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Case Report: Open Access

A Rare Case Report On Recurrence of Orbital Retinoblastoma

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ABSTRACT

Retinoblastoma is the most common intraocular malignancy in children. It has been reported through studies and researches that survival rate of extra ocular retinoblastoma ranges between 50% and 70% which is still low. Therefore better research should be carried out so that we can improve the survival rate and visual outcome of the patient. The survival of retinoblastoma patients still remains low, primarily due to a delayed presentation, resulting in larger proportions of extra ocular disease compared with the developed world, where majority of the disease is intraocular. This is a case of 6 year boy presenting with pain, swelling along with inflammation of the right orbit for 15 days which was enucleated six month ago. Recommendations are made on the role of chemotherapy and radiotherapy to reduce the morbidity of patient. After the case presentation, a review of therapeutic modalities for treatment of retinoblastoma will be discussed, with particular focus on enucleation and the management of recurrent retinoblastoma.

Keywords: Retinoblastoma, Recurrence, Treatment

INTRODUCTION

Retinoblastoma is an intraocular tumor of neuroectodermal origin. It most commonly occurs unilaterally, more than 70% of the cases. It is a life threatening and sight threatening disease. Enucleation is the main modality of treatment. Among all the enucleated eyes 4.2% cases shows recurrence [1]. Most important risk factor for recurrence is compromised surgical margin and extra scleral invasion [2,3]. Retro laminar Optic nerve invasion and choroid invasion is the most important risk factor for recurrence. Adjuvant therapy required in those situations [4].

CASE REPORT

A boy of 6 years age coming to the outpatient department of Ophthalmology, Agartala Govt. Medical college, Agartala, with the complain of pain and swelling of the right orbit for 15 days. It was associated with itching and discharge from the right eye. It was associated with fever also. The patient had undergone enucleation of the right eye 6 month back elsewhere.

The parents of the baby noticed a white pupillary reflex six month ago after which they consulted with an ophthalmologist. After all the necessary investigations, he was diagnosed as a case of intraocular retinoblastoma. He was advised enucleation and adjuvant chemotherapy. The baby underwent enucleation of the right eye. Specimens were sent to the pathology department and it was diagnosed as a case of retinoblastoma. Prothesis was implanted during the time of enucleation surgery. Parents were advised for follow up and chemotherapy after 1 month. But the parents were disinterested in any of the follow up procedures and so unfortunately the patient did not receive any chemotherapy after enucleation. After three months the patient developed severe pain and swelling of the enucleated eye, associated with watering and discharge. On examination, a large proptotic mass was noted which was spreading the eyelids apart. It was Purple red in color, irregular surface with ulcero proliferative nature, hard in consistency, fixed to underlying structures, movements restricted. Lids were swollen and vessels over the skin were engorged. On contrast enhanced CT scan on the orbit shows soft tissue mass lesion approximately 5.6 cm × 4.7 cm × 4 cm seen completely filling the intraconal and extraconal compartments of the left orbit. CT scan of the brain showed no signs of intracranial extension. Lumber puncture, body scan and bone marrow biopsy were performed to find out any metastasis. But there were no signs of metastasis at that moment. Initially the patient was managed conservatively. After the diagnosis was confirmed we planned for combined radiotherapy and chemotherapy. But the patient only received 10 Gy radiotherapy, after that the parents refused

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further treatment and the patient was discharged on request. No subsequent documentation was available on the patient's course, but it was reported that the patient passed away due to metastatic disease **(Figures 1-4)**.

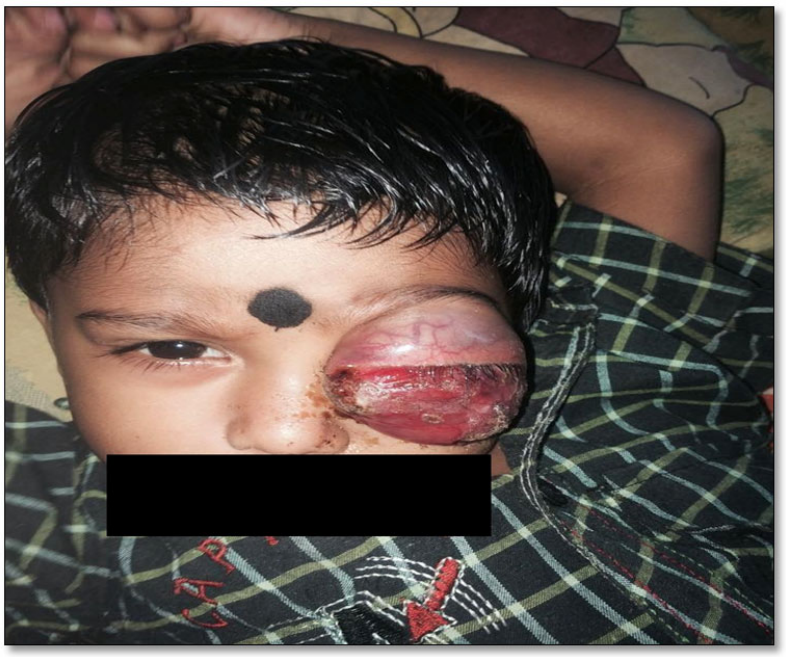

Figure 1. Clinical photo of the patient.

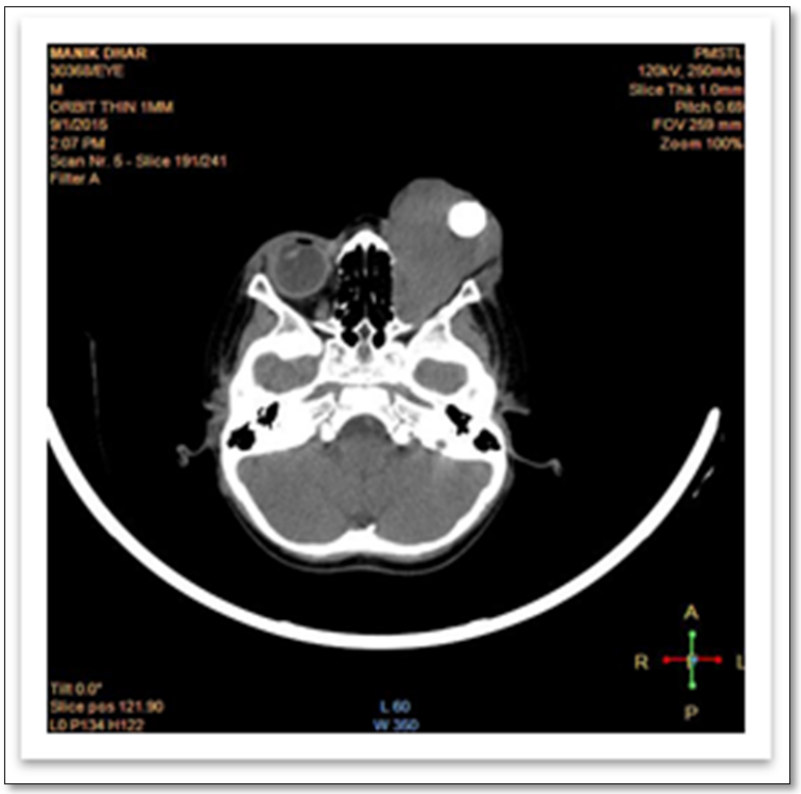

Figure 2. CT scan of the orbit.

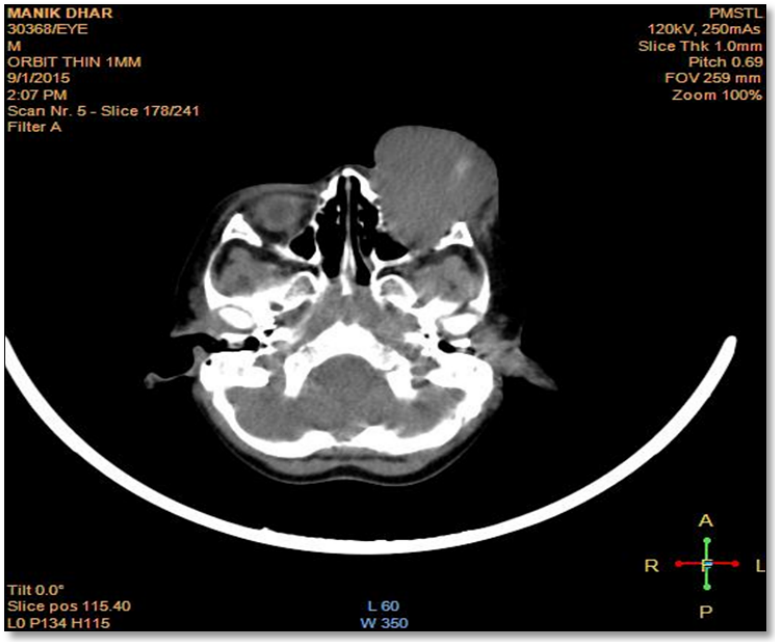

Figure 3. CT scan of the orbit showing the growth.

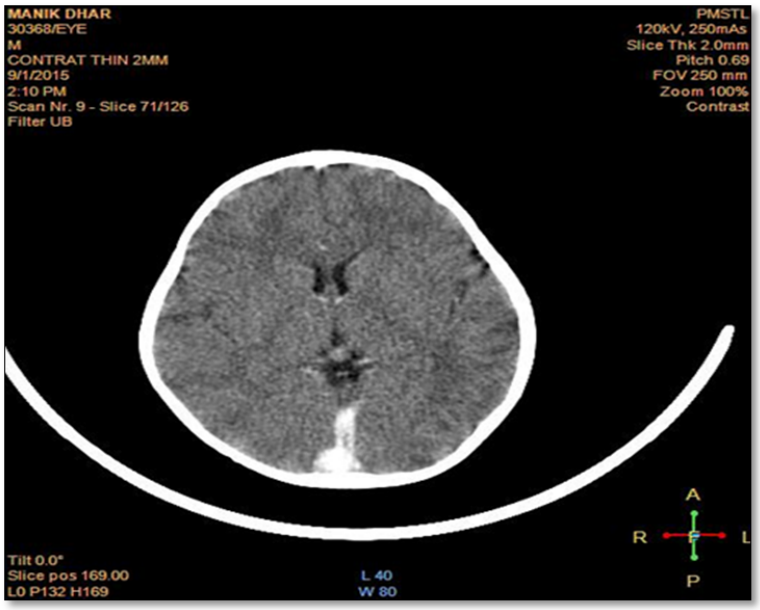

Figure 4. CT brain showing no signs of any metastasis.

DISCUSSION

Retinoblastoma is the most common intraocular malignant neoplasm of childhood affecting 1:17,000 to 1:20,000 children [5]. Most cases are unilateral; though up to 25% cases can be bilateral. It presents with white pupillary reflex, strabismus, nystagmus, proptosis, palpable orbital mass and lid swelling with ecchymosis. Sometimes extrusion of prosthesis after enucleation can represent recurrence.

Goal of treatment is to maintain life, to retain the eye, to preserve vision and to facilitate good cosmetic result. Management is based on the size and involvement of the tumor. Small sized tumors can be managed by trans pupillary thermotherapy, laser photocoagulation and cryotherapy for tumors anterior to ora serrate [6].

Medium sized tumors can be managed by plaque brachytherapy and EBRT along with chemotherapy. Invasive treatment modalities like enucleation and exenteration are reserved for large tumors with or without

optic nerve involvement. It should be followed by chemotherapy along with radiotherapy. Chemotherapy is of paramount importance in its management [7].

After enucleation, orbital bed and optic nerve examination is important. Inadequate amount of resection of optic nerve and/or spillage of tumor cells during surgery can cause recurrence. Goble et al. demonstrated the effectiveness of combination therapy enucleation, radiation and systemic chemotherapy in large tumors [8]. They conclude that aggressive therapy for orbital retinoblastoma should include local surgical biopsy, radical orbital radiotherapy and systemic chemotherapy.

If after careful enucleation, the surgical margin of the optic nerve is positive or if an orbital recurrence is present, removal of any tumor mass surgically with local irradiation and systemic chemotherapy is the management of choice. In review of our case presentation, it is hard to comment on the enucleation that took place. It is not known whether the optic nerve was extracted with as much length as possible.

Nevertheless, the patient certainly should have completed his course of initial radiation. When the patient presented with a large proptotic mass, he should have undergone the combination therapy of exenteration, radiation and chemotherapy. Unfortunately, the patient succumbed to a disease which still has a high mortality rate even with aggressive treatment.

CONCLUSION

If a child presents with ocular retinoblastoma, the utmost step is to decide whether the affected eye can be saved with the non-invasive procedures which are listed above. If however, the eye cannot be saved, then careful enucleation with extraction of a long optic nerve sample is advisable.

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