Original Research Paper

Volume-9 | Issue-7 | July - 2019 | PRINT ISSN No. 2249 - 555X

Oncology

RETINOBLASTOMA : TIME RIPE TO SCREEN?

Vinay Munikoty Venkatesh	Assistant Professor, Division of Pediatric Hematology Oncology, Department of Pediatrics, St. John's Medical College Hospital, Bangalore, Karnataka, India
Vandana Bharadwaj	Associate Professor, Division of Pediatric Hematology Oncology, Department of Pediatrics, St. John's Medical College Hospital, Bangalore, Karnataka, India
Tarangini Durgappa*	Assistant Professor, Division of Pediatric Hematology Oncology, Department of Pediatrics, St. John's Medical College Hospital, Bangalore, Karnataka, India *Corresponding Author
Ashish Mallige	Senior Resident, Division of Pediatric Hematology Oncology, Department of Pediatrics, St. John's Medical College Hospital, Bangalore, Karnataka, India

ABSTRACT Retinoblastoma is the most common pediatric ocular tumor. Early diagnosis and treatment translates to not just excellent survival but also preservation of intact vision in these children. The developing countries face a huge burden of advanced stage disease primarily due to delayed diagnosis. This article highlights the need for institution of routine screening for retinoblastoma.

KEYWORDS :

INTRODUCTION

Retinoblastoma is the most common intraocular malignancy in childhood. It contributes to 3% of all pediatric cancers. The scenario in developing countries as compared to developed countries is different, in the sense that burden of the disease is more and the survival rates are less. The outcomes are impressive if diagnosed early and treated appropriately. Diagnosis made in a advances stage often confers poor prognosis. This review highlights the need for screening methods for early diagnosis.

INCIDENCE

The incidence of retinoblastoma is 1 in 15,000 to 18,000 live births. As per the American registry, 280 new cases of retinoblastom were diagnosed in 2014(1). The age adjusted incidence rates in US is 11 per million population and 4.1 per million population in under 5 and 15 years, respectively(2).

The Bombay cancer registry documented an incidence of 4.2 cases per million population in <15 years age group in 1998(3). However with growing awareness, accessibility to health facilities and better diagnostic tools the documented incidence has greatly increased. About 1500 cases new cases are diagnosed in India each year and age adjusted rates (<5 years) accounts to 28 cases per a million population (4). This means the burden is several times higher in developing countries.

Age at diagnosis

The average age at diagnosis is 18 months and 36 months in developed and developing countries, respectively (3). The prime factors for delayed diagnosis in developing countries include the lack of awareness and lack of screening tools to diagnose early disease. The delay in the diagnosis often leads to more advanced disease at presentation which ultimately translates to poor survival.

Delay at diagnosis

The tumor if left untreated usually invades the entire globe and destroys the vision(5). The common sites of metastasis include central nervous system (brain parenchyma and cerebrospinal fluid), bone and bone marrow. Without treatment, the median time to metastasize is 4-6 months from diagnosis. The single most important prognostic factor influencing outcome is stage (spread) of the disease at diagnosis. Intra-ocular tumors (Group A to E) have excellent prognosis wit survival as high as 85-100%. The survival plummets to as low as 0-30% in locally advanced and metastatic disease.

The clinical profile of patients treated in developed countries is predominantly early stage with only 2-5% cases diagnosed with advanced disease. This has translated into excellent survival outcomes. The developing countries on the other hand are faced with the burden of advanced disease which accounts for almost 40% of all cases. This results in poor overall survival(6).

Survival

With the advent of better chemotherapeutic regimens and use of focal or radiotherapy appropriately, the survival rates for retinoblastoma has improved dramatically. There has been a conceptual evolution from saving the life to saving the globe and vision. Other factors that have improved survival are increasing awareness, facilities for early diagnosis and a good referral net to treatment centers.

The five year survival rates have improved from 86% in 1970 to 99% in 2005 in developed countries. The 5-year overall survival rate for children with retinoblastoma in United States is 94% (6). In a study done by Bombay cancer registry survival at 1 year was 78%, at 3 years was 73% and at 5 years was 68%, which is pretty poor as compared to developed countries (2). A child who remains recurrence free for 5 years after diagnosis is considered cured(7).

Does retinoblastoma validate a routine screening?

The attributes for a disease to qualify for routine screening (described by Wilson and Jugner in 1968) include:

1. The natural course of the disease should be well known,
2. It must have an acceptable treatment protocol that changes the outcome of the disease.

Retinoblastoma clearly satisfies both these criteria.

Newborn screening is done for rare disease like inborn errors of metabolism and storage disorders, the incidence of which is 1 in 30000 to 40000 children. With a much higher incidence and better therapeutic outcome, retinoblastoma necessitates routine screening.

A simple torch light examination of eyes and a focused history for familial inheritance of this tumor is sufficient to identify cases at an early stage. The loss of red reflex on flash light examination should guide the clinician for an early referral to ophthalmologist for a complete fundus examination. The screening of premature babies for ROP (retinopathy of prematurity) is made routine in most of neonatal intensive units. Similarly we propose that a routine screening methodology needs to be adopted for early referral of such suspected cases. These screening evaluations could be accommodated during the routine immunization visits at the health facility, annual school health check up, visit to the pediatricians for minor ailments etc.

REFERENCES:

1. Cancers facts and figures 2014: Special section: Childhood and Adolescent cancers, American cancers Registry
2. Age-adjusted and age-specific SEER cancer incidence rates, 2004-2008.
3. Yeole BB, Advani SH, Lizzy S: retinoblastoma- An epidemiological Appraisel with

reference to population in Mumbai, India: Asian Pacific J Cancer Prev, 3, 17-21
4. National guidelines in management of retinoblastoma: Indian Council of Medical research 2010
5. Hurwitz RL, Shields CL et al. Retinoblastoma: Principles and Practice of Pediatric Oncology, 6th ed Pizzo PA, Poplack DG, Philadelphia 2011. p809
6. Meel R, Radhakrishnan V et al: Current therapy and recent advances in management of retinoblastoma: Indian J Med Pediatr Oncol. 2012 Apr- Jun;33(2):80-88
7. Broaddus E, Topham A et al. Survival with retinoblastoma in USA: 1975-2004. Br J Ophthalmology 2009;93:24