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# Clinical features of children with retinoblastoma in Adam Malik Hospital, Medan, Indonesia

Bidasari Lubis

*Department of Child Health, Medical School, University of North Sumatra/H Adam Malik Hospital, Medan, Indonesia*

**ABSTRACT** We reviewed patients with retinoblastoma treated at the Department of Child Health, Adam Malik Hospital, Medan from January 1995 to December 1999. Clinical staging was based on Reese-Ellsworth classification (when the disease was limited to the eyeball) or on St Jude Children's Research Hospital (when the disease has extended beyond the eyeball). A total 19 retinoblastoma patients were available, from which 9 were males. Fifteen patients were less than 5 years old, and the average age at diagnosis was 3 years 8 months. In most patients the disease started at the age of less than 5 years, and they presented themselves with advanced stage as usually seen in developing countries. In nine patients the retinoblastoma affected the right eye, 8 affected the left eye, and in 2 patients the tumor was bilateral. The most common presenting symptom on admission was proptosis. Pathological confirmation was obtained in all patients. Bone marrow puncture was done in 15 patients; of whom 13 showed bone marrow metastasis. Since 1998 the standard treatment for patients with proptosis in stage III or IV of the disease was pre-operative chemotherapy followed by enucleation, radiation and chemotherapy. [**Paediatrica Indonesiana 2001; 41:234-236**]

**Keywords:** *retinoblastoma, diagnosis, clinical features, prognosis*

RETINOBLASTOMA IS THE MOST COMMON PRIMARY MALIGNANT intraocular tumor of childhood and is interesting in several respects. It occurs in approximately 1 in 18,000 live births, without sex or race predilection.<sup>1</sup> Delay in diagnosis is common in developing countries; many patients seek medical attention only after orbital extension has appeared. The diagnosis is established by typical clinical features of the tumor and judicious use of ancillary diagnostic studies, including ophthalmoscopic and radiologic examinations, particularly ultrasonography and computed tomography. Early diagnosis is important because retinoblastoma has a rapid growth rate.<sup>1-4</sup> Treatment planning must be individualized on the basis of the overall clinical manifestations; various combinations of treatment may

be needed to achieve a satisfactory result. The prospect of successful treatment depends upon whether the tumor is still confined within the eye at the time of diagnosis. The prognosis in retinoblastoma depends on the size and location of tumors and the degree of ocular and extraocular involvement. Extension into the orbital tissue, or evidence of metastases, is an indication of a poor prognosis.<sup>4-7</sup> Reports on retinoblastoma in Indonesia have been scarce. We reviewed the clinical characteristics of patients treated at the Department of Child Health, Adam Malik Hospital, Medan since January 1995- December 1999.

## Methods

Subjects of this study were patients with retinoblastoma admitted to the Pediatric Department, North Sumatra University/H Adam Malik Hospital, Medan from January 1995 to December 1999. The

clinical data consisted of age, sex, clinical history, symptoms and signs, laboratory findings, lumbar puncture, bone marrow puncture, head CT-scan, histopathological examination, and ophthalmologic examination. Staging of the disease was based on the recommendation of Reese-Ellsworth when the disease was limited to the eyeball, and of St Jude Children's Hospital for disease that has extended beyond the eyeball. All patients had histological examination of the tumor tissue.

## Results

Nineteen retinoblastoma patients were included in this study, from which 9 were males. The average age at diagnosis was 3 years 8 months (range: 1 year 6 months to 6 years), from which 15 patients were diagnosed at the age of less than 5 years. The right eye was equally affected to the left one, i.e. 9 patients who had retinoblastoma of the right eye and 8 had the tumor of the left eye. Only in 2 patients were the tumor involved both eyes on admission. See **Table 1**.

**TABLE 1. PATIENTS' CHARACTERISTICS**

Characteristic	Total
<b>Sex</b>	
Males	9
Female	10
<b>Age group</b>	
<5 years	15
≥5years	4
Range: 1 year 6 months to 6 year (Average: 3 years 8 months)	
<b>Location of tumor</b>	
Right eye	9
Left eye	8
Both eyes	2

Most patients were admitted to the Department of Child Health because of proptosis (14/19), the other 5 patients were referred by ophthalmologists following surgical intervention, i.e. enucleation. It appears that before 1997 children with retinoblastoma were admitted to Ophthalmology Department, while after 1997 they were admitted to the Department of Child Health.

The majority of patients had advanced disease (**Table 2**). The most common primary condition on admission was proptosis, i.e., in 14 patients. From 1995 to 1997 ophthalmologist consulted 5 patients after nucleation but since 1998 to 1999 ophthalmologist consulted 14 patients to be evaluated and stag-

ing of tumor and planning of the treatment. From 19 patients retinoblastoma mostly less than 5 years (79%) and there is no other difference according to gender. Bone marrow aspiration was performed only 15 patients, 13 showed metastases on marrow.

**TABLE 2. PRIMARY CONDITION**

Condition of the eye	Total
Post enucleation + radiation	3
Post enucleation	1
Relapses post enucleation	1
Proptosis	14

## Discussion

Retinoblastoma is typical malignant intraocular tumor of infancy and childhood and is fortunately very rare. From 1995 till 1999 we only found 19 patients with retinoblastoma in our department. Nasution R et al. in 1980-1988 found 46 cases retinoblastoma in Dr. Pirngadi Hospital Medan.

The diagnosis of retinoblastoma is established by recognition of typical clinical features of the tumor, ophthalmoscopic examination by ophthalmologist in combination with radiologic examination particularly ultrasonography and computed tomography. Bone marrow puncture and lumbar puncture are also needed. Donalson et al, found no sex predilection and we did not found difference according to gender in retinoblastoma patients. Some authors said that retinoblastoma cases usually found in children less than 5 years of age and in our retinoblastoma patients mostly in less than 5 years.<sup>2-5</sup>

Early recognition of this malignancy is important and life saving; however most of the patients in our series presented with was proptosis, which was very late. This probably reflects the condition in most developing countries, where late diagnosis is a rule. Diagnosis being made only after enlarged eye or gross orbital extension.

Treatment of retinoblastoma depends on the stage of the tumor at diagnosis; it is usually followed by long term follow up and periodic examination of the child. Treatment options vary and include: enucleation, total irradiation, focal irradiation, cryotherapy, photocoagulation and chemotherapy. Enucleation of the eye is still the commonest form of treatment and may result in complete cure. Retinoblastoma are very radiosensitive and radiotherapy is the cornerstone of conserva-

tive management, usually to a total dose of 3500-4000 rads in three to four weeks. The indications for chemotherapy are not clearly established, and some controversy still exists about its value. Chemotherapy is generally used in patients with orbital, intracranial and metastatic disease. In most solid tumor patients chemotherapy is given before surgical treatment but in retinoblastoma pre-operative chemotherapy is not yet routine. Decision about further treatment enucleation, chemotherapy and radiotherapy depend on which stage, whether involvement is unilateral or bilateral, whether there is vision or any potential for vision, whether the tumor is confined to the globe or there is extension to the optic nerve.

Pre-operative chemotherapy had been given in mostly patients since 1998-1999 because the patients commonly with metastatic disease (stage III-IV). From this study we are planning to evaluate the direct benefits of pre-operative chemotherapy for stage III and IV retinoblastoma. The management of retinoblastoma can be extremely difficult, and physicians must often make difficult decisions about the use of various treatment modalities. The hope for the future is that more children will preserve the affected eye with some or all of its vision and with minimal side effects from treatment.<sup>6-8</sup>

In summary retinoblastoma patients in our department were mostly diagnosed at the age of less

than 5 years and there was no gender preponderance. The most common primary condition on admission was proptosis. This advanced stage at diagnosis made the prognosis of most of 'patients' grave.

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