. In view of the absence of CNS systemic signs and/or symptoms the eye surgeon felt it inappropriate to subject the patient to comprehensive laboratory and hospital investigation for possible CNS or visceral involvement.

Discussion

The classical dermal sign, a flat (rarely tuberous) cutaneous facial angioma, which involves one or more branches of the Vth cranial nerve initially attracts attention to the victims of this disease, yet it is the hidden associated conditions which are potentially more ominous. These may be ocular, neurological and/or visceral. Ipsilateral glaucoma occurs in between 10% and 30% of patients with Sturge-Weber syndrome; if it develops *in utero* or

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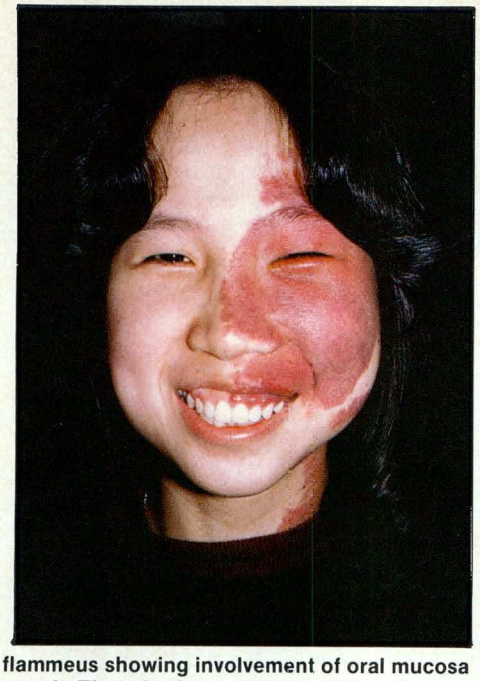
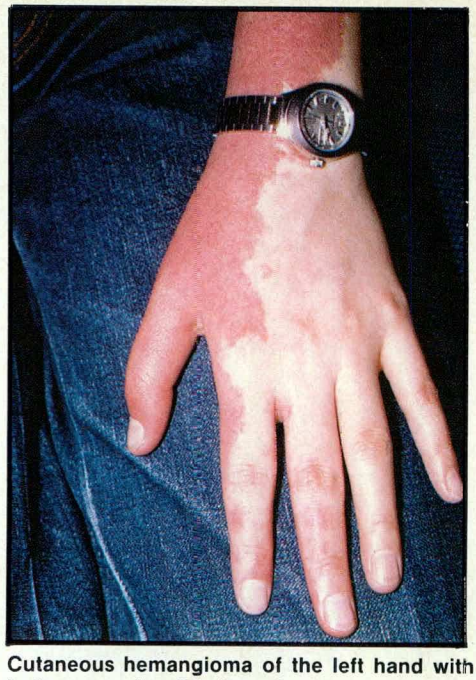


Fig. 1 Nevus flammeus showing involvement of oral mucosa and the neck. There is also a slight facial hemihypertrophy.



Cutaneous hemangioma of the left hand with distribution over the C6 dermatome. Fig. 2

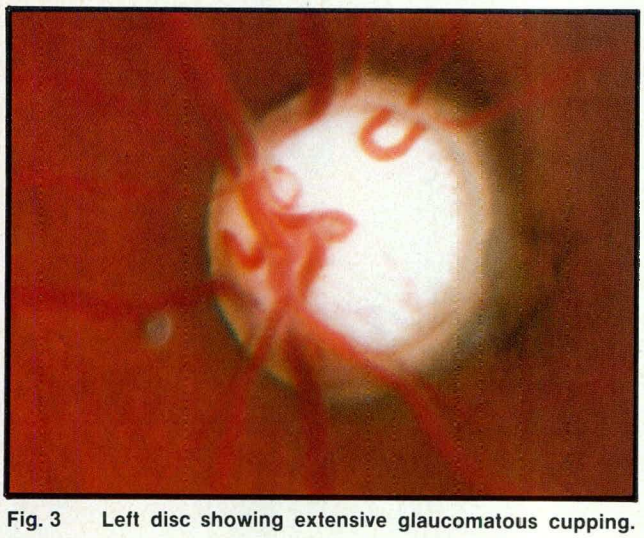
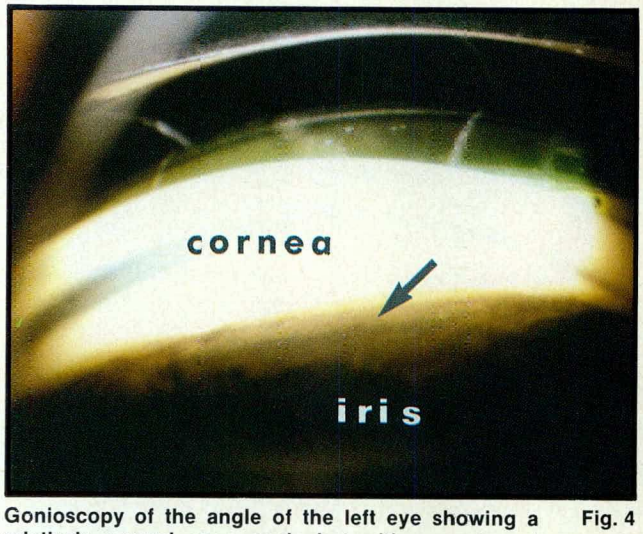


Fig. 3 Left disc showing extensive glaucomatous cupping.



Gonioscopy of the angle of the left eye showing a relatively normal open angle but with mesodermal tissue on the trabeculum (arrow). Fig. 4

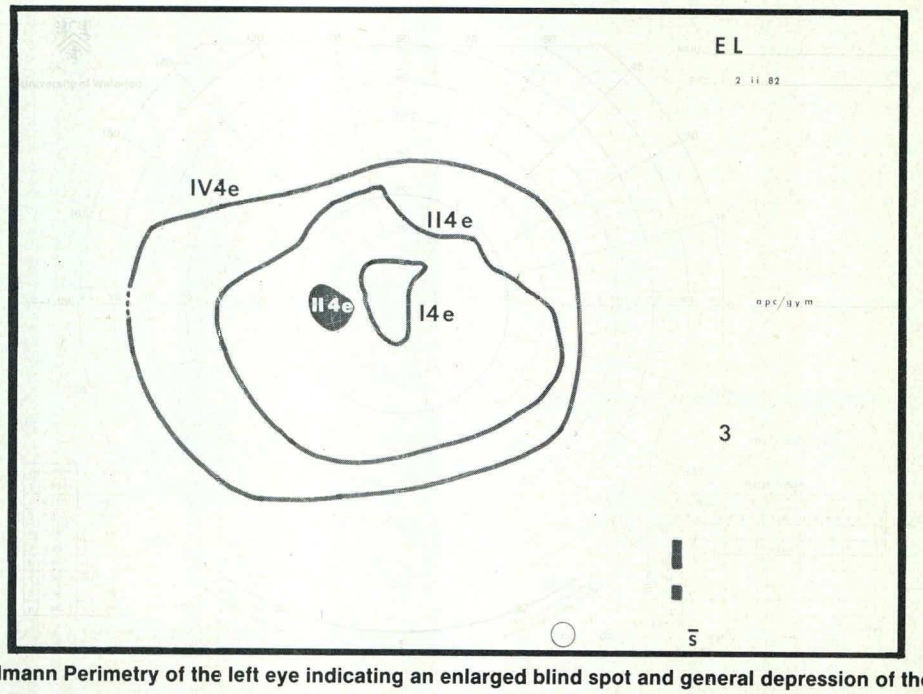


Fig. 5 Goldmann Perimetry of the left eye indicating an enlarged blind spot and general depression of the visual field.

early in life bupthalmos results. Bilateral and contralateral glaucoma occur only rarely[1] and the incidence suggests that some of these cases may not be related to the syndrome. The glaucoma may be due to developmental malformations in the angle similar to other forms of congenital glaucoma or may more frequently be associated with aqueous outflow interference caused by vascular anomalies of the limbal or episcleral regions.[2] Other Sturge-Weber patients have choroidal hemangiomata[3] which may lead to retinal detachment, rubeosis iridis, PAS (peripheral anterior synechiae) and secondary glaucoma. Leptomingeal angiomas and underlying cerebral cortex calcification may lead to local contralateral seizures and hemiparesis or even general seizures[4]; more severe cerebral involvement results in mental disturbance and retardation.

Fortunately, associated visceral findings are rare but Zion[5] describes angiomata of the lung, G.I. tract, pituitary, ovary and pancreas. Unlike most of the other phakomatoses the hereditary nature of Sturge-Weber syndrome is not proven although a number of families show transmittance in incomplete and varying forms.[6] The disease is not uncommon and may affect all racial groups[5] with no sexual prediliction. The facial angioma is so pathognomonic that the alternatives for differential diagnosis are very limited, however in this case the lesion of the left hand is suggestive of the related Klippel-Trénaunay-Weber syndrome[7]. This was ruled out as there was no evidence of hypertrophy of the bone and soft tissue of the hand.

Conclusion

This young woman has been diagnosed as having Sturge-Weber syndrome with both an extensive cutaneous angioma and ipsilateral glaucoma yet without meningeal or other involvement thus representing Schirmer's form of the disease. Although she reports poor vision since birth the normal size of the eye and cornea suggest a later onset of the markedly elevated IOP. The IOP of the left eye is maintained at an acceptable level by medical means but lifetime compliance with such a regimen is unlikely. Surgical procedures for this type of glaucoma, which include goniotomy, trabeculotomy and cylocryotherapy, are effective in some cases.

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