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CIRCUMSCRIBED CHOROIDAL HEMANGIOMA IN STURGE-WEBER SYNDROME

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PURPOSE

To report a case of choroidal hemangioma in a patient with Sturge-Weber syndrome (SWS).

METHODS

Analysis of medical records and imaging.

RESULTS

A 27-year-old patient presented with progressive worsening of visual acuity for 1 year. Her personal history includes a port wine stain on the right hemiface since birth and advanced glaucoma diagnosis at the age of 23, which was partially controlled by trabeculectomy surgical treatment. The patient was referred for evaluation due fundus alteration. Upon clinical examination, she had best corrected visual acuity of 20/80 in the right eye and 20/20 in the left eye according to the Snellen chart. Fundoscopy revealed reddish hyperpigmentation in the posterior pole region. Angiofluoresceinography showed an extensive area of early and slightly progressive hyperfluorescence. OCT revealed no foveal depression, increased thickness of inner and outer retinal layers with intraretinal cysts, bulging of the posterior sclera, and increased choroidal thickness with increased reflectivity. Ocular ultrasound demonstrated the presence of a solid, echogenic, dome-shaped lesion with homogeneous content in the temporal periphery adjacent to the macula suggesting a circumbscribed choroidal hemangioma. A diagnosis of SWS was made indicating the necessity of neuroimaging exams, which showed no abnormalities. Currently, the patient continues to be clinically monitored and awaits for local therapy evaluation on the right eye.

REFERENCES

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Singh AK, Keenaghan M. Sturge-Weber Syndrome. [Updated 2023 May 1]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK459163

DISCUSSION

SWS is a sporadic phacomatosis condition with characteristic cutaneous and neurological manifestations. It is clinically diagnosed by the presence of typical port wine stains and neurological features and/or glaucoma. Choroidal hemangiomas can be present in up to 40% of cases. These rare vascular hamartomas, believed to have a congenital origin, are non-proliferative tumors that progressively enlarge due to venous congestion within the mass. Ophthalmologists should be fully aware of this rare syndrome to provide earlier counseling and management options for these patients to improve or preserve visual acuity

IMAGES

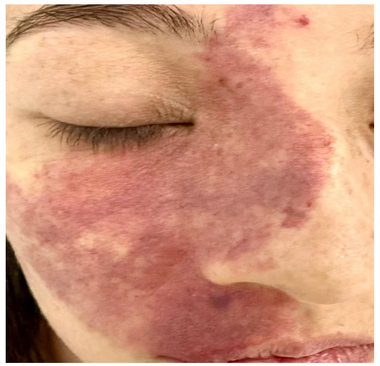

Image 1: Port wine stain

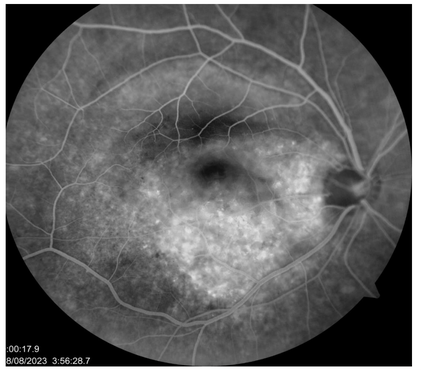

Image 2:Angiofluoresceinography 17s

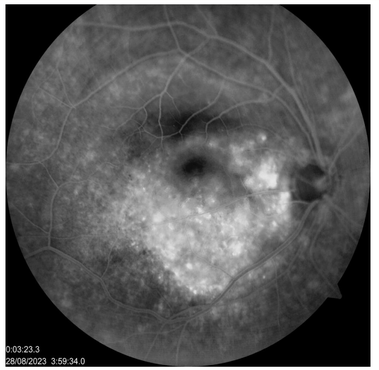

Image 3: . Angiofluoresceinography 23s

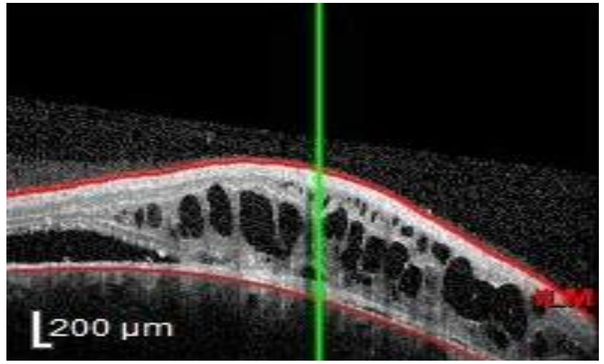

Image 4: OCT

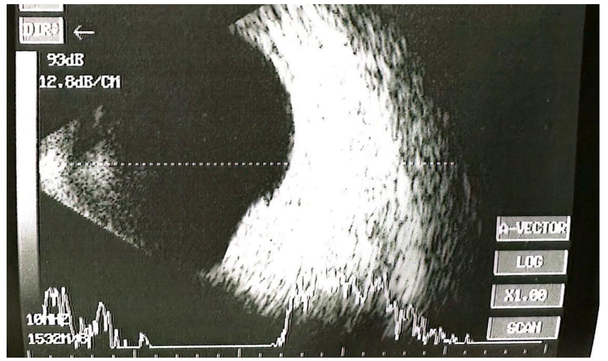

Image 5: Ocular ultrassound