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CASE REPORT

Partial thickness sclerectomy to treat exudative retinal detachment secondary a submacular choroidal hemangioma in a Sturge–Weber syndrome

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Abstract
Purpose To demonstrate the safety and efficacy of partial thickness sclerectomies to treat exudative retinal detachment secondary to choroidal hemangioma, non-suitable with photodynamic therapy in a 5-year-old child with Sturge–Weber syndrome.
Methods A 5 year-old child presented exudative retinal detachment secondary to choroidal subfoveal diffuse hemangioma. The child was non-compliant to undergo a photodynamic therapy. A partial thickness sclerectomy was made in each quadrant under general anesthesia.
Results The retina was re-attached with improvement in vision from 20/400 to 20/80. Two years after primary surgery, the retinal detachment relapsed. Drainage of the subretinal fluid was obtained by the revision of the sclerectomies. After obtaining retinal reattachment, photodynamic therapy was applied to treat the hemangioma. No complications were reported after treatment.
Conclusion Sclerectomies may be considered an efficient and safe surgical option for the management of exudative retinal detachment secondary to choroidal hemangioma in patients non-suitable for photodynamic therapy, waiting for photodynamic therapy to be practicable directly on the hemangioma.

Keywords Choroidal hemangioma · Exudative retinal detachment · Retinal detachment · Sclerectomy · Sturge–Weber syndrome · Photodynamic therapy

Case report

A 5-year-old boy, with Sturge–Weber syndrome, was referred for retinal detachment to the S. Anna Clinical Institute, Brescia, Italy. He presented progressive vision loss in his left eye (LE) over the previous months. He had a diffuse facial port-wine nevus involving the skin of both cheeks, the nose, the chin, the lips, both the upper and lower lids of the left eye and only the nasal side of the right eye. A secondary glaucoma had been treated with previous goniotomy in the LE elsewhere, 3 years before. Best-corrected visual acuity (BCVA) was 20/20 in the right eye (RE) and 20/200 in the LE.

Slit-lamp examination revealed a normal RE and a buphthalmic aspect of the LE. In the LE, biomicroscopy showed moderate disk cupping, with 0.6/0.7 cup/disk ratio, and a bright red appearance of the fundus, with retinal detachment involving the lower

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part of the retina, including the macula. No breaks were found at inspection.

At the optical coherence tomography (OCT) (Spectralis; Heidelberg Instruments Inc.; Germany), the subretinal fluid was noted to be more represented in the lower quadrants, with a gravitational aspect. Intraocular pressure (IOP) was 16 mmHg in the RE and 17 mmHg in the LE, respectively. The LE was under medical treatment with timolol/dorzolamide drops.

Fundus infrared photograph was normal in the RE and revealed, in the LE, some hyperpigmented multiple flecks-like spots, around the fovea with a circular pattern (Fig. 1). Fundus autofluorescence (Spectralis) showed a hypoautofluorescence focused within the spots delimited area (Fig. 1). The OCT (Spectralis) indicated a retinal detachment of the fovea, increasing in thickness toward the lower quadrant with a gravitational aspect, a subfoveal nodular formation under the RPE layer (Fig. 2). The choroid in the RE was 543 microns (Fig. 3) and 2.5 times thicker than the central retinal thickness. In the LE, the choroidal thickness was not measurable with the OCT (Fig. 2) due to excessive thickness.

A-scan and B-scan ultrasonography (Aviso; Quantel Medical; France) confirmed a lower retinal detachment and revealed a thickened choroid with maximum thickness of 0.97 mm.; in correspondence with the hemangioma, the maximum thickness increased up to 1.71 mm (Fig. 4); the scleral thickness was not measureable accurately with the B scan available, but it appeared to be thicker than the choroid.

The axial length (AL) was 22.77 mm in the RE and 22.88 mm in the LE, respectively.

Surgery was performed under general anesthesia. The sclera was exposed performing a 360 degrees lumbar conjunctival peritomy. The sclera appeared extremely and unusually vascularized. After careful hemostasis, a partial thickness scleral flap (5 × 5 mm square shaped and 1 mm thick) was made in the central part in each exposed quadrant, between the rectus muscles, with the posterior margin at 14 mm from the limbus, taking care to avoid the vortex veins. The underlying choroid, after removal of scleral flap, was not fully visible, due to the thickness of the sclera. The conjunctival wounds were closed with 7.0 reabsorbable suture (Fig. 5).

A biopsy of the hypervascularized sclera was obtained for analysis. Histological examination of the scleral flaps showed reactive telangiectasias with mild-to-moderate perivascular infiltrates, supported by neutrophils associated with sporadic eosinophils (Fig. 6). No angiolymphoproliferative lesion was found in the sclera.

Three weeks after treatment, fundus imaging showed a significant reduction in the exudative detachment in the macular area and inferiorly. Three months after treatment, the retinal detachment resolved almost entirely, remaining visible only as residual subretinal fluid under the fovea at OCT (Fig. 7). BCVA improved to 20/80, and the IOP was stable under medical treatment. For 2 years, the functional results were stable with a BCVA of 20/80 and a normal IOP under medical treatment, despite a minimum persistent retinal detachment involving the fovea.

After 25 months, the retinal detachment recurred. A review of the previous sclerectomy was performed

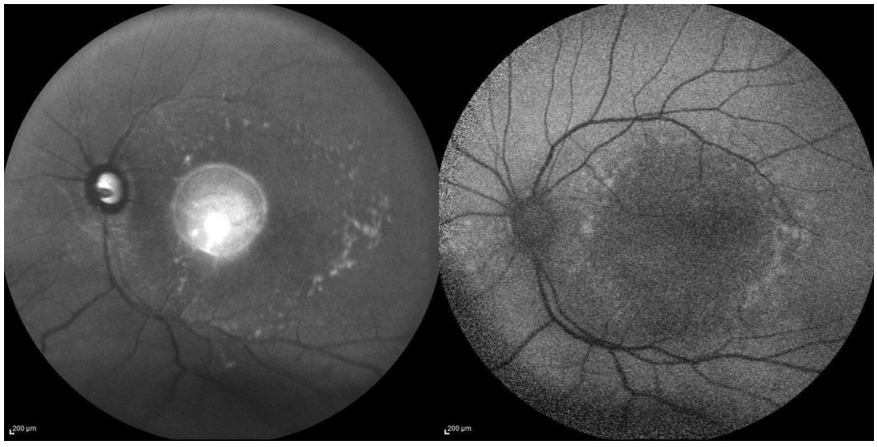

Fig. 1 Fundus IR photograph showing multiple flecks-like spots around the fovea with a circular pattern and fundus AF showing a hypoautofluorescence focused within the spots delimited area

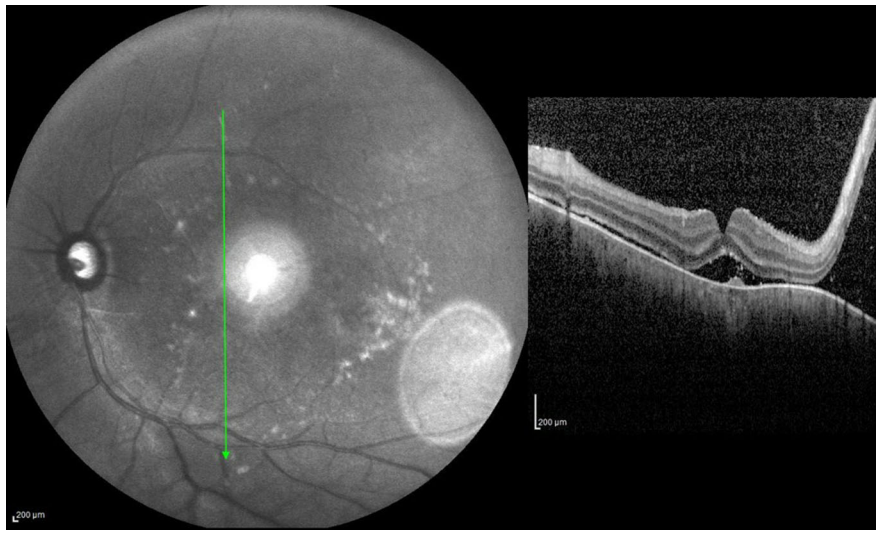

Fig. 2 OCT indicating an inferior retinal detachment of the fovea and a sub-foveal nodular formation under the RPE layer

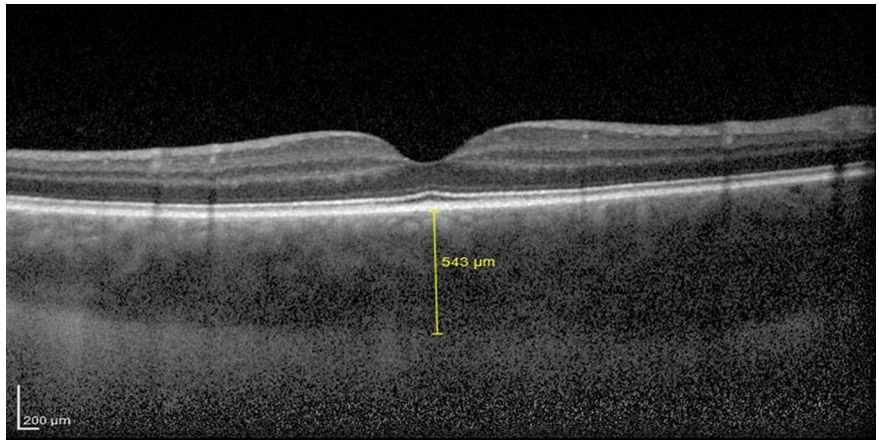

Fig. 3 The choroid thickness in the RE

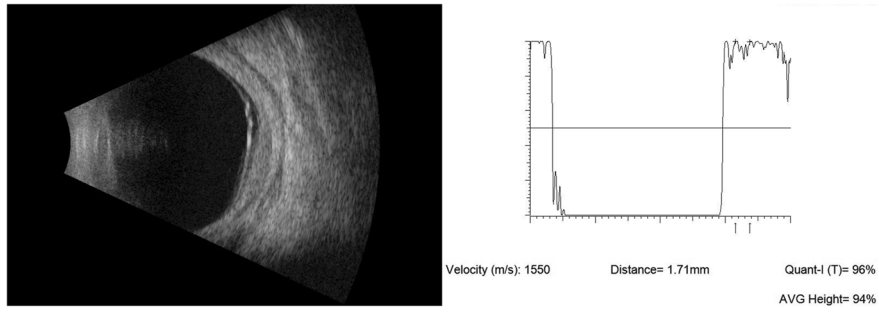

Fig. 4 A-scan and B-scan ultrasonography confirming a lower retinal detachment and reveal a thickened choroid and a thickness increased in correspondence of the hemangioma

to allow the drainage of the subretinal fluid. The retinal detachment resolved within 1 month, with recovery of preoperative BCVA (Fig. 8).

To avoid recurrence of retinal detachment, and becoming the child 7 years old and more compliant, photodynamic therapy was applied to treat the choroidal hemangioma. A standard dose of verteporfin based on weight was performed with an exposure time of 83 s.

After 2 months, no retinal detachment was observed.

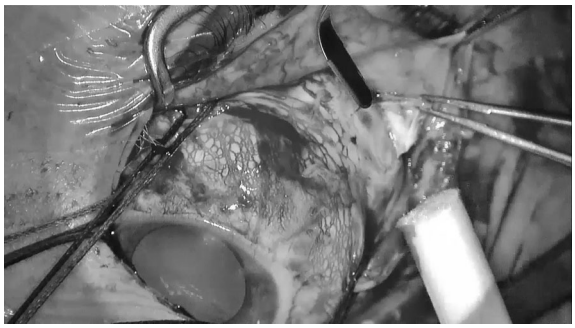

Fig. 5 Intraoperative images showing the creation of scleral flap

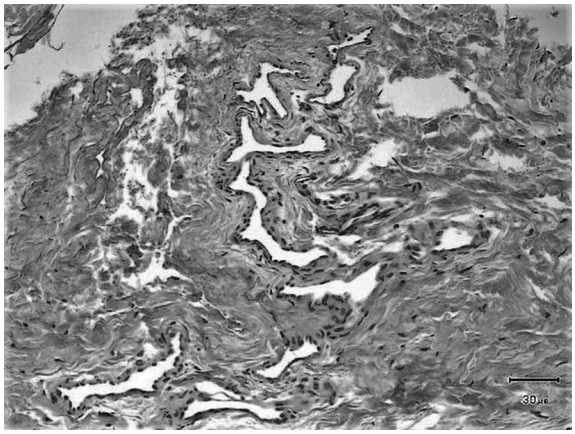

Fig. 6 Histological image of the scleral flap colored with hematoxylin-eosin. Scleral vascular proliferation compatible with hemangioma

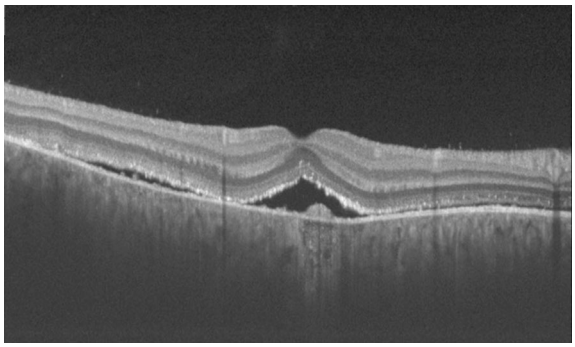

Fig. 7 OCT 3 months after surgery showing a residual subretinal fluid under the fovea

After a follow-up of 1 year, there was no recurrence of retinal detachment.

No side effects after treatment were observed.

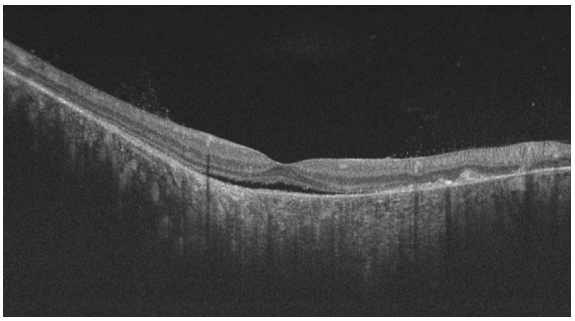

Fig. 8 OCT 1 month after the review of the previous sclerectomy

Discussion

Approximately one-third of the patients affected by Sturge–Weber syndrome have ocular abnormalities, secondary to venous malformation, often at the level of the choroid [1]. Ocular pathology associated with Sturge–Weber syndrome includes diffuse choroidal hemangioma, exudative retinal detachment, macular edema, pigmentary changes within the RPE, subretinal fibrosis, and orange pigment changes. Sturge–Weber syndrome patients are also at risk of glaucoma due to either anterior chamber angle mal-development or raised episcleral venous pressure [2].

Accepted diagnostic modalities include ophthalmoscopy, visual field, fundus fluorescein angiography (FA), indocyanine green (ICG) angiography, ultrasonography, autofluorescence (AF), and optical coherence tomography (OCT) with enhanced depth imaging (EDI) [3]. As the diagnosis was made using those techniques, the authors decided not to perform magnetic resonance imaging (MRI), even if it could be a useful technique to distinguish between malignant and benign tumors affecting the choroid [4].

Exudative retinal detachment associated with Sturge–Weber syndrome is a condition with a primary characteristic: It is generated by uveal effusion, with accumulation of serum under the neurosensorial layers. This condition is difficult to manage and often follows a relapsing course.

There are different treatments proposed for symptomatic serous retinal detachment associated with choroidal hemangiomas including photodynamic therapy (PDT), plaque brachytherapy, external beam and proton beam radiation, stereotactic radiosurgery, transpupillary thermotherapy, laser photocoagulation, oral propranolol and anti-VEGF injections [3].

All of them are reported in limited case series and carry potential complications and side effects. External beam radiation therapy is often inefficient and needs additional radiation therapy, which carries the risk of radiation retinopathy, cataract, and optic neuropathy [3].

Photodynamic therapy is not easily applied to a 5-year-old patient, and the potential side effects of verteporfin or anti-VEGF in children are still almost unknown.

In this case, histological examination of the removed scleral flaps showed reactive telangiectasias affected by mild-to-moderate perivascular infiltrates supported by neutrophils associated with sporadic eosinophils (Fig. 6). No angiolymphoproliferative lesion was found in the sclera. These features indicated vascular changes, secondary to irritative exogenous factors of possible pharmacological or reactive nature not further classifiable.

The rationale associated with the use of this surgical technique was inspired by the observation that this technique is applied in other exudative diseases such as uveal effusion syndrome (UES) [5].

In the pathogenesis of UES, some components that induce abnormalities of the sclera are reported: scleral protein permeability, reduced scleral hydraulic conductivity, vortex vein compression, increased choroidal vessel permeability and chronic choroidal inflammation [6]. These factors are related to increased scleral thickness, similarly to our case.

The authors suspected that the abnormal scleral thickness of the child might increase the resistance to the trans-scleral outflow of intraocular fluid, thus causing, as in UES, an exudation of subretinal liquid [7]. We believe that sclerectomies may work by two different mechanisms. First, the sclerectomy facilitates the drainage of uveal exudation in the immediate postoperative period. Then, more importantly, it seems to decompress vortex veins indirectly by relaxing the scleral tension [6].

To summarize, drainage sclerectomies resulted in the significant improvement of exudative retinal detachment and a subsequent improvement in visual acuity. The effect may come from indirect decompression of vortex veins by relaxing scleral tension. This maneuver allows to maintain the visual acuity. In small children or non-collaborative subjects to the PDT, sclerectomy or scleral drainage is an option to preserve the visual function. In this case, the intervention allowed to reach the age when it was possible to do PDT with dry retina.

The normal dose of verteporfin by weight and 83 s of macular application was sufficient.

In conclusion, our data suggest that this is an effective surgical option for the management of exudative retinal detachment clinically significant associated with diffuse choroidal hemangioma in the Sturge–Weber syndrome.

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