Ruthenium-106 Plaque Brachytherapy for the Treatment of Diffuse Choroidal Haemangioma in Sturge–Weber Syndrome

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A 10-year-old girl with a diffuse choroidal haemangioma was treated with plaque brachytherapy. The outcome was assessed with serial fundus examinations, ultrasound for tumour thickness, and best-corrected visual acuity (BCVA). Treatment with brachytherapy led to complete resorption of subretinal fluid, regression of the haemangioma, and an improvement in the BCVA from 6/60 to 6/15 in 1 month, which improved further to 6/9 in 3 months after treatment.

Key Words: brachytherapy, diffuse choroidal haemangioma, radiotherapy, ruthenium

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Choroidal haemangioma is a benign vascular tumour, which can be circumscribed or diffuse. Haemangiomas of the brain, orbit and the periocular skin coexist with diffuse choroidal haemangioma as part of the Sturge–Weber syndrome.

Diffuse choroidal haemangioma may be associated with an exudative retinal detachment with macular involvement, photoreceptor cell loss and cystoid degeneration of the sensory retina causing visual loss[1] and is classically treated with external beam radiotherapy.[2,3] We report a case of a diffuse choroidal haemangioma with exudative retinal detachment that showed remarkable response to plaque brachytherapy.

Case Report

A 10-year-old girl presented with blurred vision in the right eye of 1-month duration. On examination, her best-corrected visual acuity (BCVA) was 6/60 in the right eye and 6/6 in the left eye. She had diffuse nevus flammeus involving the right upper eyelid, cheek and the nose suggestive of Sturge–Weber syndrome. Anterior segment was normal. There was no evidence of glaucoma. Fundus examination showed total exudative retinal detachment and diffuse choroidal haemangioma. There was a nodular elevation arising from the diffuse choroidal haemangioma measuring about 12 mm in greatest diameter, localised to the posterior pole [Figure 1A]. Ultrasound B-scan confirmed diffuse thickening of the choroid. The tumour was thickest at the posterior pole measuring 5.25 mm [Figure 2A]. Computed tomography scan of the brain and systemic evaluation were unremarkable.

The tumour was treated with a notched 20 mm diameter Ruthenium-106 plaque (Bebig Isotopen, Germany) centered on the nodular elevation with a tumour apex dose of 3000 cGy,

dose rate of 68 cGy/h and a treatment time of 44 h.

At 1-month posttreatment, the maximum thickness of the haemangioma had decreased to 2.57 mm with complete resorption of the subretinal fluid [Figures 1B, 2B]. The BCVA was 6/15. After 3 months, the BCVA was 6/9 and the tumour was nearly flat. No radiation-induced complications were noted after 22 months following brachytherapy and the visual acuity was stable.

Discussion

Mac Lean and associates were probably the first to use brachytherapy for the treatment of choroidal haemangioma.[4] They used transscleral diathermy with scleral suturing of radon seeds. Episcleral plaque brachytherapy is currently considered an effective treatment for large circumscribed choroidal haemangiomas with subretinal fluid.[5,6] The safety and efficacy of Cobalt 60 and Iodine 125 plaques has been demonstrated.[5,6]

Lens-sparing external beam radiotherapy has been recommended for diffuse choroidal haemangiomas with subretinal fluid.[2,3] However, there is slow absorption of subretinal fluid usually taking several months.[2] Often, there is a recurrence or persistence of the fluid necessitating additional external beam radiotherapy with the risk of cataract, radiation retinopathy and optic atrophy.[2]

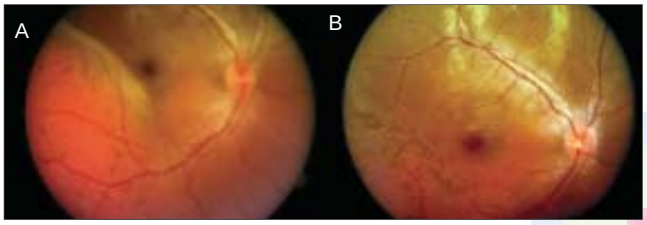

Figure 1: (A) Fundus photograph showing diffuse choroidal haemangioma with a nodular elevation and exudative retinal detachment at initial presentation.(B) Three months posttreatment fundus photograph showing regression of the tumour and resolution of the subretinal fluid

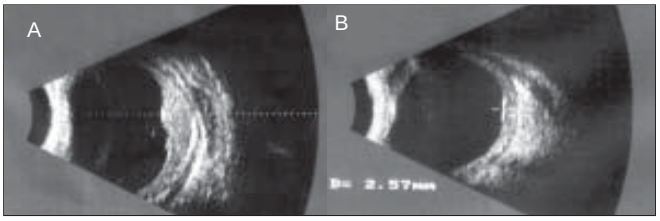

Figure 2: (A) Ultrasound showing the elevated nodular component of diffuse choroidal haemangioma before treatment. (B) Regression of the nodular component 1 month after treatment

As far as could be ascertained (medline search) there is only one report in the literature treating diffuse choroidal haemangiomas with plaque brachytherapy. Zografos and associates used Cobalt-60 applicators in two patients with diffuse choroidal haemangiomas.[5] They found complete resolution of the subretinal fluid and tumour regression in these cases with results comparable to the circumscribed form.

The advantage of plaque brachytherapy is that it delivers precise focal radiation to the target area with minimal damage to the normal ocular structures. We used a large diameter plaque with a notch to accommodate the optic nerve and centered it on the thickest portion of the diffuse choroidal haemangioma. In our patient, the subretinal fluid was completely resolved in 1 month with consequent improvement in visual acuity. The diffuse choroidal haemangioma showed rapid regression in thickness demonstrable on ultrasound. The rapidity of clinical response and lack of complications was impressive. Plaque brachytherapy may thus prove to be a viable treatment option for diffuse choroidal haemangiomas with subretinal fluid in patients with Sturge–Weber syndrome.

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