RETINOBLASTOMA IN LATE CHILDHOOD

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ABSTRACT

Retinoblastoma, the commonest intra-ocular tumor of childhood, is rare after the age of 5 years. We report a case of retinoblastoma in a 10 year-old boy, who presented with a fungating proptosed tender mass involving the entire left eye and the orbit. A clinical diagnosis of retinoblastoma was made. Exenteration of the left orbit was done. Histopathology revealed well-differentiated retinoblastoma with the involvement of optic nerve section. CSF and bone marrow aspirates did not show any malignant cells. This paper highlights the delayed presentation of retinoblastoma in late childhood and discusses the treatment modalities of orbital retinoblastoma.

Though retinoblastoma in late childhood is uncommon, it should be considered in the differential diagnosis of leukocoria and a fungating proptosed mass.

Key Words: *Retinoblastoma, late childhood, fungating proptosed mass.*

INTRODUCTION

Retinoblastoma, the commonest intra-ocular tumour in childhood, is diagnosed before 5 years of age in 90% of cases.^{1,2} It has, however, been reported in adults^{3,4} even in a 74-year-old man.⁵ Presentation of retinoblastoma with a fungating proptosed mass in late childhood or in adults is unusual.¹ We found a case of retinoblastoma

presenting as a proptosed fungating orbital mass in a 10-year-old boy at B.P.Koirala Institute of Health Sciences, Dharan, Nepal.

CASE REPORT

A 10- year-old boy presented to the Department of Ophthalmology, B.P.Koirala Institute of Health Sciences, Dharan, Nepal with the complaint of

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painful rapidly growing foul smelling mass involving his left eye for three months. It was preceded by the presence of a white pupillary reflex associated with diminution of vision in the affected eye for six months. Family history was unremarkable. Past medical history was not contributory. Examination revealed a fungating proptosed tender mass of firm consistency with irregular surface and necrotic areas involving the entire left eye and orbit (Fig. 1). The right eye was

Fig. 1, The clinical picture of fungating proptosed mass in the left orbit.

normal in all aspects. The preauricular and submandibular lymph nodes were not palpable. CT scan of the orbit showed a mass involving the entire eyeball with areas of calcification as well as the involvement of optic nerve and periorbital tissue. CT of the head disclosed normal gray and white matter and no signs of intracranial masses. CSF and bone marrow aspirates did not reveal any malignant cells. A clinical diagnosis of retinoblastoma was made. The patient underwent exenteration of the left orbit under general anesthesia. Grossly the exenterated mass with oedematous and erythematous eyelids measured 7x6x3 cm. The tumor was gray white in color, occupied the entire globe and extended beyond the sclera. Focal areas of necrosis and haemorrhages were also identified. Sections examined showed small round cells with hyperchromatic nuclei and scanty cytoplasm. At places, the cells were arranged

in rossets and fleurettes (Fig. 2, Fig. 3). Resected

Fig. 2, Section showing well differentiated retinoblastoma (H & E stain, X40).

end of the optic nerve showed tumour infiltration. The diagnosis of well differentiated retinoblastoma

Fig. 3, Higher magnification showing rossets and fleurettes (H & E stain, X100).

with involvement of the optic nerve section was offered. In the postoperative period, the orbit was allowed to granulate. The patient was referred for combined chemotherapy and radiotherapy.

DISCUSSION

The history of painless leukocoria initially for 6 months in the case presented here was significant to suspect retinoblastoma by health workers. Medical consultation, however, at that stage was not, sought for which shows poor awareness about the disease. However, what is highlighted in this

paper is the delayed presentation of retinoblastoma with fungating mass in late childhood.

Reported modes of presentation of retinoblastoma are leukocoria, squint, hypopyon, hyphema, proptosis, secondary glaucoma, and orbital cellulitis.⁶ Because of its rarity in late childhood, the fungating proptosed mass or leukocoria due to retinoblastoma may be misdiagnosed. Primary health care professionals⁷ as well as the parents require education about the importance of ocular symptoms in pediatric patients.

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.⁷ If retinoblastoma could be diagnosed in its early stage, less aggressive treatment modalities like photocoagulation, cryosurgery, cobalt plaques, external beam radiation, and diathermy would be extremely useful.⁸

Enucleation and exenteration are more invasive modalities, which are applied if the disease is too far advanced to salvage useful vision in the affected eye, or when other treatments have failed.⁹ Enucleation has to be carried out with special care not to spread tumor into the orbit during surgery and to obtain a long section of optic nerve by applying traction suture through the medial and lateral rectus insertions¹⁰ or a hemostat to the medial rectus stump.¹¹ Lateral canthotomy has been advised in a tight orbit.¹¹ No clamps or snares on the optic nerve stump should be used as they can leave crush artifact.¹¹

The removal of all the orbital structures including lids, eyes, extraocular muscles, nerves and orbital fat is termed as exenteration. It is reserved for treatment of primary orbital mass or orbital

recurrence of retinoblastoma.¹⁰ Orbital retinoblastoma can be treated with exenteration, local irradiation and systemic chemotherapy in combination. Studies have shown that among the patients with orbital retinoblastoma, 5% survive when treated with exenteration alone, while 38% survive with adjuvant chemotherapy.¹² The "cure" rate of retinoblastoma with exenteration and local irradiation is 30.9% vs 9% with exenteration alone.¹³ A long-term survivor with orbital recurrence of retinoblastoma treated with radiation and systemic chemotherapy has been reported.¹⁴ Exenteration alone may not be effective in removing the entire recurrent tumor, but in combination with radiation may affect a more complete local cure.¹⁵ The role of chemotherapy is to treat any microscopic systemic metastasis.¹⁵

CONCLUSION

1. Health education on the modes of presentation of retinoblastoma particularly leukocoria should be promoted among the population for the early detection and management of retinoblastoma.
2. Retinoblastoma is uncommon in late childhood, but its diagnosis should be considered in cases of fungating proptosed orbital mass preceded by leukocoria.

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