A Case of Rare Choroidal Tumor, Choroidal Osteoma

Shah RK, Byanju R

Bharatpur Eye Hospital

Bharatpur, Chitwan, Nepal.

Corresponding Author

Ritesh Kumar Shah

Bharatpur Eye Hospital

Bharatpur, Chitwan, Nepal.

E-mail: rites_70@hotmail.com

Citation

Shah RK, Byanju R. A Case of Rare Choroidal Tumor, Choroidal Osteoma. *Kathmandu Univ Med J.* 2017;60(4):353-5.

ABSTRACT

Choroidal osteoma is a rare benign tumor of unknown etiology, commonly found in young asymptomatic females in their second or third decade of life. It typically appears as an orange-yellow to yellow-white lesion in juxtapapillary region which can extend over to involve macula. Lesion solely located in macular region is less common. Diagnosis is based on fundoscopic appearance and findings of ancillary tests like B scan ultrasonography, optical coherence tomography (OCT), fundus fluorescein angiography (FFA) and computerized tomography (CT) scan. We report a case of unilateral macular choroidal osteoma in a fifteen-year-old girl with normal vision.

KEY WORDS

Benign tumor, Choroidal osteoma, Macula

INTRODUCTION

Choroidal osteoma, a rare ocular tumor characterized by the presence of bone within the choroid, was first described at the meeting of the Verhoeff society by Van Dyk in 1975.[1] Although the exact etiology is unknown, the origin of the tumor is believed to be choristomatous.[2] It typically presents as unilateral yellowish-white to orange-yellow lesion in juxtapapillary or macular region in young females in their second or third decades of life.[3-5]

Understanding of the incidence and course of this disease is limited due to its rare occurrence and thus management is still a challenge. We report a case of choroidal osteoma in a relatively younger patient of 15 years old in a rather uncommon location.

CASE REPORT

A fifteen-year-old girl was referred to the retina clinic of Bharatpur eye hospital from pediatric ophthalmology department for evaluation of an amelanotic sub-retinal mass in the posterior pole of her left eye. She had been under routine eye check up for evaluation of her refractive error for last two and half years. She had last visited our hospital six months back. Records revealed that her ocular examination then was unremarkable except a slight change in her refractive error. She did not mention any history of recent or chronic illness, recent medication use and prior history of surgery. She also denied any family history of ocular disease.

At presentation, best corrected visual acuity was 6/6, N6 in each eye. Anterior segment examination with slit lamp biomicroscope was unremarkable in both eyes. Intraocular pressure was 19 mm of Hg in RE and 20 mm of Hg in LE. Left eye fundus examination revealed an orange-yellow choroidal lesion in the superior macula, extending under the fovea. The lesion was circular in configuration with irregular, well defined margin of nearly 4 disc diameter in size. (fig. 1) There was no subretinal fluid or hemorrhage and any subretinal membrane. Fundus of the right eye was normal. A and B scan ultrasonography showed a high reflectivity lesion with remarkable posterior shadowing. (fig. 2) Fluorescein angiography showed early hyperfluorescence with late staining of the lesion. There was no increase in the size of hyperfluorescence in the late frame angiogram suggestive of lack of leakage. (fig. 3a-c) OCT of the left eye revealed an elevated retinal pigment epithelium adjacent to the disc with no subretinal fluid or membrane. (fig. 4) Non-contrast CT of orbit (fig. 5) showed 5x6 mm curvilinear calcific region in the posterior aspect of the left globe.

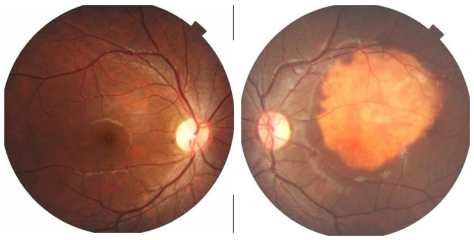

Figure 1. Color fundus photograph of LE showing macular choroidalosteoma and normal RE.

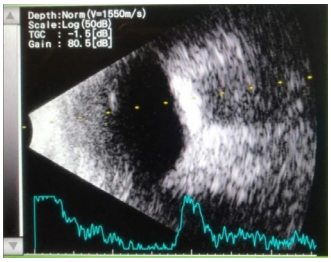

Figure 2. Ultrasonography of LE showing high reflectivity spikes of the lesion causing posterior acoustic shadowing.

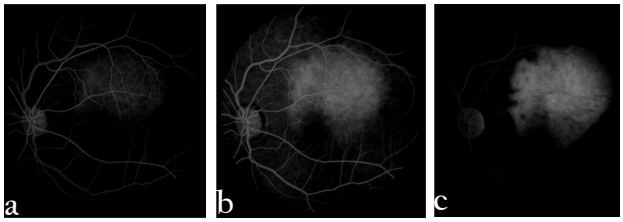

Figure 3. Fluorescein angiogram a) early phase showing patchy hyperfluorescence b) mid phase c) late phase showing late staining without leakage.

A diagnosis of macular choroidal osteoma of left eye was established based on fundus examination, radiological finding and fluorescein angiogram. The diagnosis and its natural course were discussed with the patient and her family.

She was advised for routine follow up, once in every six months, to look for signs of choroidal neovascularisation and other complications. The patient was also educated for self assessment of vision and metamorphopsia.

DISCUSSION

Choroidal osteoma is a rare entity comprising only 4% of all choroidal mass lesions.[6] It is typically seen in healthy, young asymptomatic woman and generally the lesion is found incidentally. Mean age at the time of diagnosis is 26 years. In early stage the lesion is orange-yellow in colour indicating calcified tumor whereas the late lesions appear yellowish-white due to decalcification. Decalcification is usually associated with depigmentation of retinal pigment epithelium (RPE) leading to vision loss.[7] The typical location of the lesion is peripapillary or juxtapapillary region which can extend to the macula. Rarely the mass may be found solely in the macular area as seen in this case.[8,9]

The mechanisms involved in visual impairment in cases with choroidal osteoma are from RPE atrophy overlying the decalcified osteoma, sub retinal fluid (SRF) over the mass

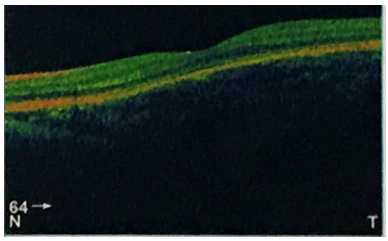

Figure 4. OCT of LE macula showing elvated RPE due to underlying choroidal mass.

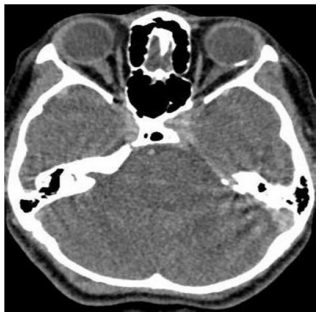

Figure 2. Axial scan CT orbit showing calcific lesion in posterior aspect of left globe.

and most commonly from choroidal neovascularisation (CNV).[10] Aylward and colleagues found that the 10-year probability of CNV and poor visual outcome (VA≤ 6/60) was 47% and 58% respectively.[11]

Combination of fundus appearance, echography, FFA and CT can help in proper diagnosis of the patients with choroidal osteoma. B scan imaging demonstrates highly reflective, acoustically dense lesion with marked acoustic shadowing behind the globe that gives it the appearance of a pseudo-optic nerve.[4,12] CT scan shows a lesion of bone density in the posterior pole of the eye temporal to optic disc. Fluorescein angiography reveals early mottled hyperfluorescence which is followed by late and persistent diffuse hyperfluorescence.[13] OCT may show areas of varying reflectivity depending on the calcification of the lesion. It may also provide information on overlying retina and presence of SRF.[14]

Treatment of choroidal osteoma is indicated when there is documented tumor growth or accompanying complications like choroidal neovascularisation and SRF.

Management of a growing osteoma depends on its specific location. Extrafoveal ones require treatment with Photodynamic therapy (PDT) to induce decalcification and prevent further growth under the foveola and the subfoveal tumors may require calcium supplementation to maintain a calcified mass.[15] Decalcification of osteoma has been seen to be associated with choriocapillaris atrophy, RPE atrophy, photoreceptor loss, and visual acuity loss.[16,17]

Clinically a large number of choroidal lesions can simulate choroidalosteoma. Some of them are amelanotic malignant melanoma, metastatic carcinoma, leukemic or lymphomatous infiltrates, circumscribed choroidalhemangioma, disciform macular scars, and resolving subretinal hemorrhage. Melanomas as compared

to osteoma have more thickness. Similarly metastatic tumor and choroidal nevi lack distinct margin.[5,18] Ophthalmic ultrasound and CT scan shows characteristic calcification in cases with osteoma.

Treatment of CNV with transpupillary thermotherapy (TTT) and photodynamic therapy has been tried with good results. Newer studies have shown that intravitreal injection of Bevacizumab and Ranibizumab have effectively caused regression of CNV secondary to osteoma.[19-22]

Serous retinal detachment overlying osteoma, which is not associated with CNV, is another significant cause of visual loss have also been managed successfully with TTT and intravitreal Bevacizumab.[23,24]

Choroidalosteoma is a rare benign tumor with non-specific clinical picture and hence needs to be identified from malignant tumor to avoid incorrect diagnosis and treatment. Although vision can be normal at the time of diagnosis, as in this case, its long term visual effect cannot be predicted precisely. Due to the rarity of this condition, not much is known about its natural course and long term management. Probably, long term studies with larger sample size would be required to learn about the prognosis and management strategies of this benign but sight threatening condition.

REFERENCES

1. Gass JD, Guerry RK, Jack RL, Harris G. Choroidal osteoma. *Archives of Ophthalmology*. 1978;96:428–35.
2. Williams AT, Font RL, Van Dyk HJ, Riekhof FT. Osseous choristoma of the choroid simulating a choroidal melanoma. Association with a positive 32 P test. *Arch Ophthalmol.* 1978;96:1874-7.
3. Aylward GW, Chang TS, Pautler SE, Gass JD. A long-term follow-up of choroidalosteoma. *Arch Ophthalmol.* 1998;116:1337-41.
4. Shields CL, Shields JA, Augsburger JJ. Choroidalosteoma. *Surv Ophthalmol* 1988;33:17-27.
5. Shields CL, Sun H, Demirci H, Shields JA. Factors predictive of tumor growth, tumor decalcification, choroidal neovascularization, and visual outcome in 74 eyes with choroidalosteoma. *Arch Ophthalmol.* 2005;123:1658-66
6. Liu CC, Kou HK, Tsai MH, Su CC, Kao ML, Chen YJ, et al. Choroidal masses: A fourteen-year analysis. *Chang Gung Med J.* 2001;24:502-11.
7. Chen J, Lee L, Gass JD. Choroidalosteoma: Evidence of progression and decalcification over 20 years. Clin Exp Optom. 2006;89:90-4.
8. DJ. Browning. Choroidalosteoma: observations from a community setting. Ophthalmology. 2003;110:1327–34.
9. Gass JDM. New observations concerning choroidalosteomas. *International Ophthalmology.* 1979;1:71–84.
10. Alameddine RM, Mansour AM, Kahtani E. Review of choroidalosteomas. Middle East Afr J Ophthalmol. 2014;21(3):244-50.
11. G. William Aylward, Tom S. Chang, Scott E. Pautler, Gass JDM. A long-term follow-up of choroidalosteoma. *Arch Ophthalmol.* 1998;116:1337-41.
12. Ide T, Ohguro N, Hayashi A, YamamotoS, NakagawaY, NagaeY et al. Optical coherence tomography patterns of choroidalosteoma. *Am J Ophthalmol.* 2000;130:131 4.
13. Bloom PA, Ferris JD, Laidlaw A, Goddard PR. Appearances of choroidalosteomas with diagnostic imaging. *British Journal of Radiology.* 1992;65:845-8.
14. Empeslidis T, Imrani U, Konidaris V, Mushtaq F, Fotiou P, Kumar P et al. Diagnosis and Monitoring of Choroidal Osteoma through Multimodal Imaging Case. *Reports in Medicine*. 2014. Article ID 393804
15. Shields CL, Materin MA, Mehta S, Foxman BT, Shields JA. Regression of extrafoveal choroidal osteoma following photodynamic therapy. *Arch Ophthalmol.* 2008;126(1):135-7.
16. Shields CL, Perez B, Materin MA, Mehta S, Shields JA Optical coherence tomography of choroidalosteoma in 22 cases: evidence for photoreceptor atrophy over the decalcified portion of the tumor. *Ophthalmology.* 2007;114(12):e53-8.
17. Shields CL, Sun H, Demirci H, Shields JA. Factors predictive of tumor growth, tumor decalcification, choroidal neovascularization, and visual outcome in 74 eyes with choroidalosteoma. *Arch Ophthalmol.* 2005;123(12):1658-66.
18. Honavar SG, Shields CL, Demirci H, Shields JA. Sclerochoroidal calcification: clinical manifestations and systemic associations. *Arch Ophthalmol.* 2001;119:833-40.
19. Sharma S, Sribhargava N, Shanmugam MP. Choroidal neovascular membrane associated with choroidalosteoma (CO) treated with trans-pupillary thermo therapy. *Indian J Ophthalmol.* 2004;52:329-30.
20. Battaglia Parodi M, Da Pozzo S, Toto L, Saviano S, Ravalico G. Photodynamic therapy for choroidal neovascularization associated with choroidalosteoma. *Retina.* 2001;21:660-1.
21. Mercé E, Korobelnik JF, Delyfer MN, Rougier MB. Intravitreal injection of bevacizumab for CNV secondary to choroidalosteoma and follow-up by spectral-domain OCT. Article in French. *J FrOphtalmol.* 2012;35:508-13.
22. Wu ZH, Wong MY, Lai TY. Long-term follow-up of intravitreal ranibizumab for the treatment of choroidal neovascularization due to choroidalosteoma. *Case Rep Ophthalmol.* 2012;3:200-4.
23. Yahia SB, Zaouali S, Attia S. Serous retinal detachment secondary to choroidal osteoma successfully treated with transpupillary thermotherapy. *Retinal Cases Brief Rep.* 2008;2:126-7.
24. Song JH, Bae JH, Rho MI, Lee SC. Intravitreal bevacizumab in the management of subretinal fluid associated with choroidalosteoma. *Retina.* 2010;30:945-51.