Brief Report

Management Of Choroidal Neovascularisation In Choroidal Osteoma

Introduction

Choroidal osteoma is believed to be first described at the 1975 Meeting of Verhoeff Society but the term choroidal osteoma was coined by Gass in 1978 when he described four healthy young women with characteristic ophthalmoscopic findings of slightly elevated, yellowish, choroidal tumour with sharp geographic borders. These tumours demonstrate evidence of bone formation in the choroid and are believed to be choristomatous in origin. It is commonly juxtapapillary or peripapillary, but may extend to the macula. It is rare that it would be found only in the macula. The shape is commonly oval or round with well defined scalloped or geographic margins. Occasionally decalcification can occur and is characterized by thin, atrophic, yellow-gray regions with associated RPE atrophy. Decalcification can occur spontaneously or as a result of laser photocoagulation or PDT. Choroidal neovascular membranes (CNVM) can also develop. The majority of patients with choroidal osteoma maintain good vision. Long term poor visual acuity in patients with choroidal osteoma is associated with subretinal fluid, RPE alterations, and subretinal hemorrhage from choroidal neovascularization. In a follow-up study of 36 patients, the probability of loss of visual acuity (20/200 or worse) was more than 50% by 10 years. Choroidal neovascularization is the most frequent cause of visual loss in choroidal osteoma with more than half of the patients expected to develop choroidal neovascularization. Management of choroidal neovascularisation in this entity has been difficult and various modalities of treatment have been reported with variable success. Combination therapy is likely to be an effective therapy in the management of this often resistant CNVM and very reports are available in the literature. This case report is on combination therapy in the management of CNVM in a young patient with choroidal osteoma.

Case report

A 25 year old male patient presented to us with complaints of metamorphopsia and defective vision in his left eye of two weeks duration. There was no associated pain or redness. He had no known systemic illness. On examination his best corrected visual acuity in the right eye with -2.00 DCyl 900 was 6/6, N6 and in the left eye with -0.50 DCyl 900 was 6/12, N9. Colour vision was normal. Amsler grid examination of left eye revealed wavy gridlines infero-temporal to fixation. Anterior segment was unremarkable. Intraocular pressure was 16 mm Hg in right eye and 14 mm Hg in the left eye. Right eye fundus was normal. The left eye showed a well-circumscribed orange-red lesion around 5 DD in size, superotemporal to the optic disc with the inferior edge adjacent to the superior disc margin. Subretinal haemorrhage was present near the inferotemporal edge of the lesion just superior to the fovea with sub-retinal fluid (SRF) spreading to the fovea (Fig 1).

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Fig 3; HRA FFA showing the classic extrafoveal CNVM with early bright well defined hyperfluorescence (A) and intense late leakage (B).

Fig 4; HRA ICG imaging showing the relative hypofluorescence of the osteoma(A) with late staining (C) and the classic hot spot extrafoveally with an adjacent point of increased fluorescence (B) seen clearly in the late phases (D).
Spectral domain HRA OCT revealed thickening above the RPE at the site of CNVM and shallow subretinal fluid at the fovea (Fig 2). B Scan revealed highly echogenic lesion 7.52 mm in diameter and 1.94mm in thickness above the optic disc with significant back shadowing suggestive of calcification. A-scan ultrasonography showed a high spike corresponding to the anterior surface of the lesion suggestive of choroidal osteoma (Fig 5). Simultaneous FFA + ICG angiography was done. The lesion showed persistent hypofluorescence corresponding to the osteoma with an area of early well defined hyperfluorescence that increased towards the later phases in FFA suggestive of CNVM (Fig 3). The ICG showed another small hotspot in ICG near the inferior edge of the primary hyperfluorescent lesion (Fig 4). A diagnosis of Choroidal Osteoma with secondary predominantly classic extra-foveal choroidal neovascular membrane (CNVM) was made. Intravitreal bevacizumab 1.25 mg/0.05 cc was injected under sterile precautions and 2 weeks later focal laser was done for the extrafoveal CNVM. On subsequent follow up after one month, Fundus examination showed scarred CNVM and OCT revealed complete resolution of SRF and retinal oedema (Fig 6). Subsequently he was kept under observation with Home Amsler monitoring. The Vision remained stable at 6/6, N6 with correction, with regressed CNVM (Fig 7) and static osteoma until the completed 12 months follow up.
Discussion

The etiology of the choroidal osteoma is unknown. Factors implicated in its development, however, include inflammation, trauma, hormonal state, calcium metabolism, environment, and heredity. None of these factors appear to be either a sole, or an established, factor in causing patients to develop the condition. For instance, the hormonal hypothesis does not explain why males are affected by the condition or why these lesions can be observed in pre-pubertal patients. It has been postulated that choroidal osteoma is a choristoma, i.e., normal tissue arising at an abnormal location, but this explanation begs the question of why females are affected more frequently than males and why there is continuous development and growth of the lesion in adulthood. No consistency has been established with serum calcium, phosphate, or alkaline phosphatase levels. The reasons for vision loss from choroidal osteoma include CNV, subfoveal fluid, and photoreceptor degeneration. The cause of CNV development is unknown. It has been hypothesized that the thinned, degenerated retinal pigment epithelium overlying the osteoma allows the growth of new blood vessels.

Several treatments are tried but with limited success. Laser photocoagulation of CNV associated with choroidal osteoma was less effective owing to depigmentation of RPE that often reduces the absorption of laser energy. The surgical removal of subfoveal CNV has been performed successfully, but the visual result has been poor. Recently, PDT and transpupillary thermotherapy have been tried, but the visual result are variable. Intravitreal bevacizumab has been given and improvement of visual acuity and regression of CNV was observed in a few reports. Ranibizumab has also recently been used successfully for the treatment of CNV secondary to choroidal osteoma either as monotherapy or as part of combination therapy.

As monotherapy with anti-VEGF agents have not been effective in all series and because these CNV are often extrafoveal and PDT is an expensive therapy in our part of the world, combination of anti-VEGF therapy with focal laser would be an effective strategy in the management of this pathology. We have used a combination of intravitreal bevacizumab and focal laser for the treatment of CNV secondary to choroidal osteoma, and observed regression of CNV and recovery of visual acuity. This is the first report of such a combination for treating CNV related to choroidal osteoma. We propose that combination of intravitreal bevacizumab and focal laser may be a cost effective and useful treatment in extrafoveal CNV secondary to choroidal osteoma. However long-term follow-up and further studies are required to confirm the role of combination therapy of intravitreal bevacizumab and focal laser in these resistant CNV. Also subfoveal CNV cannot be treated with conventional laser treatment and these patients may require photodynamic therapy.

References


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