

## **LOCALIZED CHOROIDAL HAEMANGIOMA ASSOCIATED WITH STURGE - WEBER SYNDROME: A CASE REPORT**

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### **ABSTRACT**

Sturge-Weber syndrome (SWS) is a sporadic congenital neurooculocutaneous disorder that may present with glaucoma and vascular malformations of the conjunctiva, episclera, choroid and retina. We report a case of localized choroidal haemangioma associated with SWS Type I. A 26-year-old male with a port-wine stain birthmark on right side of the face was admitted to our clinic with complaints of photophobia, pain, and blurred vision in his right eye. Anterior segment examination of the right eye revealed dilatation and tortuosity of conjunctival vessels. Snellen visual acuity was 0.4 (20/50) and intraocular pressure - normal. Fluorescein angiography demonstrated a small-spotted hyperfluorescent mass nasal to the optic disc and exudative retinal detachment in the central zone. Testing of the left eye found no abnormalities. Brain CT scans showed bilateral tram-track calcifications. This case report arouses certain clinical interest because of its rare incidence, continued asymptomatic development, and delayed diagnosis only after the presentation of exudative retinal detachment with subsequent visual deficit. Neuroophthalmological monitoring of patients with SWS may be useful for early detection of ocular involvement before the appearance of serious visual complications.

**Key words:** Sturge-Weber syndrome, localized choroidal haemangioma, exudative retinal detachment, neuroophthalmological monitoring

### **INTRODUCTION**

Sturge-Weber syndrome (SWS) is a sporadic congenital neurooculocutaneous disorder (3, 7, 20). Ocular abnormalities can include glaucoma and vascular malformations of the conjunctiva, episclera, choroid and retina (2, 5, 13, 17). Although glaucoma is the most common ocular involvement, diffuse choroidal hemangioma is another characteristic feature of SWS that may be found in 31% to 71% of SWS cases (2, 7, 11, 13, 16). This choroidal haemangioma usually evident of birth appears as an orange or red diffuse choroidal thickening, producing a “tomato-ketchup” appearance on funduscopy (3, 4, 5, 17). Retinal pigment epithelium degeneration, fibrous methaplasia and

cystic retinal degeneration as secondary changes of the diffuse choroidal haemangioma contribute to visual loss and visual field defects (7, 14, 16, 18). Except these findings, the diffuse choroidal hemangioma of SWS may have localized areas simulating a circumscribed choroidal haemangioma (1, 6, 12, 14, 19). Hystologically both types of angioma are of the cavernous variety.

Data from previous clinical observation reveal that circumscribed choroidal haemangiomas occur only sporadically, without any associated local or systematic anomalies (7, 11, 14, 18). In contrast, literature review demonstrates that localized angiomas of the choroid are less common, but could be a part of SWS (3, 6, 8, 10, 13, 15, 17). They are diagnosed predominantly between the second and fourth decade of life, when a secondary exudative retinal detachment is present generally resulting in progressive visual loss (5, 7, 11, 12, 16, 20). Accordingly, we report a case of localized choroidal haemangioma associated with SWS Type I leading to ocular complications with subsequent vision deficit.

### **CASE PRESENTATION**

A 26-year-old male with a port-wine stain birthmark on the forehead and upper eyelid of right side of his face (pic 1) was admitted to our clinic with complaints of photophobia, pain, and blurred vision in his right eye. Examination of the anterior segment of the right eye revealed dilatation and tortuosity of conjunctival vessels. Localized choroidal vascular malformations with exudative retinal detachment were presented nasal to the optic disc. Snellen visual acuity was 0.4 (20/50) and intraocular pressure - normal (18 mm Hg - Schiötz). The left eye ophthalmological evaluation was normal. Fluorescein angiography demonstrated a small-spotted hyperfluorescent mass nasal to the optic disc in the choroidal filing phase of the angiogram and an exudative retinal detachment in the central zone (fig. 1). Brain CT scans showed bilateral tram-track calcifications (fig. 2). Medical history revealed presentation of motor simple partial seizures affecting the left facial side and arm at the age of five months.

## DISCUSSION

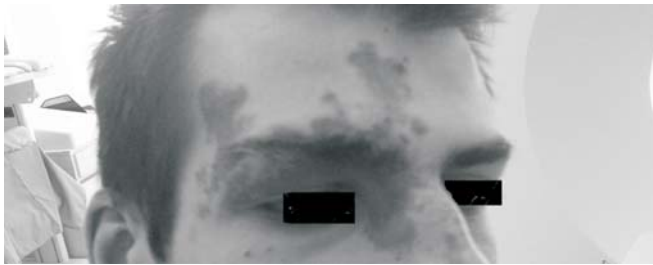
Sturge-Weber Syndrome belongs to a group of neurocutaneous disorders manifested by facial and leptomeningeal angiomas (3, 6, 7, 10, 18). The Roach Scale classifies it into complete trisymptomatic and incomplete mono- or bisymptomatic forms. The 3 general types (I, II, and III) of SWS are clinically defined by the association of cutaneous, central nervous system, and ocular abnormalities (3, 7, 17, 20). Close to these data, we report a case of a 26-year-old male diagnosed as a complete form of SWS Type I on the basis of external clinical sign (facial hemangioma) and CT findings (bilateral cerebral angiomas).

The most common ocular abnormalities of SWS include glaucoma, conjunctival or episcleral hemangiomas, and either diffuse or localized choroidal hemangiomas (2, 4, 6, 9, 20). The literature review reveals that localized choroidal hemangiomas associated with encephalotrigeminal angiomatosis are rare and usually asymptomatic (3, 5, 7, 15, 17). That's why, their diagnosis prior to manifestation of different ocular complications such as progressive visual loss or visual field anomalies presents a clinical challenge. Our case presentation of localized choroidal hemangioma with continues asymptomatic development is in accordance with data from previous reports (1, 8, 13, 20).

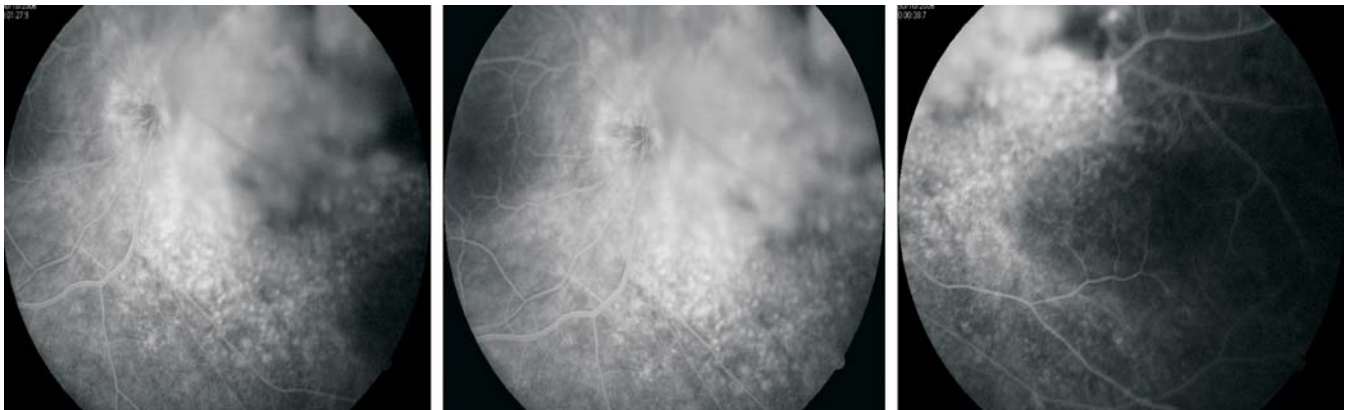
Evidence exists that with time, choroidal hemangioma may cause various retinal complications such as pigment epithelium degeneration, fibrous metaplasia, cystic degeneration, and detachment (2, 5, 11, 14, 17). Also retinal vascular tortuosity, iris heterochromia, and cataracts are found in patients with SWS. Our neuroophthalmological findings of secondary exudative retinal detachment and the corresponding clinical manifestation of progressive vision decrease contribute to these data. Obviously the long-term ophthalmological examination with early recognition of ocular changes improves the successful medical management of SWS.

## CONCLUSION

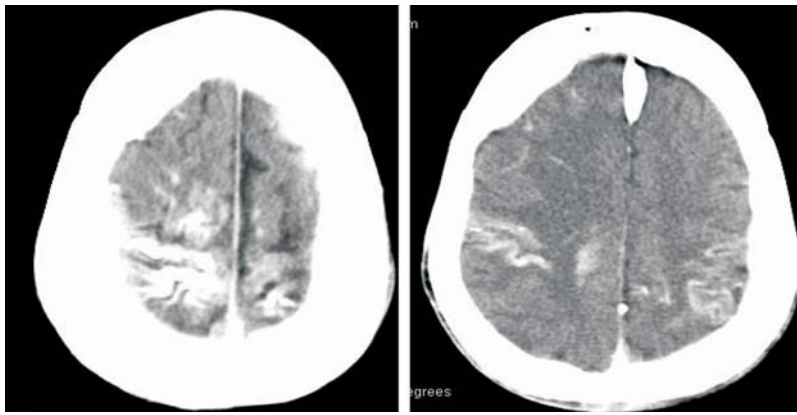
This case report of localized choroidal hemangioma associated with SWS Type I arouses certain clinical interest because of its rare incidence, continued asymptomatic presentation, and delayed diagnosis only after the development of exudative retinal detachment with subsequent visual deficit. Based on our own notices and literature review, we suggest that in patients with SWS neuroophthalmological monitoring may be useful for early detection of ocular involvements before the appearance of serious visual complications.



**Pic 1.** A port-wine stain birthmark on the forehead and upper eyelid of right side of face.



**Fig. 1.** Fluorescein angiography demonstrates a small-spotted hyperfluorescent mass nasal to the optic disc in the choroidal filling phase of the angiogram and an exudative retinal detachment in the central zone.



**Fig. 2.** Brain CT scans show bilateral tram-track calcifications.

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