

ABSTRACT

Introduction: Retinoblastoma (RB) is one of the most common childhood intraocular malignant tumors with bilateral cases being more likely inheritable forms. This study aims to assess the clinical characteristics and treatment outcomes in affected children, who were treated at Lady Ridgeway Hospital in Sri Lanka, as well as to assess the proportion who developed trilateral RB and secondary neoplasms.

Methodology: A longitudinal study was conducted at the Pediatric Eye Unit at Lady Ridgeway Hospital for Children in Sri Lanka from January 2007 to January 2022. Data was collected from 258 children with RB who were treated and followed up at the hospital during this period after obtaining ethical approval.

Results: This study included 258 patients with eye tumors, among which 52% were males and 48% were females. Leukocoria was the most common complaint (n=180, 70.03%) and in 156(60.7%) cases eyes were affected unilaterally and in 101(39.29%) respondents' eyes were affected bilaterally. The mean age of unilateral cases was 8.48 ± 4.144 months and the mean age of bilateral cases was 7.32 ± 3.975 months. Only 10 children had a history of consanguinity. Also, a majority of 224(88.53%) patients did not have a similar family history. Moreover, the majority of 69 (36.3%) cases had a moderately differentiated RB, followed by well-differentiated RB (N=45; 23.7%) and poorly differentiated RB (N=37; 19.5%). Furthermore, the minority of 2% of cases had their optic nerve involved and the rest of 98% had no optic nerve involved.

The majority of cases at diagnosis were in Group E (45.25%), followed by group D (20.39%), group A (13.68%), group B (11.73%) and group C (8.10%). Bilateral cases had a higher proportion of tumors in stage A at diagnosis (n=45, 46.4%) compared to unilateral cases (n=2, 66.7%). Unilateral cases had a higher proportion of tumors in stage B (n=9, 90%) and C at diagnosis (n=12, 92.3%) compared to bilateral cases.

The majority of the Grade A tumors were treated with only laser therapy (n=49, 89.79%), and the majority of Grade B tumors (n=38, 90.47%), Grade C tumors (n=27, 91.90%), and Grade D

tumors (n=72, 98.63%) were treated with laser therapy together with systemic chemotherapy and Grade E was mainly treated with enucleation (n=154, 95.06%). In the majority of patients, enucleation was done (n- 210 eyes, 58.65%) and it was mainly done on Group E tumors 154(95.06%). Regarding the association between treatment modality and the current situation of the tumor, the majority of children who underwent laser therapy, cryotherapy, systemic chemotherapy, and intravitreal chemotherapy had regressed. Among the treated eyes, the majority of 111(31.00%) eyes are fully regressed and there were only 9(2.51%) eyes, which are still regressing. Among enucleated eyes, a majority of 192(53.63%) eyes had healthy sockets following the intervention. Moreover, there are 18(5.02%) eyes with new active tumors according to the current status of each tumor. Also, most patients (N=251, 97.2%) were following up at the time of the study. The prevalence of trilateral retinoblastoma was 1% and in secondary neoplasm, it was 0.3%.

Conclusions: Leukocoria was the commonest symptom of retinoblastoma, with unilateral involvement being the majority. Bilateral cases had a higher proportion of tumors in stage A at diagnosis, and unilateral cases had a higher proportion of tumors in stages B and C at diagnosis, compared to bilateral cases. The mean age of unilateral cases was 8.48 ± 4.144 months and of bilateral cases was 7.32 ± 3.975 months. Only a few patients had a history of consanguinity and a similar family history. Moreover, the histopathology of enucleated eyes revealed that most cases had a moderately differentiated RB. Furthermore, only a minority of cases had their optic nerve involved. Further studies with larger sample sizes are recommended to confirm these findings.

Keywords: Clinical characteristics of Retinoblastoma, Sri Lanka,