



Multidisciplinary Approach for Retinoblastoma Management

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We read with keen interest the article by Shah et al “Pathologic Risk Factor in Retinoblastoma: An Institutional Experience Based on Analysis of Enucleated Eyes”(Shah A et al, 2012). They have highlighted high-risk histopathological features in Retinoblastoma with retrolaminar optic nerve invasion being the most common and also showed statistically significant correlation of high risk histological features with tumor size and AJCC stage of tumor. Their aim to assess the frequency and spectrum of high risk histopathological features in enucleated specimens of retinoblastoma was successful which may guide the clinician in timely planning for subsequent neoadjuvant therapy and prevent further ocular morbidity and mortality in children. We would like to congratulate authors for this article and with

invoked interest it led us to go in more depth and we wish to discuss several aspects of the study that can potentially influence the results.

The high risk histological features include one or more of the features including retrolaminar optic nerve invasion, massive choroidal invasion, combination of pre- laminar or laminar optic nerve invasion and focal choroidal invasion or tumor invasion into the anterior chamber, sclera, and extrascleral soft tissue (Kashyap S et al, 2012). Massive invasion of the choroid is a significant risk factor for extraocular relapse, justifying the use of adjuvant chemotherapy in an attempt to prevent it. Bosaleh et al in their article have classified massive choroid invasion according to 3 definitions: (a) extending at least 3 mm in any dimension, (b) through the choroid’s whole thickness, and (c) more than 50% of the thickness and/or more than 1 cluster (Bosaleh et al, 2012).

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Metastasis after enucleation have been mentioned in an article by Kopelman et al. Invasion of the tumor into the optic nerve or orbit were the risk factors most highly predictive of death from retinoblastoma. Choroidal invasion was not significantly associated with a fatal outcome (Kopelman et al, 1987). So, in further studies we could follow up the patients to find out the outcome for explaining prognosis in future. Metastatic and mortality rates were directly related to the extent of optic nerve invasion, and survival for patients with superficial optic nerve invasion up to the lamina was 83–86% which was almost the same as in patients with no optic nerve invasion. While it was 33%–55.6% for patients with post lamellar invasion, and 19%–36% for patients with optic nerve cut edge invasion (Hiasat et al, 2019).

Secondary glaucoma may also be predictive of massive choroidal invasion (Berry JL et al, 2017). Clinical factors predictive of optic nerve invasion included secondary glaucoma, exophytic growth pattern, and tumor thickness >15 mm. The main clinical risk feature predictive of choroidal invasion was iris neovascularization. Other clinical features predictive of high-risk retinoblastoma include older age at presentation (>2 years), longer duration lag period from symptoms to diagnosis (>3 months), hyphema, pseudohypopyon, staphyloma, and orbital cellulitis (Kaliki S et al, 2013). In collaboration with ophthalmologists involved in patient's treatment we can find risk for metastasis in future needing enucleation and this can be the basis for further studies in retinoblastoma.

In Lumbini Eye Institute & Research Center, recently, we had a case of unilateral retinoblastoma who underwent enucleation, in which MRI showed no extra globular extension and optic nerve and extraocular muscle were normal. But in histopathology there was an invasion of lamina cribrosa and optic nerve head. Patient was referred to a higher centre for systemic neoadjuvant chemotherapy but due to poor socio-economic status the patient went late and had orbital metastasis. The prediction and diagnosis of high-risk retinoblastoma are crucial because untreated high-risk retinoblastoma carries at least 24% risk for metastatic disease and adjuvant systemic chemotherapy reduces the risk to 0% to 4% (Kaliki S et al, 2013). Poor socioeconomic factors also play a role in prognosis. The histopathologic risk level is significantly associated with the delay in medical intervention, indicating the potential influence of parental and health-provider delays on disease severity (Xiao et al, 2019).

Almost all the patients with retinoblastoma undergo CT or MRI. In one of the articles by Hiasat et al, they concluded that MRI did not reliably predict prelaminar, laminar, or post laminar invasion of the optic nerve. The accuracy of MRI in detection of optic nerve invasion is variable according to the severity and extent of nerve invasion (Hiasat et al, 2019). Correlating MRI and histopathology can be a separate basis of discussion for knowing sensitivity and specificity of MRI scans and great addition to this type of study.

Best way to approach Retinoblastoma is

multidisciplinary. The examining, surgically managing and referring ophthalmologist provides detailed information of the child and the histopathological examination is by pathologist. According to the HPE report, follow-up examination by an ophthalmologist and referral for neoadjuvant therapy to an ocular oncologist in time is crucial. Moreover, social awareness and education on retinoblastoma will enable early presentation of children to the hospital with suspected disease for the timely management. As socio economic factors are

responsible for the delay in presentation and lag in treatment, treatment charge should be free of cost as far as possible and treatment facilities must be present in every district so that patients need not be referred to the central region for further treatment, which would help in preventing childhood blindness and mortality.



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