Kathmandu University Medical Journal (2010), Vol. 8, No. 2, Issue 30, 247-250

Case Note

Retinoblastoma in a 37 years old man in Nepal: A case report

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Abstract

Retinoblastoma is extremely rare in adults. We report a case of Retinoblastoma diagnosed by histopathology in an enucleated specimen of a 37 year old patient who presented with pain and diminished visual acuity with intraocular mass and serous detachment. CT head and orbit showed uniform hyperdense mass in nasal quadrant of left globe projecting into adjacent vitreous cavity. Chest X Ray and USG abdomen was unremarkable. Initially provisional diagnosis of amelanotic melanoma was made. In view of nil visual prognoses enucleation was done. Sections from the enucleated eyeball showed diffuse proliferation of tumour cells. These tumour cells were small sized with scanty cytoplasm, and oval and hyper chromatic nuclei. A diagnosis of poorly differentiated Retinoblastoma was made with TNM (AJCC) stage as T1NXMX.

This paper highlights the delayed presentation of retinoblastoma in adult. Although retinoblastoma is rare in adult, we suggest keeping retinoblastoma in differential diagnosis of intraocular mass until proven histopathology report is available.

Key words: Retinoblastoma, Adult, Intraocular mass

A retinoblastoma occurring in an adult always evokes interest. These tumours are most common before the age of three and rarely occur after the age of ten. However, retinoblastoma has been reported even in the age of 74 years[1]. The common modes of presentation of retinoblastoma are leukocoria, squint, hypopyon, hyphema, proptosis, secondary glaucoma, and orbital cellulitis[2]. Retinoblatoma arises from a multipotent precursor cell that has the ability to develop into any type of inner or outer retinal cells. Histopathologic findings include the Flexner- Wintersteiner rosette, Homer Wright rosette, and the fleurette. Extensive areas of necrosis and calcification within the tumour are highly characteristic. The loss of the tumour suppressor retinoblastoma gene plays a critical role in the development of retinoblastoma[3].Patient with retinoblastoma is always at the risk of metastasis. Most metastases are detected within the first 2 years after diagnosis[4].

We report a retinoblastoma in 37 years man; the report is probably the oldest histopathologically proven case in Nepal.

Case report

A 37 years man presented in Tilganga Institute of Ophthalmology, Kathmandu, Nepal with history of pain and diminished vision in left eye for 9 months. Patient also gave history of on and off redness in the same eye for the same duration. Past ocular history and medical history was unremarkable. Family history was not contributory.

Visual acuity in right eye was 6/6. Intraocular pressure was 12 mmHg. Anterior segment and fundus finding were within normal limits.

Left eye had PL+PR accurate vision in all four quadrants. Intraocular pressure was 40 mmHg. Anterior segment showed circumciliary congestion, old KPs with superficial punctuate keratopathy, 360^0 of neovascularisation, ectropion uvea, dilated and fixed pupil with RAPD. Fundus examination revealed a whitish elevated mass (about 6 DD size) nasally in the ciliary body region with serous exudation, sheathing of vessels. The Optic disc was pale with an inferior notch and 0.9:1 cupping. Regional lymph nodes were not palpable. Systemic examination revealed no abnormality.

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B Scan USG showed dome shaped lesion with a diameter of 10 mm and height of 4 mm and surrounding retinal detachment in the left eye. CT head and orbit showed uniform hyperdense mass in nasal quadrant of left globe projecting into adjacent vitreous cavity. Chest X Ray and USG abdomen was unremarkable. Consultation with internist was made and no signs of metastasis were noted.

Intraocular tumour with possible amelanotic choroidal melanoma was provisionally diagnosed and in view of nil visual prognoses enucleation of left eye was done. Gross specimen of eyeball measured anteroposteriorly 3 cm, vertically 2.9 cm and transversely 2.9 cm. Optic nerve measured 0.3x0.2 cm. Cut section showed whitish mass measuring 1.2X0.8 cm.

Sections from the enucleated eyeball showed diffuse proliferation of tumour cells. These tumour cells were small sized with scanty cytoplasm, oval and hyper chromatic nuclei. These tumour cells were also oriented around the blood vessel. Area of tumour necrosis with calcification and few multinucleated giant cells were also seen. Uvea, sclera, cornea and optic nerve was free of tumour. Lymphovascular invasion was not seen. A diagnosis of poorly differentiated Retinoblastoma was made with TNM (AJCC) staging as T1NXMX.

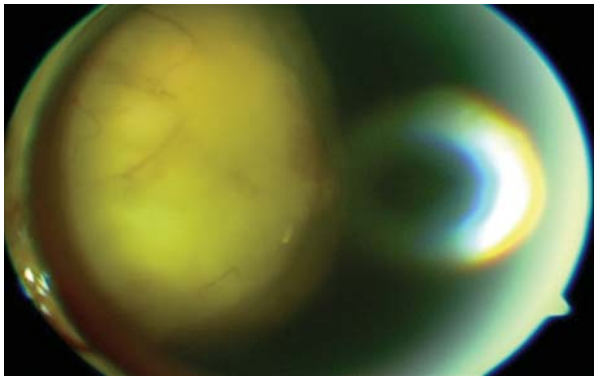

Fig 1: Fundus photograph showing the retinoblatoma

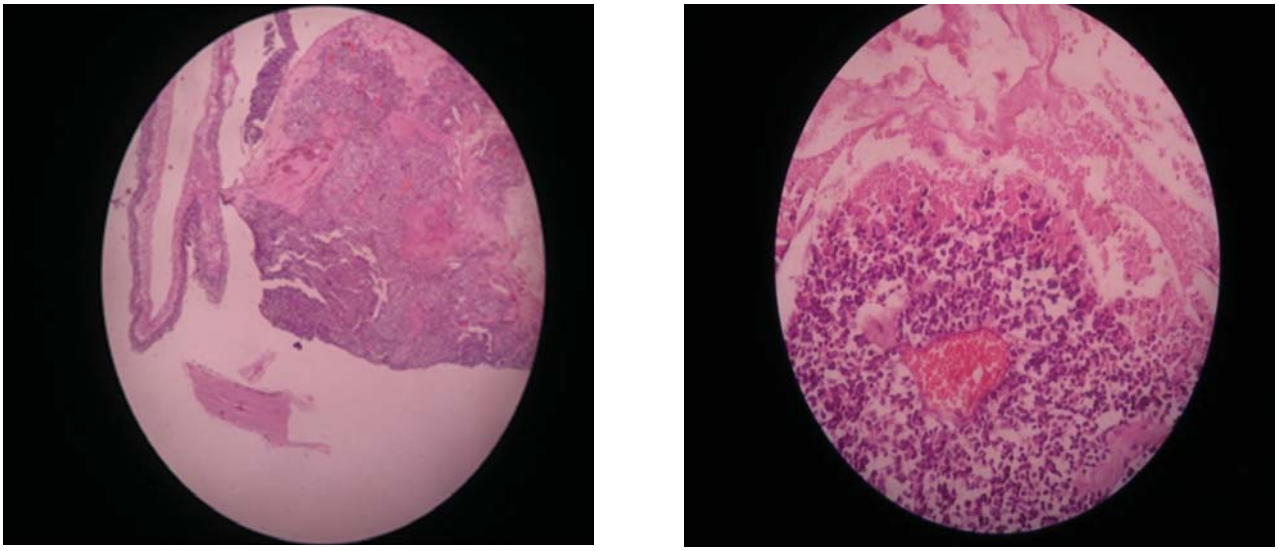

Fig 2: Histopathology slide showing tumor cells with area of coagulative necrosis in the right and small round cell tumor with high N: C ratio and hyper chromatic nuclei in the left

Discussion

The history of pain and decreasing vision in an otherwise healthy adult in 37 years of age and finding of intraocular mass was sufficient to consider as intraocular tumour possibly amelanotic melanoma of choroid. However, what is highlighted in this paper is Retinoblastoma was diagnosed in histopathological evaluation of enucleated specimen. It is very likely to misdiagnose the case because of its rarity in late adulthood.

Reinoblastoma has been reported even in a 74-year-old man[1]. In 1929 Verhoeff reported a retinoblastoma in a man 48 years old. Rasmussen in 1944 reported another case in a man of 48, and at that time he reviewed the literature for adult cases. In Wintersteiner's series of 429 cases of retinoblastoma the oldest patient was 16 years. Magbe reported a case in a 20-year-old woman; Gerard and Morel in a 35-year-old man, Torence A Makley in 53 years old man and Gerard and Detray in a 66-year-old woman[5]. Saiju R reported a retinoblastoma in 9 years of boy in Nepal[6].Badhu B previously reported a retinoblastoma in 10 years boy in Nepal[7]. If retinoblastoma could be diagnosed in its early stage, less aggressive treatment modalities like photocoagulation, cryotherapy, cobalt plaques, external beam radiation, and diathermy would be extremely useful[9]. Management of retinoblastoma should be guided by the objectives to save life, to retain anatomical integrity of the eye, to preserve vision, and to obtain good cosmetic results[10]. Enucleation is more invasive modality, which is applied if the disease is too far advanced to salvage useful vision in the affected eye, or when other treatments have failed[11].

The small round cell tumour from eye is unless otherwise proven is retinoblastoma despite any age. The differentiation status is determined based on the percentage of Flexner–Wintersteiner rosettes, Homer–Wright rosettes, and fleurettes seen in the histopathological slides. Tumours with rosettes (with or without fleurettes) in more than 80% of the areas were considered well differentiated and the rest were considered poorly differentiated. It was also observed that bilateral well-differentiated tumours to occur significantly earlier compared to bilateral poorly differentiated tumors. A similar trend was also noted between unilateral well-differentiated and unilateral poorly differentiated retinoblastoma Thus, poorly-differentiated tumors were noted to occur significantly later than well differentiated tumours, irrespective of the laterality of the tumour[12].

Differentiated and poorly differentiated tumors may represent events at different levels of retinal cell development. Alternatively, the earlier occurrence of well-differentiated tumors compared to poorly differentiated tumors might represent tumour progression and differentiation—a process where less severe well-differentiated tumors progress to more severe poorly differentiated tumors over time[13,14,15]. However, the theory of differentiation will be difficult to prove in retinoblastoma because there is often a delay in presentation and diagnosis of tumours, making it difficult to study tumours from their onset.

Conclusion

Although intraocular mass in adult is unlikely to be retinoblastoma, it can't be ruled out on clinical background. Histopathological report may demonstrate Retinoblastoma even in adult. So, careful and thoughtful evaluation is needed in any intraocular mass.

Reference

1. Finlay JR, Byron H. Retinoblastoma in the adult: review of literature and report of a case associated with benign melanoma. Acta XIX Concilium Ophthalmologicum (New Delhi). 1962;2:1168-78.
2. Mietz H, Hutton WL, Font RL. Unilateral retinoblastoma in an adult: report of a case and review of literature. Ophthalmology. 1997;104:43-7.
3. Albert DM, Jackobiec FA. Principles and Practice of Ophthalmology, volume 3. 3rd edition. UK: W.B. Saunders Company; 1994.
4. Yttebsorg J, Arnesem K. Late recurrence of retinoblastoma. Acta Ophthalmol. 1972;52:367
5. Makley TA. Retinoblatoma in 52 years man. Arch Ophthalmology. 1963;69(3):325-7.
6. Saiju R, Thakur J, Karmacharya PC, Sah DN. Nepal Med Coll J. 2006;8(3):171-5.
7. Badhu B, Sah SP, Kumar S. Retinoblastoma in Late Childhood. Journal of Nepal Medical Association. 2001:40:86-9.
8. Abramson DH. Treatment of Retinoblastoma. In:Blodi, FC, d. Contemporary Issues in Ophthalmology, Vol.2, retinoblastoma. New York: Churchill living stone; 1985.p. 63-93.
9. Abramson DH, Ellsworth RM. The Surgical Management of Retinoblastoma. Ophthalmic Surgery. 1980;11(9):596-8.
10. Goddara AG, Kingstoma JE, Hungerfordb JL. Delay in diagnosis of retinoblastoma: risk factors and treatment out come. B J Ophthalmol. 1999;83:1320-3.
11. Shields JA, Shields CL, De Potter P. Enucleation Technique for Children with Retinoblastoma. Journal of Pediatric Ophthalmology & Strabismus. 1992;29(4):213-5.

12. Madhavan J et al. The Relationship between tumor cell differentiation and age at diagnosis in Retinoblastoma J Pediatr Ophthalmol Strabismus. 2008;45:22-5.
13. Helm J, Enkemann SA, Coppola D, Barthel JS, Kelley ST, Yeatman TJ. Dedifferentiation precedes invasion in the progression from Barrett's metaplasia to esophageal adenocarcinoma. Clin Cancer Res. 2005;11:2478-85.
14. Santos L, Loo C, Chandraratnam E, Gune S. Anaplastic carcinoma dedifferentiation of solid variant of papillary thyroid carcinoma. Pathology. 2004;36:196-211.
15. Yano H, Iemura A, Fukuda K, Mizoguchi A, Haramaki M, Kojiro M. Establishment of two distinct human hepatocellular carcinoma cell lines from a single nodule showing clonal dedifferentiation of cancer cells. Hepatology. 1993;18:320-7.