INTRODUCTION

Choroidal osteoma is a rare benign tumor. We report a male child diagnosed with bilateral choroidal osteoma, high myopia and secondary choroidal neovascularization (CNV) membrane in one eye. Co-existence of posterior staphyloma made the clinical diagnosis of choroidal osteoma difficult due to the osteoma filling the depression of the posterior staphyloma. On review of the literature we were unable to find a similar case of co-existent choroidal osteoma and posterior staphyloma reported earlier. CNV secondary to choroidal osteoma was treated with intravitreal bevacizumab and it responded well. Regular follow-up is essential for recurrence of CNV and decalcification of the osteoma.

We report a case of bilateral choroidal osteoma-macular in the right eye and peripapillary with macular involvement in the left eye along with high myopia and posterior staphyloma in a male child associated with a CNV membrane in the left eye treated with the off-label use of intravitreal bevacizumab (Avastin; Genentech, San Francisco, CA, USA).

CASE REPORT

A 10-year-old male patient presented with the complaints of decreased vision in the left eye for the last 4 days. On examination, the best corrected visual acuity (BCVA) in the right eye was 20/40, N6 and in the left eye 20/100, N9. Anterior segment examination was normal and the fundus examination of both eyes showed myopic discs with a posterior staphyloma and an extensive orangish-yellow flat lesion in the macular area of the right eye and in the peripapillary and macular area with submacular hemorrhage in the left eye [Figure 1a and b].

Ultrasound B-Scan of both eyes showed a high reflective echo with shadowing at the posterior pole of both eyes. Patient underwent fundus fluorescein angiography (FFA) for both eyes which showed early hypofluorescence with late hyperfluorescence in the macular area of both eyes along with the peripapillary area in the left eye suggestive of the choroidal neovascular membrane.

ABSTRACT

Choroidal osteoma is a rare benign tumor. We report a male child diagnosed with bilateral choroidal osteoma, high myopia and secondary choroidal neovascularization (CNV) membrane in one eye. Co-existence of posterior staphyloma made the clinical diagnosis of choroidal osteoma difficult due to the osteoma filling the depression of the posterior staphyloma. Typical findings on fundus fluorescein angiography, optical coherence tomography, B-scan and indocyanine green angiography confirmed the diagnosis. A review of literature was performed. CNV secondary to choroidal osteoma was treated with intravitreal bevacizumab and it responded well. Regular follow-up is essential for recurrence of CNV and decalcification of the osteoma.

Key words: Bevacizumab, Choroidal Neovascular Membrane, Choroidal Osteoma
osteoma. There was an area of early hyperfluorescence which increased in size and intensity in the macular area of the left eye suggestive of an active CNV [Figure 2a]. The indocyanine green angiography showed macular choroidal hypofluorescence in both eyes with an ill-defined hyperfluorescence plaque seen in the delayed frames of the left eye suggestive of a CNV [Figure 2b]. Spectral domain optical coherence tomography of the left eye showed a steep curvature of the retinal layers due to posterior staphyloma along with a subfoveal CNV [Figure 3].

He was diagnosed to have bilateral choroidal osteoma with an active subfoveal CNV in the left eye. He underwent a loading dose of three intravitreal injections of 1.25 mg/0.05 ml bevacizumab (off-label use) delivered to the inferotemporal quadrant of the left eye under strict aseptic precautions and short general anesthesia after an informed consent was signed by the parents.

At 1 year follow-up after the last injection showed a BCVA of 20/60, N6 in the left eye. Fundus examination of the left eye showed choroidal osteoma in the macula with no evidence of submacular hemorrhage. FFA of the left eye showed staining of the CNV with no evidence of dye leakage.

DISCUSSION

Choroidal osteoma is a relatively rare benign intraocular tumor composed of mature bone that typically replaces the full thickness of the choroid. This tumor classically manifests as an orange-yellow plaque deep to the retina in the juxtapapillary or macular region. Nearly 80% are unilateral though Gass has described unilateral osteomas becoming bilateral. These tumors are said to show signs of evolution - growth of tumor, decalcification and CNV formation causing profound visual loss.

Our patient has an osteoma involving the peripapillary and macular area, which is rare. We missed the diagnosis of the osteoma initially as the lesion appeared flat though the tumor is usually a little elevated. This was due to the tumor filling the depth of the co-existent posterior staphyloma. Based on our experience we recommend other clinicians to remain suspicious for a similar presentation when examining a myopic fundus with a posterior staphyloma. There was a large area of submacular hemorrhage overlying the osteoma, which may predispose to the formation of CNV along with an irregular surface of the tumor.

The possible reasons for the development of the CNV in these cases includes thin degenerated retinal pigment epithelium (RPE) overlying the osteoma allowing the growth of new blood vessels or an extension of the osteoma itself due to the presence of osteoclasts on histopathology.

The various treatments for CNV secondary to a choroidal osteoma include laser photocoagulation, photodynamic therapy (PDT), transpupillary thermotherapy (TTT) and surgical removal of the CNV. Laser photocoagulation especially for an extrafoveal CNV with osteoma, was found to be poorly responsive to laser photocoagulation due to depigmentation of the RPE and degenerated RPE/Bruch’s membrane complex, resulting in the reduced uptake of laser energy. PDT requires multiple treatments and there is a risk of a drop in vision after the treatment. Visual results have been poor with TTT and surgical removal of the CNV associated with osteoma.
In our case, the osteoma induced CNV was treated with a loading dose of three intravitreal bevacizumab injections (off label use) resulting in rapid regression of the CNV with a marked improvement in visual acuity. This treatment shows better visual recovery than the other treatments. Bevacizumab may have better penetration in these cases due to a very thinned and degenerated RPE/Bruch’s membrane complex.\(^8\)

There are reports of CNV secondary to choroidal osteoma in adult patients treated with intravitreal bevacizumab.\(^8,9\) However to the best of our knowledge this is the first report of a choroidal osteoma induced CNV treated with bevacizumab in a young child.

REFERENCES


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