·Case Report·

A case of circumscribed choroidal hemangioma in Sturge-Weber syndrome in China

Xiao-Lei Yin[1,2], Jian Ye[2]*, Rong-Di Yuan[2]*, Shu-Xing Ji[2]**

[1]Department of Ophthalmology, 305 Hospital of Chinese PLA, Beijing 100017, China
[2]Department of Ophthalmology, Daping Hospital, the Third Military Medical University, Chongqing 400042, China
*These authors contributed equally to this work.
Correspondence to: Xiao-Lei Yin. Department of Ophthalmology, 305 Hospital of PLA, Jia 13, Wenjin Street, Xicheng District, Beijing 100017, China. yinxiaolei971221@163.com
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Abstract

• We present a case of circumscribed choroidal hemangioma (CCH) in Sturge-Weber syndrome in a 30-year-old woman with congenital port-wine stains on the left side of face involving the upper eyelid, cheek and the nose, and she had undergone facial hemangioma surgery 3 years ago suggestive of Sturge-Weber syndrome. She presented with a 1-month history of rapidly decreased visual acuity (VA) to counting fingers in the left eye which had no prior history of visual problem. And there was no evidence of glaucoma. At 3 months after the treatment of the standard photodynamic therapy (PDT) the VA was 20/200. For some reasons, we have no idea about the changes of tumor thickness and subretinal fluid. We confirmed the curative effect of PDT treatment for CCH because of the significantly improved VA in the bad eye.

• KEYWORDS:Sturge-Weber syndrome;circumscribed choroidal hemangioma; photodynamic therapy
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INTRODUCTION

In Sturge-Weber syndrome, the most common vascular lesion is a stain birthmark that can vary in color from light pink to deep purple and is caused by an overabundance of capillaries around the trigeminal nerve just beneath the facial surface. Unlike capillary angiomas, the birthmark will not resolve over time, and it may be associated with blood vessel abnormalities involving the eye on the same side of the face, the choroidal haemangioma [1-2]. Circumscribed (or diffuse) choroidal hemangioma is a rare, benign vascular tumor that may subsequently lead to the development of subretinal fluid, exudative retinal detachment with macular involvement, photoreceptor cell loss and cystoid degeneration of the sensory retina causing visual loss [3], in contrast to the well-circumscribed choroidal hemangiomas seen in patients without the syndrome. We report a case of circumscribed choroidal haemangioma (CCH) in Sturge-Weber syndrome in China.

CASE REPORT

A 30-year-old woman with congenital port-wine stains on the left side of face involving the upper eyelid, cheek and the nose had undergone facial hemangioma surgery 3 years ago suggestive of Sturge-Weber syndrome (Figure 1A). She presented with a one-month history of rapidly decreased visual acuity (VA) in the left eye. She had no prior history of visual problem. In the examination, the VA was 20/20 in the right eye and counting fingers in the left eye, and the intraocular pressure was 14.57mmHg and 9.36mmHg, respectively. The cornea was clear and anterior segment was normal. There was no evidence of glaucoma. Her fundus photograph showed CCH with associated subretinal fluid in the left eye (Figure 1B). Magnetic resonance imaging (MRI) confirmed diffuse thickening of the choroid (Figure 1C) and fundus fluorescence angiography (FFA) revealed subretinal perivascular exudation (Figure 1D). The standard photodynamic therapy (PDT) protocol was used for the treatment[4]. At 3 months after the treatment the VA was 20/200 in the left eye. However, unfortunately for the economic problem, the patient was satisfied with the VA of the bad eye and she refused to take other examinations, so we have no idea about the changes of tumor thickness and subretinal fluid.

DISCUSSION

Sturge-Weber syndrome is a rare neurocutaneous syndrome that occurs with a frequency of approximately 1 per 50 000 and produced early in fetal development and account for the high frequency of association between unilateral involvement of facial ectoderm, globe, and occipital cortex of the brain [1]. The syndrome occurs almost entirely sporadically and with equal frequency in both sexes [5]. Although the syndrome is congenital, there is generally no

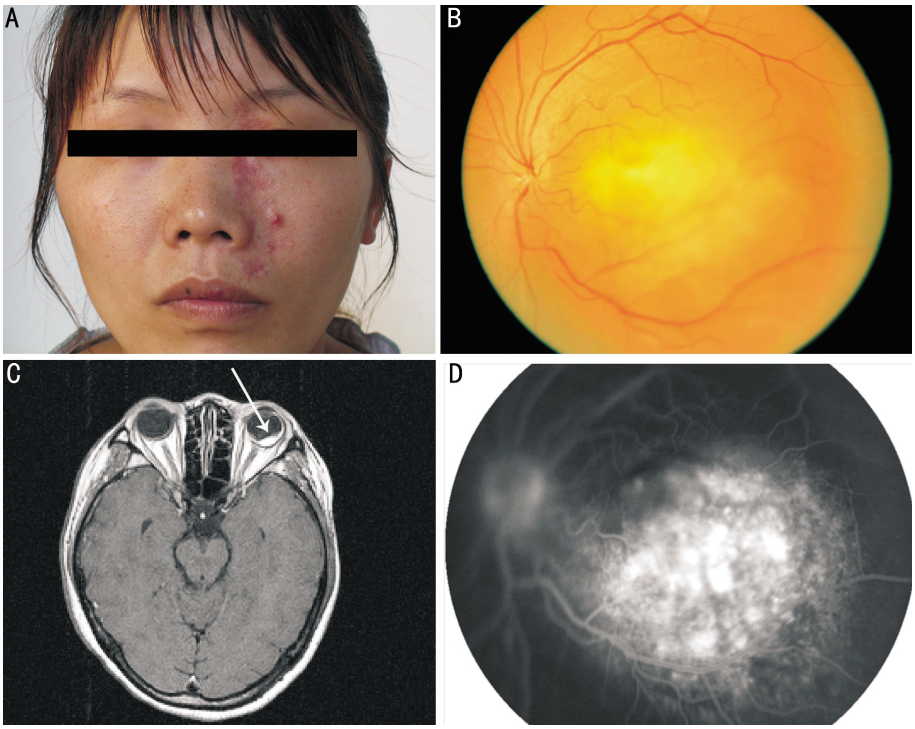

Figure 1 A circumscribed choroidal haemangioma in Sturge-Weber syndrome in China A: Port-wine stains on the left side of face; B: CCH with associated subretinal fluid in the left eye; C: MRI confirmed diffuse thickening of the choroid; D: FFA revealed subretinal perivascular exudation

heritability, and even if there were familial cases it was few[6]. The port-wine stains are irregular in shape and are usually in the distribution of the ophthalmic branch of the trigeminal nerve. Choroidal hemangioma, and other parts of Sturge-Weber syndrome almost always ipsilateral to the port-wine stain, may be associated with formation of subretinal fluid, serous retinal detachment, and cystoid macular edema can cause progressive visual loss [3]. After PDT was first successfully used for the treatment of symptomatic CCH in 2000 [7], many groups confirmed it was a safe, effective treatment for CCH, although it might lead to the development of focal chorioretinal atrophy following either modified or standard treatment protocol, and might be associated with transient visual disturbances[4]. In the current study we presented a case of PDT treatment for CCH in Sturge-Weber syndrome. Even if we did not know the changes of tumor thickness and subretinal fluid after therapy for some reasons, the VA of the patient improved significantly. We confirmed the curative effect of PDT treatment for CCH. Nevertheless, questions emerge such as, although the choroidal haemangioma with Sturge-Weber syndrome has no pronounced heritability, whether it has racial tendency or not. For the classic vascular malformations possibly result from a failure of the primitive cephalic venous plexus regressing during vascular development [6], why the initial choroidal haemangioma does not blur vision?

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