·Clinical Research·

Treatment review of sight threatening circumscribed choroidal haemangioma

James FT Li Yim[1], Teresa Sandinha[1], Jan M Kerr[1], Diana Ritchie[2], Ewan G Kemp[1]

[1]Tennent Institute of Ophthalmology, Glasgow, Scotland, UK
[2]Beatson Oncology Centre, Glasgow, Scotland, UK
Correspondence to: James FT Li Yim. Tennent Institute, Gartnavel General Hospital, 1053 Great Western Road, Glasgow G120YN, Scotland, UK. jamesly@doctors.org.uk
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Abstract

• AIM: To describe our clinical experience in treating circumscribed choroidal haemangioma (CCH) in a tertiary referral centre over a fifteen year period prior to photodynamic therapy.

• METHODS: The departmental database and photographic records of a tertiary referral center were used to identify patients who were treated for CCH between 1992 and 2007. Their case records were reviewed.

• RESULTS: Visual acuity improved (>2 Snellen lines) in eleven patients (69%) remained stable in one patient (6%) and deteriorated in four patients (25%). Six of the seven treated with brachytherapy and three of the four treated with transpupillary thermotherapy achieved better visual acuity after treatment. 86% of patients treated within six months of onset of symptoms and 50% of patients treated after six months of onset of symptoms noted an improvement in visual acuity. Only one patient in our series had a final VA of 6/60 or worse. Mean follow-up was thirty-five months.

• CONCLUSION: Visual outcome is better when treatment is performed within 6 months of symptoms. The majority of patients achieved an improvement in visual acuity without any adverse effect following treatment.

• KEYWORDS: circumscribed choroidal haemangioma; plaque brachytherapy; transpupillary thermotherapy
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INTRODUCTION

Choroidal haemangioma are benign vascular tumours that can be classified as circumscribed or diffused, the latter occurring in association with the Sturge-Weber syndrome. Clinically, a circumscribed choroidal haemangioma (CCH) appears as an amelanotic orange-red elevated mass on funduscopy. Ultrasound findings are high internal reflectivity on A-scan, dome shaped elevation and acoustic solidity on B-scan. Fluorescein angiography typically shows hyperfluorescence in the early prearterial phase and staining in the late phase. CCH are rare and can remain asymptomatic. However, progressive enlargement may occur [1-3] with ensuing visual impairment caused by exudative retinal detachment, retinal oedema, RPE changes or choroidal fibrosis. Many treatments have been used for symptomatic CCH such as cryotherapy, xenon arc or argon photocoagulation (PC) [4], transpupillary thermotherapy (TTT) [5-8], brachytherapy (PL) [9-12], proton beam therapy (PBT) [13-15], external beam radiotherapy (EBT) [12,16], stereotactic radiotherapy [17] and more recently photodynamic therapy (PDT) [18-23]. The treatment modalities previously used in our department for CCH were: transpupillary thermotherapy and radiotherapy in the form of ruthenium 106 scleral plaque brachytherapy, proton beam therapy and external beam therapy.

Transpupillary thermotherapy uses a diode laser emitting at 810nm with a broad beam and raises the temperature in the treated tissue to 65℃ at the apex and 40℃ internally. This is the critical temperature that induces ischemic necrosis. Complications include cystoid macular oedema, preretinal fibrosis and focal iris atrophy [5], and possibly branch retinal vascular occlusion [24]. Low dose radiation, less than or equal to 20Gy appears successful in controlling both circumscribed and diffuse choroidal haemangiomas associated with progressive retinal detachment. Lens sparing treatment avoids the risk of cataract. External beam therapy and brachytherapy have not been directly compared but brachytherapy appears to be effective for selected well circumscribed lesions. With brachytherapy the radiation dose is prescribed to the apex of the tumour. This results in corresponding higher surface dose due to exponential attenuation of radiation dose at depth. Treatment depth of 5mm results in a maximum surface dose in the order of

50Gy and very low risk of any late radiation toxicity except for juxtapapillary tumours where there is a risk of radiation dose to the optic disc and proximal optic nerve. For these cases external beam radiation is preferable. Complications of radiation treatment that have been reported are: radiation retinopathy[13] and cataract[25].

Photodynamic therapy (PDT) is currently the treatment for posteriorly located CCH. However, the long term results are not yet known and anteriorly located lesions cannot be targeted with PDT. The aim of this paper is to describe our clinical experience in treating sight threatening circumscribed choroidal haemangioma with other modalities from 1992 to 2007.

MATERIALS AND METHODS

Subjects Twenty-one patients were identified with a diagnosis of CCH. Two of them were lost to follow up; three did not require any treatment and remain under review. Sixteen patients received treatment (eleven males and five females; age range 14-75 years, mean 45 years old). Seven had brachytherapy, four had transpupillary thermotherapy, one proton beam therapy, two external beam therapy, one TTT followed by PL, 1 EBT followed by TTT. The most common indication for treatment was decreased visual acuity (twelve patients). Other indications were: progressive field defect in two patients and increase in size of CCH in one patient. The pre-treatment visual acuity ranged from 1/120 to 6/5. After a mean follow-up of 35 months, best corrected post treatment visual acuity ranged from 1/60 to 6/5.

Methods We performed a retrospective review of patients case records diagnosed with CCH between 1992 and 2007. Patients were identified through the departmental database and photographic records. Case records were reviewed for data collection. Location of CCH, indication for treatment, duration of symptoms prior to diagnosis, visual acuity (VA) pre-treatment and best post-treatment, time interval from diagnosis to treatment and treatment modalities were recorded.

This cohort of patients was referred to our tertiary centre with a suspicious choroidal lesion or a CCH requiring further treatment not available locally. Diagnosis of CCH is made clinically following fundus examination, B scan ultrasonography and fluorescein angiography. Treatment is recommended when there is visual impairment or threat due to tumor location and associated subretinal fluid if present.

Choice of treatment was based on accessibility-TTT for posterior pole lesion, PL for anteriorly located lesions not accessible with TTT up to 5mm thick and PBT or EBT for lesions thicker than 5mm. TTT and PL were administered in our department while EBT at the Beatson Oncology Centre. PBT is performed at the Douglas Cyclotron Unit, Clatterbridge Centre for Oncology, Cheshire.

Ethical approval was sought from the West of Scotland Research Ethics Committee and was deemed not required as this was a retrospective study. The tenets of the Declaration of Helsinki were observed.

RESULTS

Visual acuity improved (defined as a gain of two or more lines on Snellen chart) in eleven patients (69%) following treatment and one patient (6%) retained the same level of visual acuity. Four patients (25%) experienced a deterioration in their vision (loss of one or more lines) after treatment. Of the seven patients treated with brachytherapy alone, only one experienced a decrease in visual acuity while the others all noted an improvement in visual acuity. Among those four treated with TTT alone, only one patient noted a reduction in visual acuity. Only one patient in our series had a final VA of 6/60 or worse. This was an untreated patient who was referred with and retained a visual acuity of 1/36 and then lost to follow-up.

Six out of seven patients (86%) treated within 6 months of onset of symptoms compared to three out of six patients (50%) treated after 6 months of onset of symptoms noted an improvement in visual acuity following treatment.

Duration of symptoms prior to diagnosis ranged from two to eighteen months. Time to treatment from diagnosis ranged from same day to 24 months whereas time to treatment from onset of symptoms ranged from 2 to 28 months. No complications were reported following any of the forms of treatment used in this series after a mean follow-up of 35 months.

DISCUSSION

From our experience, it appeared that more patients had a better outcome when treated within six months of onset of symptoms compared to those treated six months after onset of symptoms (86% versus 50%). Similarly, in the series of Shields *et al* [26], 42% of patients treated within six months and 72% of those treated after six months from the onset of symptoms had a poor visual outcome.

In terms of treatment modalities, an improvement of 2 or more lines was noted in 75% of those treated with TTT alone and 86% with brachytherapy alone. Overall, an improvement in visual acuity was noted in 69% of our patients with only one patient having a visual acuity of less than 6/60 after a mean follow-up of 35 months. Although our retrospective study involved a small population referred to a tertiary centre which precludes strong conclusions, this

Table 1 CCH outcome following TTT or PL treatment

Author	*n*	Treatment	VA improvement (≥2 Snellen lines) (%)	Mean follow-up (months)
Current study	4	TTT	75	35
Garcia-Arumi *et al*[6]	8	TTT	50	12
Kamal *et al*[7]	6	TTT/ICG	67	2-12
Fuchs *et al*[8]	10	TTT	40	3-13
Gündüz *et al*[5]	10	TTT	50	8-44
Current study	7	Ruthenium 106 (Ru 106)	71	35
Madreperla *et al*[11]	8*	Ru 106	75	25
Aizman *et al*[9]	5	Palladium 103	60	19
Zografos *et al*[10]	31	Cobalt 60	81	24

*Two patients treated with Iodine 125 plaque

Table 2 Published series of visual outcomes for treatment of CCH

Author	*n*	Treatment	Improved VA (%)	Worse VA (*n*)	VA < 6/60 (*n*)	Mean follow-up (months)
Zografos *et al*[10]	31	Cobalt PL	81	19	6	24
Hannouche *et al*[13]	13	PBT	62	0	31	26
Madreperla *et al*[11]	23	PC , PL(8) , EBT	61	9	NA	66
Schilling *et al*[16]	36	EBT	39	22	NA	54
Lee *et al*[14]	3	PBT	33	0	2	24
Zografos *et al*[15]	31	PBT	71	NA	3	12
Shields *et al*[26]	96	PC, TTT(3), PL(15), EBT(2)	86	30	62	3 *
Aizman *et al*[9]	5	Palladium PL	60	0	1	19
Current study	16	TTT, PL, PBT, EBT	69	4	1	35

*Patients followed up for at least 3 months, mean follow-up not available

review of our outcomes compares favourably with those reported in the literature (Table 1, 2) (TTT/ICG-Transpupillary thermotherapy with indocyanine green enhancement)

We currently recommend treatment within 6 months of onset of symptoms with PL for anteriorly located lesions not accessible with PDT. The visual outcome is influenced by the location of the haemangioma and the duration of symptoms prior to treatment. The majority of patients may expect to retain or achieve some improvement in their vision following treatment. No side effects were observed after a mean follow-up of 35 months. This paper adds to the small number of patients who benefited from Ruthenium plaque brachytherapy for CCH reported in the current literature.

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