Original Research Paper Radiology

CT Scan Evaluation of Retinoblastoma

Saket Nigam	MD, Asst Professor, Department of Radiodiagnosis Rama Medical College and Hospital, Kanpur
Namrata Nigam	MD, Asst Professor, Department of Radiodiagnosis Era Medical College, Lucknow
Ankita Raj	MDS, Reader, Department of Maxillofacial Surgery Rama Dental College and Hospital, Kanpur
Amrita Raj	MDS, Reader, Department of Oral Pathology Rama Dental College and Hospital, Kanpur
Amit Pandey	MDS, Reader, Department of Periodontics, Rama Dental College and Hospital, Kanpur

ABSTRACT

A prospective study was carried out in patients suspected of retinoblastoma to evaluate the role of CT in diagnosis and staging. The presenting features were leukocoria and proptosis for which CT was advised after clinical evaluation. Calcification is seen in all patients with retinoblastoma and high sensitivity of the CT for detecting calcium makes it cost effective and reliable diagnostic modality. CT scanning also increase accuracy in differentiating retinoblastomas from other simulating lesions. In addition simultaneous scanning of brain can be used to evaluate intracranial extension.

KEYWORDS proptosis, calcification, leukocoria

INTRODUCTION:

Retinoblastoma is the most common intraocular childhood malignancy. Although few reports exist of retinoblastoma in adults, onset beyond 6 years of age is very rare. The most common initial sign of retinoblastoma is leukocoria, where the light emanating through the pupil is white reflecting off the tumor instead of red light reflecting off the retina. Later as the disease progresses the patient develops proptosis. Diagnosis of retinoblastoma is typically multidisciplinary. In patients with a newly diagnosed ocular lesion, a clinical examination is the first step in characterizing, the lesion. Imaging in the form of ocular ultrasound(US), computed tomography (CT), or magnetic resonance imaging (MRI) is used to confirm the diagnosis and its extension in patients with retinoblastoma. Cross sectional imaging is also beneficial in certain prognostic factors such as tumor invasion of the optic nerve and choroid. CT depict intralesional calcifications, the presence of which may confirm the diagnosis of retinoblastoma and exclude other differential diagnoses.

MATRIAL AND METHODS:

Study was conducted on 14 patients of retinoblastoma who presented to our department in last two years. The age group of the patient ranged from 3 to 8yrs. Scanning was done on multisclice CT scanner. Before performing the scan the procedure and objective of performing the scan was explained to the attendants/ parents. As the cases were later put up for contrast studies, so associated drug history (sensitivity to any drug) was also taken. Consent of the parent/ attendant were taken for contrast examination. Child who was restless was given oral sedation. CT Protocol: CT scanning of the orbit was performed with patient supine, head placed in a slightly hyperextended position . The entire orbit was encompassed, along with the adjacent portions of the brain, the cavernous sinus and portions of the paranasal sinuses.

RESULTS:

TABLE 1. *Distribution of Retinoblastoma According to the Site:*

Distribution	No. of Cases
Grade 1 (Tumor confined to the globe)	4
Grade 2 (Tumor extending retroorbitally and involving optic nerve)	8
Grade 3 (Tumor extending beyond the confines of the orbit or intracranial)	2

Table 2: Intravenous contrast enhancement in retinoblastoma:

Enhancement	No. of Cases
Mild	9
Moderate	2
Severe	2
No enhancement	1

Twelve (85.7 %)of our patients were below six years of age. Ten (71.4%) of patients showed extension of the retinoblastoma beyond the confines of the orbit of which two patients showed intra cranial extension of the disease. Most of the cases showed some amount of post contrast enhancement, no enhancement following I.V. contrast was seen in a single case. All retinoblastomas in the study showed calcification.

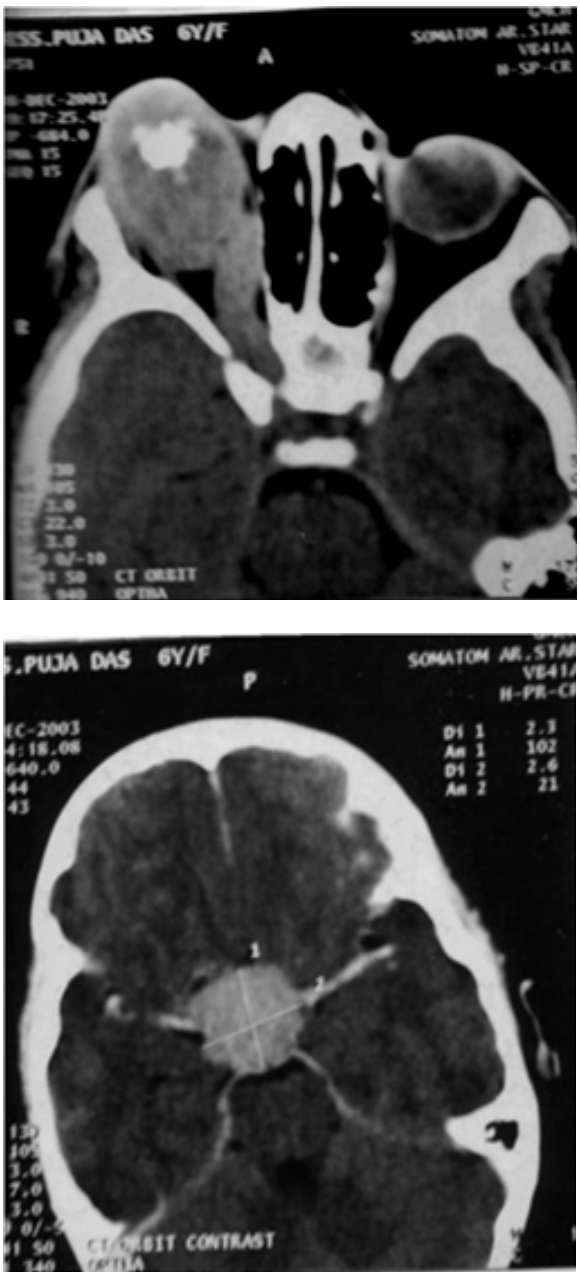

FIG1 (A,B): Plain and contrast CT of brain showing soft tissue mass with calcification, retrobulbar extension, thickened optic nerve and enhancing suprasellar mass- Retinoblastoma with intracranial extension.

Discussion:

Retinoblastoma on CT evaluation appears as a homogenous mass slightly hyperdense to vitreous, located peripherally indicating its retinal origin. Thickening of the optic nerve is a reflection of the tumor spread along the nerve, which may advance into the intracranial cavity via the subarachnoid pathway. Release of DNA from the necrotic tumor results in calcification which is present in almost all tumors. (1) Histology reports show that retinoblastomas calcify in ≤95% of cases.(2) An intrabulbar calcified mass is nearly pathognomonic of retinoblastoma. (3,4,5) However, in children older than 3 years of age, other intraocular lesions, such as retinopathy of prematurity, toxocariasis, Coats' disease, retinal astrocytoma, and optic nerve drusen may appear as a calcified masses.6,7 CT has a designated sensitivity in detecting calcifications in retinoblastoma of 81%–96%,(7-11). Calcification in our study was also seen in all cases which correlates with these studies. The calcification was confined to the intraocular component of the tumor in all cases which correlates well with study by Danzinges ***A et al*** (12) who also observed that calcification was more common in the intraocular than the extraocular portion of the tumor.

Most of the patients in our study were less than six years of age which correlates well with other studies like ***Thakur (5)*** Kivela (13),***Provenzale JM et al*** (14) stated that two third cases trilateral retinoblastoma have a positive family history. However no such correlation was seen in our study.

Retinoblastomas appeared as high density soft tissue mass on non contrast scan with most of them (64.2%) showing mild enhancement. Marked enhancement was demonstrated in a two cases of with intracranial extension. Our study tally with that of the Alan Danziger (1979) who studied 38 cases of retinoblastoma, 5 of which with intracranial extension showed marked contrast enhancement, while others showed nil to mild enhancement.

Management of retinoblastoma broadly depends on the presence or absence of extraocular involvement, its aggressiveness, whether involvement is unilateral or bilateral, and other factors that contribute to the potential for vision in the affected eye (15). CT examination of patients with suspected retinoblastomas is useful in determining retrobulbar spread, intracranial metastases, and second tumours.(16,17). The involvement of the optic nerve indicates poor prognosis, therefore, special attention is directed to investigation of the optic disc area with imaging procedures.(18) Most of the cases in our study showed retroorbital extension with involvement of the optic nerve. ***Meli FJ et al*** (19) reported eight patients with meningeal dissemination of retinoblastoma using computed tomography.

Some degree of proptosis was seen in most of the patients which was not associated with extensive periocular or orbital inflammation. Presence of inflammation may indicate sterile orbital cellulitis, which may be secondary to intraocular tumor necrosis (20).

Patients with the heritable form of retinoblastoma have a higher tendency for bilateral disease and a second primary malignancy with the most common second primary malignancy being a midline intracranial tumor originating in the primitive neuroectodermal tissue. These tumors are most commonly localized to the suprasellar or pineal region and usually manifest after the primary tumors appear in the globe. An intracranial tumor in a patient with retinoblastoma is referred to as '***trilateral retinoblastoma***' and is present in approximately 5%–7% of patients with bilateral disease (21, 22).

There have long been comparison of the two imaging modalities CT and MRI of which some autors prefer CT and others MRI. The advantage of CT is its detection of calcium and MRI's relative insensitivity to calcification. (23, 24). However, MRI provides better soft tissue characterization and involvement of adjacent structures. A choice between the two modalities becomes difficult.

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