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Causes of delayed presentation of retinoblastoma: A single centre study

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Abstract---Aim: The aim of the study was to determine the Causes of Delayed Presentation of Retinoblastoma. Materials and methods: This study was carried out at the department of ophthalmology DHQ TH Bannu from January 2022 to January 2023. In this research newly recruited patients were included who were receiving therapy for retinoblastoma during that time. For this study, parents of patients who came into the hospital were gathered. A form via the patient's information was noted. First symptom, duration after onset of initial symptom, gender, age at first visit. The data analysis software used was SPSS version 20. The importance of each factor was determined using the Pearson Chi Square test. Results A total of 80 participants were included in the study. 52(65%) patients had delayed presentation, 57(71%) had delayed referral presentation and 65(81%) had advanced diseases at the time of presentation. Various factors involved in the delay presentation. These factors are, living in a remote region; financial hardship; social pressure; knowledge of the severity of the illness; lack of transportation; Fear of enucleation and looking for alternative medications or non-medical treatments. Each factor significance was calculated through chi-square test. Conclusion: Our study concludes that the different factors associated with the delayed presentation of Retinoblastoma were living in a remote region; financial hardship; social pressure; knowledge of the severity of the illness; lack of transportation; Fear of enucleation and looking for alternative medications or non-medical treatments. The reasons why health services are not developing in rural regions might be emphasized at the national level.

Keywords---Retinoblastoma, Cancer, Malignant tumor, Leucocoria.

Introduction

The most prevalent intraocular cancer in babies is retinoblastoma that affects one in 15,000 to One in 20,000 live births (1). This leads to about nine thousand additional cases annually on a global scale. Bilateral illnesses, which is inherited and a component of a genetic cancer risk syndrome, affects 40% of patients. Ten to fifteen percent of individuals with a unilateral form and all kids with either the bilateral or familial type have an RB1 gene mutation. It cases are not inherited in 60% of situations (2, 3). In unilateral circumstances, the median age of diagnosis is twenty-four months, but in bilateral cases, it is between nine and twelve months (4). In the developed nations, leucocoria and strabismus are the most typical clinical manifestations. Nonetheless, retinoblastoma people living in developing nations usually have proptosis, buphthalmos, and bright-colored eyes (5). Many tertiary centers have chosen to treat retinoblastoma patients with intra-arterial chemotherapy (IAC) as their first-line therapeutic choice. In addition to group A, B, and C, IAC may be useful in sparing the eyes of Group D retinoblastoma patients whose disease had progressed beyond the point of systemic treatment and was headed for enucleation.(6) Extra ocular retinoblastoma still has a poor prognosis; the stated survival rate for this condition varies from 50% to 70%. When comparing the industrialized world, where the majority of cases are intraocular, to developing nations, where extra ocular illness is more common due to delayed presentation, the overall survival rate for patients with retinoblastoma remains poor.(7) A research conducted in London found that the five-year survival rate among kids with unilateral retinoblastoma rose from 85% in the 20th century to 97% in the 21st.(8) In Germany, the annual overall survival rate is more than 95%.(9) Not everywhere in the globe can be expected to see this incredible progress. According to statistics from Africa, 20% of people will survive without illness. (10) Kivelia has penned an essay outlining the progress made in the medical industry over the past century. He came to the conclusion that it may take a further 200 years for the benefits to become widely distributed (11) .this study was conducted to find out Causes of Delayed Presentation of Retinoblastoma in a tertiary care hospital

Material and Methods

This study was carried out at the department of ophthalmology DHQ TH Bannu from January 2022 to January 2023. In this research newly recruited patients were included who were receiving therapy for retinoblastoma during that time. Parental complaint time (lag time 1) was defined as the period of time between the parent's initial diagnosis of the illness and the visit to the physician or ophthalmologist. When a presentation was delayed, it was classified as having a lag time of one if it lasted more than two weeks. The period of time between the initial visit by a doctor or general practitioner to the presentation at The Hospital, was designated as the referral time (lag time 2). A referral was considered delayed if there was a lag period of more than two weeks. Total delay was calculated as the difference between the onset of the first parental complaint and the first evaluation at the tertiary care hospital (total lag time = lag time 1 + lag time 2). (12) Our patients' parents, who were primarily women, were the source of their history. The patient's information was recorded, including the first symptom, the amount of time that had passed since the symptom began, the patient's gender,

age, and the amount of time that had passed before the patient presented to the primary care physician and the tertiary care Centre. Specific physical diagnosis performed by cancer specialist and a pediatric ophthalmologist performed the examination while the patient was unconscious. Prior to beginning therapy, the disease was staged or grouped at the time of admission to a hospital with tertiary care (TNM staging and international intraocular retinoblastoma classification). The tumors were categorized as extra ocular or intraocular by means of radiological and clinical assessment. The newly diagnosed cases with extra ocular or worsening eye retinal tumors with category D or E were classified as having an advanced illness and added to the research. Parents were questioned on the difficulties they faced when caring for their children. All of the parental participants in the study completed a questionnaire outlining the main concerns with the assistance of paramedic personnel. SPSS version 20 was used to analyze the data. Each cause's significance was determined using the Pearson Chi Square test.

Results

94 patients visited to the hospital diagnosed with retinoblastoma. Out of them 80 were further assessed in the study that fulfilled the inclusion criteria. Among them the prevalence of intraocular diseases were 61(76.2%) and that of the extra retinoblastoma was 19(23.7%). (fig1) Similarly distant metastasis was diagnosed in 4 patients and intracranial extension in 6 individuals. The occurrence of unilateral retinoblastoma was n 55(68.75%) and bilateral was 25(31.25%). The mean age were 14 months for the bilateral and 25 months for unilateral diseases. 52(65%) patients had delayed presentation, 57(71%) had delayed referral presentation and 65(81%) had advanced diseases at the time presentation. As displayed in figure no 2. In downward direction, the significance of the numerous elements related to delayed identification and cure is shown in table 1.

Discussion

In Pakistan, tertiary care hospitals offer treatment options include intravenous chemotherapy, intra-vitriol chemotherapy, and focused laser consolidation, in addition to standard chemotherapy. However, in the majority of situations, the retinoblastoma team is left with no option except to do enucleation due to the presentation of an advanced illness and limited finances (13, 14, and 15). Even though there is an excellent survival rate for severe intraocular sickness if enucleation is performed on time, fear of losing one's eyes, cultural norms, and alternative medicine may all be factors in a bad outcome. In this country, there is also low treatment compliance (16). Appropriate screening undoubtedly aids in an early illness diagnosis. For a variety of reasons, the majority of malignancies in underdeveloped nations—including Pakistan—take longer to diagnose (17). This has been especially noted in the majority of retinoblastoma investigations conducted in Pakistan to date, as the majority of these individuals had advanced ailments (18). In Pakistan, the availability of innovative therapeutic alternatives, such as intravascular chemotherapy, has evolved quickly.(19) Not much has been done so far to identify early tumors (IIRC Groups A as well as B) so we can save the patient's life and globe in addition to their usable vision. Consequently, we are unable to preserve these individuals' functional eyesight. The primary

predictor identified in our study for the delayed manifestation of retinoblastoma is the presence of individuals residing in isolated rural locations with inadequate access to healthcare services. In Pakistan, 21% of people live below the poverty level line while 61.4% of people reside in rural regions.(20) The front-line healthcare worker is the main staff member. To get to a qualified general practitioner, they must trek many miles. Their suffering is exacerbated by their precarious financial situation. They may want a loan in order to get to a neighboring area for a doctor's appointment. Patients who live in restricted communities are frequently discouraged from getting appropriate medical advice due to familial pressure. The sum of all the other components is ignorance. Parents can mistake leucocoria, the initial symptom of retinoblastoma, for a lucky charm. In most of the subcontinent, seeking non-medical therapy for practically any condition is a prevalent practice. When there are no qualified medical professionals present and faith healers are easily accessible, they are frequently the first to be sought for treatment. These folks in particular have strong roots in the community's beliefs. Or fatal illnesses, a large number of individuals visit them before it's too late.(21) A significant contributing factor to postponing treatment for retinoblastoma is anxiety of enucleation. An organ loss has a profound effect on a person's mind. The parents of many patients deny enucleation in an attempt to save the child's life. They continue looking for a practitioner who is capable of treating without enucleation. This results in the loss of significant time. Similar explanations have already been documented in other emerging nations, including India, which has a similar social structure and demographics.(22) Leucocoria detection in infants, particularly in children who are at risk, can improve the course of the illness. It is impossible to dispute the importance of genetic counselling in the early diagnosis and management of cancer in cases of hereditary retinoblastoma. It is possible to use social media's influence to raise awareness of serious illnesses like retinoblastoma. Many centers in poor nations, like India, have launched awareness campaigns concerning retinoblastoma, which have aided in the disease's early discovery. Addressing these factors at the national level is crucial. In this context, improved medical education and instruction for healthcare providers about the diagnosis of retinoblastoma is crucial. The single-center study is its major shortcomings, particularly in light of the significance of the topic. Doing such surveys at the national level will assist in the development of health care guidelines.

Conclusion

Our study concludes that the different factors associated with the delayed presentation of Retinoblastoma were living in a remote region; financial hardship; social pressure; knowledge of the severity of the illness; lack of transportation; Fear of enucleation and looking for alternative medications or non-medical treatments. The reasons why health services are not developing in rural regions might be emphasized at the national level.

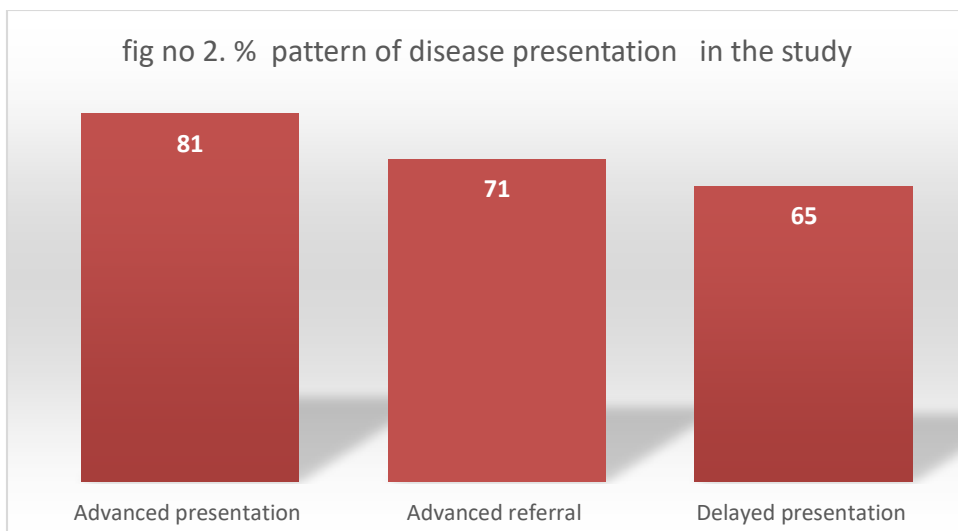
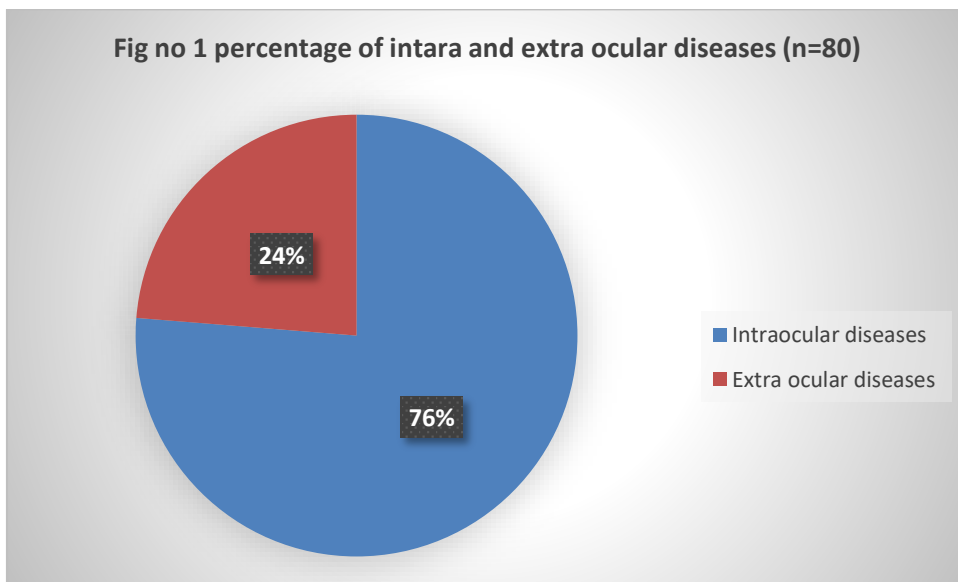


Table 1: Percentage and importance (chi-square test) of a number of causes accountable for delayed presentation of retinoblastoma

Causes	%	Importance Chi-square Test
Rural regions	56	0.593
Insufficient funds	66	0.407
social compulsion	30	0.365
Knowledge of the nature of disease	62	0.015
Absence of transportation	56	0.004
Fear of eye removal	40	0.001

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