Case Report

Recurrence of uveal malignant melanoma: a case report

Thapa M[1], Shrestha GB[1], Sharma AK[1], Karki S[2], Khanal S[1]
[1]Department of Ophthalmology, BP Koirala Lion's Centre of Ophthalmic Studies
[2]Deparment of Pathology
Institute of Medicine, Kathmandu, Nepal

Abstract

Background: Malignant melanoma of uveal tract is a rare ocular malignancy. It is one of the significant causes of ocular morbidity and mortality which is less commonly seen in children. **Case:** We report an unusual case of orbital recurrence of malignant melanoma in a 14-year-old boy who had previously undergone enucleation of the left painful blind eye 8 months ago. He was diagnosed to have uveal malignant melanoma elsewhere which was confirmed by histopathology. Orbital recurrence was managed with modified exenteration with adjuvant chemotherapy and radiotherapy. **Conclusion:** In all treated cases of uveal melanoma, close follow up examination and monitoring is necessary for early diagnosis of the recurrence and to plan for further management.

Keywords: enucleation, uveal melanoma, recurrence

Introduction

Malignant melanoma of the uveal tract is a rare malignancy. It is one of the significant causes of mortality and visual loss. This is more commonly seen in white races in comparison to black population. Approximately 50% of the diagnosed cases of melanoma of choroid and ciliary body die of the disease within two decades. There are some indications that radiation therapy may improve the survival rate (Egan et al, 1998) but survival of metastatic cases is not so good (Egan et al, 1988). Iris Melanoma is detected easily and earlier than ciliary melanoma during routine slit lamp biomicroscopy hence the prognosis of iris melanoma is better than that of the other uveal melanoma.

We report an unusual case of orbital recurrence of malignant melanoma in a 14-year-old boy who had previously undergone enucleation surgery of the left painful blind eye, which was finally diagnosed histopathologically as uveal malignant melanoma.

Received on: 04.01.2013 Accepted on:05.04.2013
Address for correspondence: Dr Madhu Thapa, MD
BP Koirala Lion's Center for Ophthalmic Studies
Maharajgunj, Kathmandu
Email: dr_madhut@yahoo.com

Case report

A 14-year-old boy presented with fleshy growth in left orbit after removal of that eye about 8 months back. He gave the history of high velocity trauma with the cricket ball to the left eye in early childhood. According to the patient, his both eyes were normal before the trauma. Following the injury, he was diagnosed as a case of retinal detachment in left eye in peripheral eye hospital of eastern Nepal. He was advised to go to tertiary eye centre immediately for possible surgery. But the patient did not seek any further treatment. Subsequently, he gradually developed pain in that eye associated with constant headache which was localized to the parietal region, not associated with nausea, vomiting or fever. He used to use some topical medicine and oral analgesics for the pain but still did not take any medical help. He visited the eye hospital only after 2 years when the pain became intolerable. In the ophthalmic record from the peripheral eye hospital, his visual acuity in the right eye was 6/6 and in the left, there was no light perception. The anterior chamber was shallow with cataractous lens in the

left eye. The IOP was raised to 50mm of Hg. The ophthalmic findings in the right eye were normal. The CT scan of the orbit and head was also done, which showed left orbital mass, with calcification in posterior vitreous cavity. The brain scan was within normal limits (Figure 1). The diagnosis of left painful blind eye with secondary glaucoma was made and left eye enucleation surgery was performed 8 months ago in the same hospital. Histopathological examination of the enucleated eye showed malignant melanoma with resected optic nerve end free of tumour cells. He was referred to oncologist but the patient did not go for further management.

On the first visit of the patient to our institution, his visual acuity in right eye was 6/6 whereas the left side had anophthalmic socket S/P enucleation .There was a fleshy growth occupying the left socket with fornicial fullness (Figure 2). The conjunctiva overlying the mass was pink and transparent with moderate discharge. The regional lymph nodes were within normal limit. Systemic examination was done by Paediatrician which was normal. CT scan of orbit and brain was also done which showed a contrast enhanced heterogenous lobulated mass in the left socket with prominent calcification. The left bony orbit was widened with area of erosion in the medial wall. The findings were suggestive of recurrence of malignant melanoma of the left orbit (Figure 3).

Incisional biopsy of the lesion showed malignant melanoma of spindle B type. Bone marrow aspiration showed normocellular marrow with no evidence of malignant cells. With the available clinical evidence along with biopsy report, the diagnosis of recurrent malignant melanoma of left orbit, without systemic metastasis was made. In our centre, lid skin sparing modified exenteration of left orbit was performed and chemotherapy was started (Figure 4). Inj Dacarbazine 275 mg in 200 ml NS over 1-2hours OD for 5 days was given, as advised by Paediatric oncologist. The histopathology of orbital mass obtained from exenteration had shown malignant melanoma of spindle A type, infiltrating lacrimal glands and eye lid muscles. All margins of the exenterated mass was infiltrated with tumour cells. Hence, after completion of chemotherapy, the patient was referred to a cancer hospital for radiotherapy.

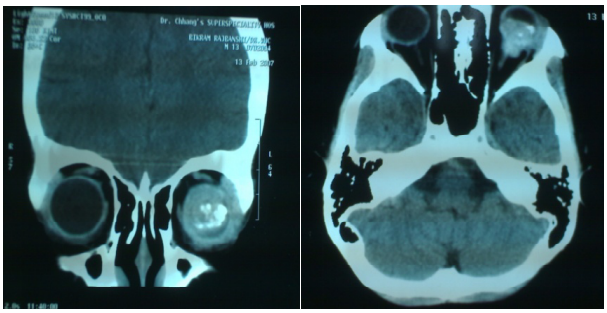

Figure 1: CT scan of orbit and brain showing intraocular calcification.(before enucleation)

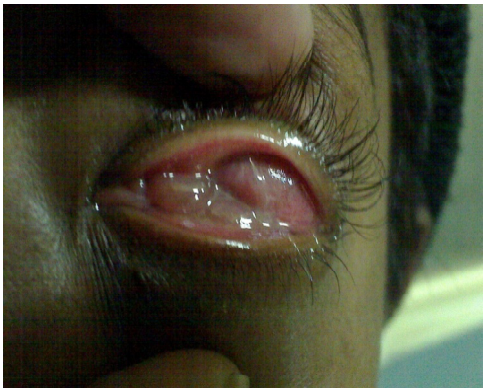

Figure 2: Fleshy lobulated tumour mass of the left socket with discharge (post enucleation)

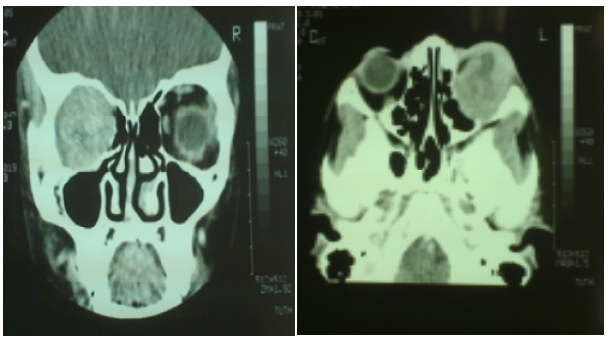

Figure 3: CT scan of orbit and head showing widening of orbit with orbital mass eroding of medial wall

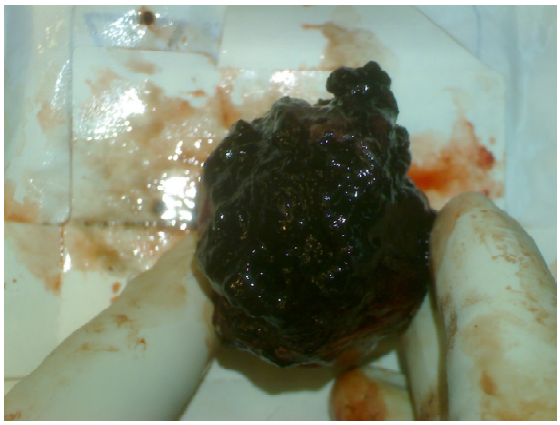

Figure 4: Specimen showing firm, darkly pigmented, lobulated mass (obtained during modified exenteration of left orbit.)

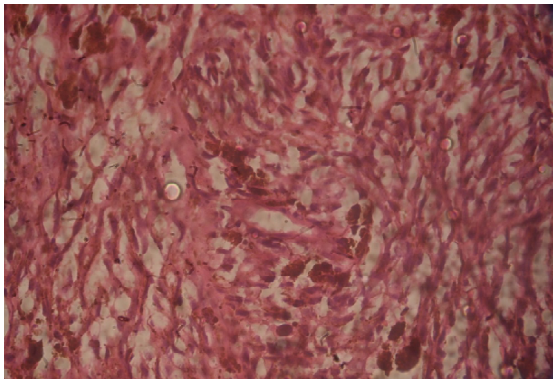

Figure 5: Photograph of histopathological section (H&E, 40X) showing spindle cells arranged in fasicles with indistinct nucleoli and melanin pigments in the cytoplasm

Discussion

Among all the primary intraocular tumours, uveal melanoma is the commonest one in adults.

The most common intraocular tumour in paediatric population is retinoblastoma (Lee et al, 2000).

Here, we describe a case of malignant melanoma which is a rare presentation in peadriatic age group. There is history of injury in the same eye before he developed ocular problem. Literatures have mentioned that many cases of uveal neoplasm are preceded by injury. Ocular trauma can be responsible for exciting obvious activity in a tumour which might have shown a latent course. Chronic intraocular inflammation may be a determining factor in some cases as seen in this case, where it took couple of years to develop clinical presentation (Nicoletti et al, 2006). In some of the cases of intraocular tumour, phthisis bulbi may be a frequent presentation where toxic effect of necrotic changes are observed.

Presentation with secondary glaucoma, as seen in this case, could be one of the presentations of intraocular tumour. When the tumour arises from ciliary body or if the tumour is large in size, it can directly compromise the angle structure increasing the resistance to aqueous out flow. Moreover, the malignant cells and macrophages with engulfed malignant cells can also clog the trabecular meshwork, giving rise to increased intraocular pressure (el Baba et al,1988).

Regarding the treatment modalities, for the medium sized tumour, enucleation and brachytherapy has shown to have the similar results as far as survival rate is concerned (COMS group, 2006). Charged Particle Radio therapy is also an alternative treatment modality in medium sized uveal melanomas (Egger et al 2002). Stereotactic photon beam irradiation therapy (Muller K et al, 2005) and thermotherapy (Sheild CL et al, 1998) has shown significant role in treatment of choroidal melanoma. Photodynamic therapy has also been tried where there was failure of brachytherapy and trans pupillary thermotherapy (Barbazetto et al, 2003). Recurrence of uveal malignant melanoma is a rare complication which can be treated with exenteration along with chemo and radio therapy (Shields et al, 1984)).

Conclusion

Malignant melanoma can be an unusual presentation in children. Recurrence of uveal malignant melanoma is a rare complication. In all cases of treated uveal melanoma, periodic ophthalmic examination is needed to diagnose the early recurrence and to plan for further management.

References

Barbazetto IA, Lee TC, Rollins IS, Chang S, Abramson DH (2003). Treatment of choroidal melanoma using photodynamic therapy. Am. J. Ophthalmol;135(6): 898–899.

Collaborative Ocular Melanoma Study (COMS) Group. The COMS randomized trial of iodine 125 brachytherapy for choroidal melanoma(2006). V. Twelve-year mortality rates and prognostic factors: COMS report no. 28. Arch. Ophthalmol. ;124(12): 1684–1693.

Egan KM, Ryan LM, Gragoudas ES (1998). Survival implications of enucleation after definitive radiotherapy for choroidal melanoma: an example of regression on time-dependent covariates. Arch Ophthalmol; 116(3):366-70.

Egan KM, Seddon JM, Glynn RJ, Gragoudas ES, Albert DM (1988). Epidemiologic aspects of uveal melanoma. Surv Ophthalmol; 32(4):239-51.

Egger E, Zografos L, Schalenbourg A et al (2003). Eye retention after proton beam radiotherapy for uveal melanoma. Int. J. Radiat. Oncol. Biol. Phys. ;55(4): 867–880.

el Baba F, Hagler WS, De la Cruz A, Green WR (1988). Choroidal melanoma with pigment dispersion in vitreous and melanomalytic glaucoma. Ophthalmology; 95(3):370-77.

Lee SB, Au Eong KG, Saw SM, Chan TK, Lee HP (2000). Eye cancer incidence in Singapore. Br J Ophthalmol; 84(7):767-70.

Muller K, Nowak PJ, de Pan C et al. (2005). Effectiveness of fractionated stereotactic radiotherapy for uveal melanoma. Int. J. Radiat. Oncol. Biol. Phys ;63(1):116–122.

Nicoletti AG, Costa DS, Ghanem RC, Carricondo PC, Santo RM, Matayoshi S (2006). Atypical choroidal melanoma: report of 3 cases. Clinics (Sao Paulo); 61(1):79-82. doi: /S1807-59322006000100014.

Shields JA, Augsburger JJ, Dougherty MJ (1984). Orbital recurrence of choroidal melanoma 20 years after enucleation. Am J Ophthalmol; 97(6):767-70.

Source of support: nil. Conflict of interest: none