INTRODUCTION

Sturge-Weber syndrome (SWS) also known as encephalo-trigeminal angiomatosis belongs to a group of disorders collectively called phakomatosis. It consists of congenital hamartomatous malformations that may affect the eye, skin, and central nervous system at different times. SWS is classified into complete trisystemic (when all three organ systems are involved), incomplete bisystemic (when the involvement is either oculocutaneous or neurocutaneous), and incomplete monosystemic (when there is only neural or cutaneous involvement).\(^1\) The syndrome likely results from an early embryologic malformation of vascular development affecting the development of nearby skin, eye and brain structures.\(^2\)

The hallmark of SWS is a facial cutaneous venous dilation, also referred to as port-wine stain, which appears as dull red patches of irregular outline, along the distribution of one or more divisions of the trigeminal nerve. Neurological features include focal or generalized motor seizures, mental retardation and neurologic deficits as hemiplegia or homonymous hemianopia. Ocular manifestations include eyelid haemangiomas, conjunctival and episcleral haemangiomas, glaucoma, heterochromia of the irides, diffuse choroidal haemangiomas, and tortuous retinal vessels with occasional arterio-venous communications.\(^1\,3,4\) These pathological changes may also induce a shift in refractive state of the involved eye which results in anisometropia or even amblyopia if it occurs at an early age and is not properly managed.

This case report describes choroidal haemangioma occurring in a teenage boy with SWS, which persecuted as unilateral hypermetropia and glaucoma.

CASE REPORT

A 16 years old male patient of Sturge-Weber syndrome was referred to glaucoma clinic for the management of unilateral glaucoma. There was also an ipsilateral hypermetropic shift. On detailed investigations, a diffuse choroidal haemangioma was diagnosed which induced this hypermetropic shift. Anisometropia in Sturge-Weber syndrome can give us clue regarding some underlying pathology, so unilateral myopia or hypermetropia should be thoroughly evaluated in such patients.

Key words: Choroidal haemangioma. Hypermetropia. Sturge-Weber syndrome. Unilateral glaucoma. Anisometropia.
alterations at macula were noted, however, any elevation or abnormal appearance of posterior pole was not appreciable which could explain the hypermetropic shift.

For further evaluation, investigations like fundus photographs, B-Scan and fundus fluorescene angiography (FFA) were advised. Simultaneous comparison of both fundi with fundus photographs clearly demonstrated a diffuse red lesion at posterior pole of RE suggestive of diffuse choroidal haemangioma which was difficult to appreciate with ophthalmoscopy (Figure 1). B-Scan ultrasonography also showed diffuse choroidal thickening (Figure 2) while, FFA revealed widespread areas of hyperfluorescence secondary to diffuse leakage of dye which confirmed the diagnosis of diffuse choroidal haemangioma (Figure 3).

Keeping in view the systemic associations of Sturge-Weber syndrome, a skull radiograph was advised and opinion from a neurologist was taken. No calcification was seen on skull radiograph while neurological examination was also unremarkable.

Regarding management, the patient was advised topical beta blocker in RE and his intraocular pressures were well controlled on follow-up visits. Retinal consultation from vitreo retina department was taken and various management options for diffuse choroidal haemangioma were discussed with the patient including simple observation with regular follow-up or prophylactic laser therapy. The patient preferred the option of observation and regular follow-up at retina clinic.

**DISCUSSION**

Decreased visual acuity in patients with SWS may be due to several possible mechanisms which include glaucoma, diffuse choroidal haemangioma, angiomia of leptomeninges, refractive errors (anisometropia) and amblyopia.

Glaucoma is mostly unilateral and ipsilateral to the port-wine stain. The risk of glaucoma is increased when the facial skin changes involve upper eyelid. Most accepted explanation for the elevated intraocular pressure is a combination of developmental angle anomalies, which have a dominant role in infantile onset glaucoma and elevated episcleral venous pressure, which is more important in later onset glaucoma. Since, the infant's eye is damaged quickly by increased IOP, secondary glaucomatous changes occur, including increased corneal diameter, tears in the Descemet membrane, buphthalmos and optic nerve damage which result in decreased vision.

Diffuse choroidal haemangioma usually affects over half of the choroid and enlarges very slowly in childhood. During adolescence, marked thickening of the choroid sometimes becomes evident with secondary changes to overlying ocular structures. The retina over the haemangioma may be attached and well preserved, attached and degenerated, or detached. Degenerative changes also occur in the overlying retina including focal chorioretinal adhesions, loss of photoreceptors, severe cystoid degeneration, marked gliosis and exudative retinal detachment.

Angioma of leptomeninges in the occipital lobe may also result in visual symptoms. As this is a neurological type of lesion, it usually produces hemianopic visual field defects. Another reason for decreased vision in these patients is refractive error which may occur as a result of myopic or hypermetropic shift in the involved eye. Glaucoma usually induces a myopic shift. Rise in IOP in the first 2 years of life when the collagen fibers are more elastic leads to enlargement of globe which results in myopic shift of the involved eye. Diffuse choroidal haemangioma on the other hand results in hypermetropic shift due to choroidal thickening causing elevation of retinal surface. As the haemangioma enlarges very slowly and remains asymptomatic in early childhood, this hypermetropic shift occurs at later stage of life.

Amblyopia is another reason for decreased vision in these patients. Amblyopia usually is anisometropic due to the difference in refractive state of affected eye as
explained above. It may also develop secondary to stimulus deprivation in cases of hazy corneas.7,8 Diffuse choroidal haemangioma can sometimes become a diagnostic challenge and may be overlooked easily on ophthalmoscopy. The colour of haemangioma resembles that of normal fundus, and the elevation may be minimal, especially in children. FFA and indocyanine green angiography (ICGA) may be important and sensitive in detecting diffuse choroidal haemangioma that is not detected by clinical examination. However, before going for such investigations, presence of hypermetropic shift in the eye can also give us a clue and aid in diagnosis of diffuse choroidal haemangioma.8-10

Refractive state of eye especially unilateral myopia or hypermetropia should be given consideration in the clinical evaluation of patients with SWS as such shift in the refractive state might be due to some underlying pathology. Early detection of such lesions can be beneficial in preventing serious sight threatening complications related to their un-noticed progression.

REFERENCES


