ORIGINAL RESEARCH PAPER

Radio-Diagnosis

RETINOBLASTOMA WITH PERINEURAL SPREAD ALONG OPTIC NERVE: A CASE REPORT

KEY WORDS:
Retinoblastoma, Perineural Spread, Optic Nerve, Intracranial Extension, Bony Erosion.

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ABSTRACT

Retinoblastoma (RB) is a most common primary intraocular malignancy affecting children less than 5 years. Retinoblastoma can spread by direct growth or through the blood. Retinoblastoma can sometimes spread through the optic nerve to the brain and the spinal cord. Perineural spread of tumor represent the ability of tumor to disseminate along nerve. Most commonly affected nerve are trigeminal and facial due to their extensive innervated territory, but any cranial nerve can be involved. The presence of perineural spread implies worsening of prognosis. Here we present case of retinoblastoma in 6 year old female patient.

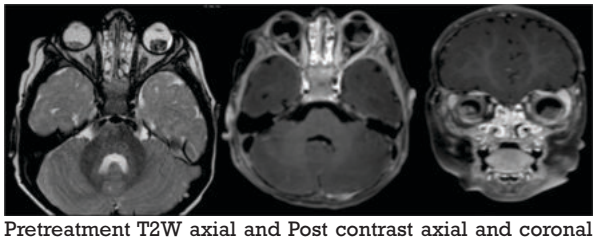
INTRODUCTION

Retinoblastoma (Rb) is a rare form of cancer that rapidly develops from the immature cells of a retina, the light-detecting tissue of the eye. The most common presenting feature is a white pupillary reflex called leukocoria, often recognized first by parents. Strabismus and decreased vision are also common. Patients with advanced disease can present with iris color changes, an enlarged cornea and globe, orbital inflammation, and exophthalmos. On presentation, approximately 60% of cases are unilateral, and the remaining 40% are bilateral. Almost half of children with retinoblastoma have a hereditary genetic defect associated with retinoblastoma. In other cases, it is caused by a congenital mutation in the chromosome 13 gene 13q14 (retinoblastoma protein). Retinoblastoma affects 1 in 16,000 births, with 8000–10,000 children diagnosed annually. There are no known geographic, racial, or sex predilections. When left untreated, retinoblastoma is almost always fatal.

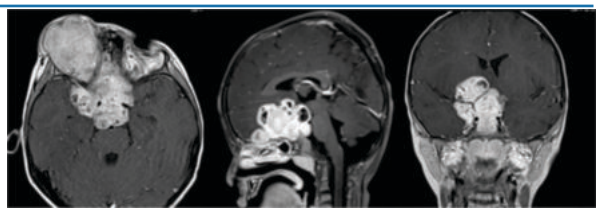
Case Report

A 6 year female patient is presented to our department with complaint of deterioration of vision and leukocoria. The patient initially presented 6 months back in a private hospital. The patient did not take any treatment at that time. After ophthalmologist clinically evaluated this patient, patient was taken for MRI for confirmation of disease and staging of same. At that time there was b/l retinoblastoma which does not show any extraocular spread. Patient has received treatment in form of enucleation of left eye and chemotherapy and kept on annual follow up but patient has developed meningeal and neural spread of disease.

Imaging Findings



Pretreatment T2W axial and Post contrast axial and coronal images of patient shows b/l retinoblastoma and Soft tissue mass is seen in posterior chamber of both orbit. It shows hyperintense signals in T1W-weighted images, hypointense signals in T2W-weighted and bright in STIR images. There is associated retinal detachment with minimal subretinal fluid collection on both eyes and no e/o extension into optic nerve on both side.



Post treatment one year later, Post contrast axial, sagittal and coronal images of Right orbit shows isointense to hypointense signals in T1W-weighted images, hyperintense signals in T2W-weighted and FLAIR images. Lesion shows heterogeneous post-contrast enhancement. Lesion shows internal calcification. Lesion shows internal non enhancing areas s/o necrosis. Lesion causes expansion of right orbit with exophytic growth with resultant proptosis.

Lesion extends along right optic nerve and optic chiasma into suprasellar and b/l parasellar regions. Lesion encases b/l ICA, origin of b/l MCA, lesion involves b/l cavernous sinus. Lesion causes erosion of body of sphenoid bone. Lesion extends into perimesencephalic and prepontine cistern. Lesion causes erosion of body of sphenoid bone. Lesion extends into perimesencephalic and prepontine cistern, Lesion abuts midbrain and pons, Lesion abuts rostrum of corpus callosum,.

Thickening and enhancement of intracanalicular segments of b/l VII/VIII nerve complex, leptomeningeal aspect of posterior aspect of medulla, and along b/l cerebellar foliae possibility of meningeal metastatic deposits.

DISCUSSION

When retinoblastoma is treated in the early stages by enucleation, the cure rates approach 95%. More advanced disease may require radiotherapy or chemotherapy in addition to enucleation. Prognosis of retinoblastoma is affected by many risk factors, the most important of which is the extent of invasion of the retinoblastoma into ocular coats and the optic nerve. There is currently no successful therapy for the treatment of patients who develop metastatic disease. Retinoblastoma can spread through direct invasion along the optic nerve to the brain, or along orbital tissue in adjacent bone, the nasopharynx via the sinuses, or the cranium via the foramina.

In second pattern of metastasis tumor cells that have invaded the optic nerve and leptomeninges and then disperse into the circulating subarachnoid fluid are characteristic. This may occur even when there is no tumor detected at the cut end of

the optic nerve. Via the circulating subarachnoid fluid, tumor cells can also reach the spinal cord, distant sites of brain, and the contralateral optic nerve. Tumor formation in these sites represents true metastasis rather than local invasion.

The third pattern of metastasis is hematogenous dissemination that results in widespread metastasis to the lungs, bones, brain, and other viscera.

Assessment of the optic nerve is essential for accurate staging. Prelaminar and intralaminar invasion of the optic nerve are not considered high-risk features. Postlaminar optic nerve invasion occurs in approximately 8% of patients and typically requires enucleation followed by systemic chemotherapy because of increased risk for metastatic disease and mortality. A definitive diagnosis of postlaminar optic nerve invasion is based on histopathology, which may not be available if eye-preserving treatment is considered. Thickening and enhancement of the postlaminar optic nerve on MR imaging usually indicates tumor extension.

Enhancement of the optic nerve at the globe–nerve junction can represent postlaminar tumor, pre- or intralaminar tumor with a posteriorly displaced lamina cribrosa, central retinal vessels, or inflammation. The most reliable direct MR imaging criteria to rule out advanced optic nerve invasion are normal optic nerve size, normal optic nerve signal on T2-weighted images, and optic nerve enhancement of ≤3 mm on postcontrast imaging

CONCLUSION

Perineural spread refers to tumor selectively travelling along a nerve away from a primary lesion and separate from main bulk of tumor. MRI is gold standard modality to evaluate nerve anatomy and to detect perineural spread. It is an important parameter to be evaluated when staging patients with retinoblastoma. The presence of perineural spread implies worsening of prognosis.

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