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Overview of Bilateral Retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia

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ABSTRACT

Background: Retinoblastoma is the most common intraocular primary malignant tumor in children. In the United States, cases of retinoblastoma are estimated to be found in 1 in 18,000 children under 5 years. The incidence of retinoblastoma ranges from 1:14.00 to 1:34.000 live births. The highest incidence occurs in developing countries. This study aimed to provide a clinical picture of bilateral retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia, as basic data for further policy-making in order to optimize services related to the management of bilateral retinoblastoma. **Methods:** This study was a descriptive observational study with 29 study subjects. Sociodemographic, clinical, and management data are presented descriptively. Data analysis was performed with the help of SPSS software version 25. **Results:** Based on the age group, the majority of bilateral retinoblastoma cases were less than 5 years old, with the most age range being 3-4 years. The majority of therapeutic management for cases of bilateral retinoblastoma is exenteration in combination with chemotherapy. **Conclusion:** A combination of surgery and chemotherapy is the most frequently used treatment for bilateral retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia.

1. Introduction

Retinoblastoma is the most common primary intraocular malignant tumor in children. In the United States, cases of retinoblastoma are estimated to be found in 1 in 18,000 children under 5 years. The incidence of retinoblastoma ranges from 1:14.00 to 1:34.000 live births. The highest incidence occurs in developing countries.¹⁻³ These tumors may be hereditary or sporadic. Approximately 90% of retinoblastoma patients have no family history of the disease, and only 10% have a family history, such as their parents, uncles, or grandparents, and if these patients have a family history of retinoblastoma, it often occurs bilaterally. The incidence of retinoblastoma ranges from unilateral (70-75% of

cases) and bilateral (25-30% of cases). Retinoblastoma is caused by mutations in the RB1 gene, which is on the long arm of chromosome 13 at locus 14 (13q14).³⁻⁵

The most common clinical symptoms are leukocoria, strabismus, glaucoma, and protrusio bulbi. The prognosis depends on the clinical stage of the tumor at the time of diagnosis. If found at an early stage, the prognosis will be better.⁶ Although retinoblastoma is the primary intraocular tumor in children, the management of this disease is complex. Planned therapy usually requires a multidisciplinary approach consisting of an ocular oncologist, a pediatric oncologist, and a radiologist. Conservative

therapy includes photocoagulation, cryotherapy, chemotherapy, and radiotherapy. Surgical therapy includes enucleation and exenteration. The choice of therapy depends on the involvement of 1 or 2 eyes, tumor size, and disease stage of the children. Management in particular is very individual. Every effort made is the main goal of survival and maintaining their eyes and vision as much as possible.^{7,8} This study aims to provide a clinical picture of bilateral retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia, as basic data for further policy-making in order to optimize services related to the management of bilateral retinoblastoma.

2. Methods

This study was a descriptive observational study to present the clinical picture of bilateral retinoblastoma. This study uses secondary data obtained from the medical record installation of Dr. M. Djamil General Hospital, Padang, Indonesia. A total of 29 patients were included in the study. The criteria for patient inclusion are new or old bilateral retinoblastoma patients, treatment to the tumor subdivision eye polyclinic, other ward inpatients who are consulted to

the department of ophthalmology at Dr. M. Djamil General Hospital, Padang, from January 2003 to December 2015 and patients with complete medical record data. This study was approved by the medical and health research ethics committee at Dr. M. Djamil General Hospital, Padang, Indonesia.

This study presents sociodemographic data of patients. In addition, this study presents clinical data on bilateral retinoblastoma patients and presents data on the management of retinoblastoma patients at Dr. M. Djamil General Hospital, Padang, Indonesia. Data analysis was carried out with the help of SPSS software version 25. Univariate analysis was carried out in order to provide data on the frequency distribution of sociodemographic data, clinical data, and management data on retinoblastoma patients at Dr. M. Djamil General Hospital, Padang, Indonesia.

3. Results

Table 1 showed the data on the distribution of the frequency of occurrence of bilateral retinoblastoma by sex. Of the 29 patients diagnosed with bilateral retinoblastoma, 14 patients were male, and 15 were female.

Table 1. Distribution of bilateral retinoblastoma cases by gender.

Gender	Number (people)	Percentage %
Male	14	48.28
Female	15	51.72
Total	29	100

Table 2 showed the distribution of bilateral retinoblastoma cases by age. Based on the age group, the majority of bilateral retinoblastoma cases were less

than 5 years old, with the most age range being 3-4 years.

Table 2. Distribution of bilateral retinoblastoma cases by age group.

Age group	Number (people)	Percentage %
Less than 1 year	4	13,70
1-2 years	10	34,48
3-4 years	13	44,82
Over 5 years	2	6,90
Total	29	100

Management of retinoblastoma can be in the form of photocoagulation, cryotherapy, radiotherapy, and chemotherapy as well as surgery at Dr. M. Djamil

General Hospital, the management of retinoblastoma can be in the form of surgery (exenteration) + chemotherapy or chemotherapy only.

Table 3. Distribution of bilateral retinoblastoma cases based on the therapy given.

Type of therapy	Number (people)	Percentage %
Chemotherapy	10	34.48
Exenteration + chemotherapy	14	48.28
Not therapy	5	17.24
Total	29	100

Management of therapy mostly carried out at Dr. M. Djamil General Hospital, Padang, for bilateral retinoblastoma cases during 2003-2015 was exenteration in combination with chemotherapy in 14 cases (48.28%).

4. Discussion

Retinoblastoma is the most common intraocular primary malignant tumor in children. These tumors can be hereditary or sporadic inherited. Nearly 75% of cases are unilateral, and 25% are bilateral. Within 13 years (January 2003 – December 2015), there were 29 patients diagnosed with bilateral retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia. Although there was no gender predilection, in this study, there were fewer male patients with retinoblastoma, namely 14 patients (48.28%) compared to 15 female patients (51.72%). In another study, it was also found that there were more male patients with retinoblastoma than women, with a ratio of 1.6:1.^{9,10}

In most studies from various parts of the world, there is no significant difference between gender in the incidence of retinoblastoma. Other studies have shown that overall, men and women have no differences in the distribution of retinoblastoma for all ages. However, for the 0-1 year age group in the bilateral retinoblastoma category, there is a male: female ratio of 1.1:1.0. For the 1-2 year age group, for bilateral tumors, it was 1.3: 1.0 while for the 2-3 year age group it was 2.2: 1.0. In another study, there was a difference in the ratio between males and females

(3:1) in bilateral cases. However, for unilateral cases, the male-to-female ratio is 1.2:1.0. But the overall ratio is 4: 3.¹¹⁻¹³

Table 2 shows the age distribution of retinoblastoma patients in this study, where the largest age range was 3-4 years, with 13 cases (44.82%). Only 13% of patients come at the age of less than 1 year. The results of this study indicate that there is a delay in patients being treated at Dr. M. Djamil General Hospital seeking treatment. In another study between 1999 and 2002, 70 cases of retinoblastoma were found, with 93% of them in children under 5 years of age; 67% of cases are bilateral. Bilateral retinoblastoma cases become less with age, while unilateral cases peak in the 2-3 year age group, with a gradual decrease occurring thereafter. The mean age was 28.17 months, and unilateral cases had a mean age of 31.81 months. Another study conducted from 1957 to 2006 found that 1234 eyes had enucleation due to retinoblastoma at a specialist eye hospital. The mean age was 2.8 years, with a range of 1 month to 14 years. In bilateral retinoblastoma, there are 2.4% of cases.¹⁴⁻¹⁶ While in other studies, The age of the patient was found to be younger, namely in the age range of 2-3 years.^{17,18} This is often found in people in developing countries, where people don't know, especially parents, about this disease because of a lack of information through health workers or the mass media. Apart from that, it is also due to economic factors, low level of education, and lack of awareness of early detection of disease.

Treatment for retinoblastoma sufferers can be in the form of radiation, photocoagulation, cryotherapy, chemotherapy, surgery, or combination therapy of the various methods mentioned above. Treatment must be seen individually, depending on the size of the tumor, location, number of lesions, bilaterality, the presence of extra-ocular tissue expansion, and the presence of signs of distant metastases.¹⁹ In this study, the most common treatment was exenteration and chemotherapy in 14 patients (48.28%). This is in accordance with most complaints when they come, namely protrusion. The treatment is surgery followed by chemotherapy. Another study also found that in Thailand, the combination of surgery, radiation, and chemotherapy was the most common (36.51%). The results of this study and other studies in Southeast Asia show that therapy given in developing countries still aims to save the lives of sufferers, while in developed countries such as America and Europe, therapy for patients with retinoblastoma is aimed at saving vision and preserving the eyeball.^{19,20}

5. Conclusion

A combination of surgery and chemotherapy is the most frequently used treatment for bilateral retinoblastoma at Dr. M. Djamil General Hospital, Padang, Indonesia.

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