ORIGINAL RESEARCH PAPER

Radiotherapy

RADIATION THERAPY IN CHOROIDAL HEMANGIOMA: BOON OR BANE

KEY WORDS: Choroidal Hemangioma, Radiation Therapy, Struge Weber Syndrome

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ABSTRACT

Choroidal hemangiomas (CH) are extremely rare benign ocular tumors that frequently develop in the posterior fundus. It manifests as two types diffuse (commonly seen in Sturge Weber syndrome) and circumscribed. Most of them remain asymptomatic until adulthood, and they can be identified during a routine examination or when they cause noticeable visual impairment. It is usually treated using a variety of therapeutic approaches including photocoagulation, transpupillary thermotherapy and anti-VEGF agents. External beam radiotherapy (EBRT) is used in patients with advanced disease with retinal detachment (RD) or in cases refractory to local therapies. EBRT results in resolution of tumor thickness and sub-retinal fluid thereby improving the visual outcomes. This case series highlights the role of EBRT as non-invasive and effective modality in managing choroidal hemangioma.

INTRODUCTION

Choroidal hemangiomas are benign vascular tumors which may present with diagnostic and therapeutic challenges due to their variable presentation and potential to cause vision threatening complications. They manifest either as circumscribed choroidal hemangioma or diffuse choroidal hemangioma. Circumscribed hemangiomas usually affect young to middle aged adults and are localized and often present with visual symptoms such as blurred vision and metamorphopsia. Diffuse hemangiomas are commonly associated with systemic conditions like struge weber syndrome, a neurocutaneous disorder characterized by facial port wine nevus, leptomeningeal angiomatosis, glaucoma, neurological deficits such as seizures and hemiplegia. These tumors can cause exudative retinal detachment, macular edema and significant visual loss if not treated.

The pathophysiology involves vascular malformations within the choroid leading to chronic subretinal fluid accumulation and exudative retinal detachment. Imaging modalities such as ultrasonography (USG Bscan), optical coherence tomography (OCT), flouroscein angiography and MRI are essential for diagnosis and monitoring.

Traditional management includes laser photocoagulation, transpupillary thermotherapy, photodynamic therapy and intra vitreal anti VEGF agents depending on tumor size and location. In recent years EBRT has emerged as a viable non invasive option for larger or refractory cases with an advantage of vision preservation.

Case Summary

CASE 1- A 10 year old male presented with progressive vision loss in the right eye. Ophthalmic examination revealed signs of exudative retinal detachment. MRI of the orbit showed diffuse thickening of the choroid in the right eye associated with extensive subretinal fluid and lesion measuring 6.2mm in maximum thickness. The patient had characteristic facial port wine stain over right upper half of face and neurologic findings consistent with struge weber syndrome. A diagnosis of diffuse choroidal hemangioma was made. Given the extent of retinal detachment and fluid accumulation, the patient was planned for external beam radiotherapy. He received 30Gy in 15 fractions using IMRT with precise conformal targeting and sparing of adjacent structures. Post treatment MRI showed complete resolution of the subretinal fluid and choroidal thickening was markedly reduced. Visual acuity improved from poor perception to 3/60. The patient tolerated treatment well without radiation complications.

CASE 2- A 12 year old male was diagnosed with circumscribed choroidal hemangioma in the right eye, presenting with total retinal detachment and decreased vision. MRI revealed a well circumscribed crescentic shaped intraocular lesion measuring 1.5x0.5x1.6cm is seen in the temporal aspect along the choroidal layer of right eye with retinal detachment and hemorrhage. The patient received a dose of 30Gy in 15 fractions with IMRT technique. Post therapy imaging showed complete retinal reattachment and clinical evaluation confirmed stabilization of visual acuity. The treatment was well tolerated and no ocular toxicities were observed.

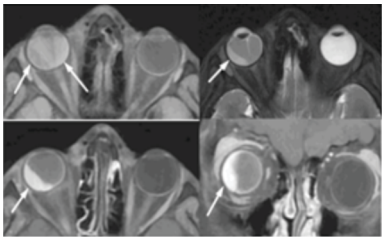

Figure 1: MRI showing crescentic shaped lesion in the temporal aspect along choroidal layer of right eye and subretinal hemorrhage with in the globe

CASE 3- A 17 year old adolescent presented with a posterior circumscribed choroidal hemangioma in the left eye. MRI findings included a crescent shaped well enhanced mass at the posterior pole, with associated subretinal fluid and retinal detachment with a maximum lesion thickness of 4.5mm. The location of the lesion is close to the macula, made local therapies less feasible. The patient was treated with IMRT, receiving 35Gy in 18 fractions. Follow up imaging demonstrated regression of the lesion and resolution of subretinal fluid. No radiation induced complications were noted.

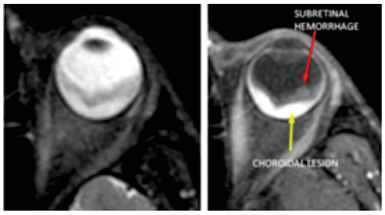

Figure 2: MRI showing well defined homogenously enhancing lesion in retinochoroidal layer of left globe along the posterior aspect.

Radiation Planning and Contouring

All patients underwent CT simulation with 1mm slices fused with MRI for precise tumor and organ at risk (OAR) delineation. Immobilization was achieved with custom thermoplastic masks.Target volumes were defined as follows

- GTV(GROSS TUMOR VOLUME)-Visible lesion on MRI and B Scan
- CTV(CLINICAL TARGET VOLUME)-GTV+margin for microscopic spread, typically includes entire choroid
- PTV(PLANNING TARGET VOLUME)-CTV+ set up error margin(3mm)

Organ at risk	Tolerance dose	Case1	Case2	Case3
Optic Nerve	<50-54Gy	30Gy	25Gy	27Gy
Retina	<45Gy	37Gy	30Gy	30Gy
Cornea	<40-45Gy	14Gy	26Gy	27Gy
Lacrimal Gland	<30-35Gy	32Gy	29Gy	30Gy

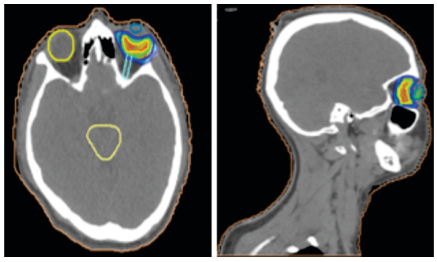

Figure 3: The axial and saggital CT slices showing the conformal dose distribution using IMRT technique.

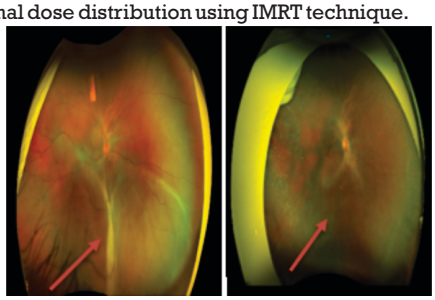

Figure 4A: OPTOS image showing retinal detachment.
Figure 4B: 6 months post RT OPTOS imaging shows complete resolution of retinal detachment.

DISCUSSION

External beam radiotherapy has emerged as a safe and effective non invasive alternative. It promotes tumor regression and reattachment of retina by reducing vascular permeability. Shields et al(2001) reported outcomes of 44 patients treated with EBRT(20-30Gy), achieving subretinal fluid resolution in 97% and visual acuity improvement in 77% . Shukla et al(2014) presented a series of cases treated with 30Gy in 15 fractions. They observed complete fluid resolution and visual improvement in 75% of patients with no long term radiation complications during follow up.Arepall et al (2014) also reported EBRT outcomes in both pediatric and adolescent patients with choroidal hemangioma particularly in cases associated with struge weber syndrome which confirmed good anatomical and visual outcomes with minimal toxicity using 20-30Gy.

CONCLUSION

External beam radiotherapy is well tolerated, non invasive and effective treatment modality for choroidal hemangiomas with exudative retinal detachment particularly when other localized therapies fail or contraindicated. Our case series supports its role in pediatric patients demonstrating not only anatomic resolution but also functional visual improvement. The use of lens sparing IMRT further minimizes radiation exposure to adjacent critical ocular structures.

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