Extraocular retinoblastoma: a neglected case

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Accepted 1 May 2017

DESCRIPTION

A 13-month-old female child was brought to our tertiary eye hospital with a history of a rapidly enlarging mass in the right eye for 4 months. Her medical history was suggestive of leukocoria in the right eye, noted 6 months ago, for which medical attention was sought elsewhere. At that time, a diagnosis of group E retinoblastoma in the right eye was made. The left eye was normal. The caretakers were explained about the advanced nature of the disease in the right eye and the need for enucleation surgery. The parents denied surgery despite counselling and opted for neoadjuvant chemotherapy and regular follow-up. However, they did not comply with the treatment and the child presented to our hospital 5 months later with a huge mass involving the right eye and a deteriorating systemic condition. Local examination showed a large extraocular mass measuring approximately 14×16 cm along its greatest dimensions, with superficial pigmentation and dryness (figure 1A,B). The child was very irritable and was advised contrast-enhanced MRI of the brain and orbits to note the extent of disease and intracranial extension, if any. However, within 2 days of presentation, the child died, most probably due to extensive systemic and CNS metastasis.

In low/middle-income countries, extraocular retinoblastoma still constitutes 20%–50% of retinoblastoma cases and is a major concern.^{1 2} Orbital invasion is associated with an increased risk of metastasis and death. In this case, treatment was declined because of social stigma and lack of awareness about the disease, resulting in severe neglect and death.

Contributors This patient was initially evaluated in our ocular oncology clinic by AP and BC and planned for immediate imaging followed by neoadjuvant chemotherapy but we lost the patient due to extensive systemic metastasis. AP, BC along with AKB wrote the case after critically analysing the educational value of the case.

Figure 1 (A,B) Large orbital mass arising from the right eye.

Learning points

- ▶ Extraocular retinoblastoma constitutes a significant proportion of cases in low/middle-income countries.
- ▶ There is a pressing need for increasing awareness about early diagnosis of retinoblastoma and its consequences, if left untreated.

Competing interests None declared.

Patient consent Consent obtained from guardian.

Provenance and peer review Not commissioned; externally peer reviewed.

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REFERENCES

- 1 Chawla B, Hasan F, Azad R, *et al.* Clinical presentation and survival of retinoblastoma in Indian children. *Br J Ophthalmol* 2016;100:172–8.
- 2 Shah PK, Narendran V, Kalpana N. Outcomes of Intra- and extraocular retinoblastomas from a single Institute in South India. *Ophthalmic Genet* 2015;36:248–50.

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To cite: Chawla B, Pujari A, Behera AK. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2016-218471

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