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The role of demographic features, pathologic subtype and classifications on prognosis in patients with Rhabdomyosarcoma referred to Iran Cancer Institute

M. Babaei[1], E. Esmati[1], A. Maddah Safaei[1], H. Nosrati[1*], M. Hadji[2], S. Ahmadi[2], S.R. Madani[3], M. Ganjalikhani[4]

[1]Department of Radiotherapy Oncology, Cancer Institute, Tehran University of Medical Sciences, Tehran, Iran
[2]Cancer Research Center, Cancer Institute, Tehran University of Medical Sciences, Tehran, Iran
[3]Shahid Beheshti University of Medical Sciences, Tehran, Iran
[4]Tehran University of Medical Sciences, Tehran, Iran

► ***Short report***

**** Corresponding author:***
Mr. Hassan Nosrati,
Fax: *+98 21 61192584*
E-mail:
hassannosrati@yahoo.com

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ABSTRACT

Background: Rhabdomyosarcoma is a rare malignant soft tissue sarcoma and the most common sarcoma of childhood. The purpose of this study was to investigate the distribution of age and sex of patients, pathological subtypes and to determine the role of staging and classification of RMS on treatment outcome. ***Materials and Methods***: This retrospective study included patients who diagnosed with RMS in Cancer Institute between 2006 and 2013. We used the Kaplan Meier and life table methods to estimate survival rate. STATA statistical software ver. 11.2 was used for statistical analyses. ***Results***: Thirty patients with a mean age of 15.1 years (±SD =18.1) were evaluated.66.6 Percent were males. Median follow-up for survivors was 57.3 months (±SD=32.4). Survival rates of patients were as follows: 6 months (94%), 1 year (87%), 3 years (69%), and 5 years (50%). The pathology review classified of tumors as botryoid (6.7%), spindle cell (6.7%), embryonal (40%), alveolar (33.3%) and undifferentiated (13.3%). 3 years survival for patients with international classification III (alveolar and undifferentiated) was 70%, International classification II (embryonal) was 67%. Due to small sample size, we can't report 3 years survival for international classification I (botryoid and spindle cell). ***Conclusion***: Our results support early age of onset (>50% of RMS cases are diagnosed before age 10 years). Age of diagnosis often gives key facts about clinical behavior and has a noticeable effect on 5 year survival; it is a prognostic factor in RMS. Disease extent as well as age and histology affect survival.

Keywords: *Rhabdomyosarcoma, survival, childhood, sarcoma.*

INTRODUCTION

Rhabdomyosarcoma (RMS) is a rare malignant soft tissue sarcoma. It arises from unsegmented, undifferentiated highly malignant mesoderm or myotom-derived skeletal muscle.

This type of sarcoma may be derived from any part of the body, but common sites are; orbit (9%), head and neck (non parameningeal) (7%), parameningeal (15%), genitourinary (31%), extremities (13%), trunk (5%), retro peritoneum (7%), and other sites (13%)[1].

Rhabdomyosarcoma is the most common sarcoma of childhood. At the time of diagnosis, the majority of patients have less than 10 years old [2]. Two peaks of age exists: 1- between 2 and 6 years and 2- adulthood [3].

Rhabdomyosarcoma often has pseudo-capsule and can spread locally along muscle plane or fascia. It may have lymphatic or hematogenous dissemination. At the time of diagnosis, lymphatic spread is detected in approximately 15% of patients [4] and the risk of hematogenous metastasis is approximately 15% [5].

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As Rhabdomyosarcoma has the potential to be occurred in any site, clinical signs and symptoms may be vary. It usually present as an asymptomatic mass. While it became symptomatic, mass effect is the prominent symptom.

Rhabdomyosarcoma treatment consists of surgery, Radiotherapy and chemotherapy [6]. 5-year survival rates of RMS is 40-54% in adults and 70% in children [7]. Rhabdomyosarcoma is rare, but highly malignant tumor that includes several histological subtypes and originates almost anywhere in the body. The purpose of this study was to investigate the distribution of age and sex of disease and pathological subtypes, and to determine the role of staging and classification of RMS on treatment outcome.

MATERIALS AND METHODS

This retrospective study included patients diagnosed with RMS in cancer institute between 2006 and 2013. This study has been done due to accepted proposal (date: 1390/12/5 and referral number: 91-01-51-17217) in cancer research center of Iran cancer institute. We followed patients from the date of diagnosis until death or year 2013 whichever came first. Follow-up information was obtained through patient's medical records and telephone interviews with them or their close relatives. Information about age, sex and date of diagnosis for each patient was abstracted from hospital records. Retrieval of the patient data was performed by one of the authors, who were in treatment team. In addition, we hired an independent interviewer to perform telephone interview and obtain the follow-up information. We believe that the information retrieval was not subject to bias.

Definitive diagnosis was based on pathological examination. Data of patients with RMS were reviewed and they were called for follow up. Details regarding age, gender, stage at diagnosis, International classification of Rhabdomyosarcoma (based on pathologic characteristics) and survival rate were collected from the medical records. Patients' treatment modalities were not analyzable because these treatments including surgery, radiotherapy and chemotherapy were the same for most patients.

Statistical analyses

Data were statistically described in terms of mean, standard deviation, median, frequencies (number of cases) and relative frequencies (percentages). We performed survival analyses among patients after excluding the patients who could not find them for the follow-up. We used the Kaplan Meier and life table methods to estimate median survival and the six-month to five-year survival rates. STATA statistical software ver. 11.2 was used for statistical analyses.

RESULTS

Characteristics of the patients

Thirty patients with a mean age of 15.1 years (±SD = 18.1) were evaluated. 66.6 Percent (20 patients) were males and 33.3 percent (10 patients) were females. The pathology review classified of tumors as botryoid (6.7%), spindle cell (6.7%), embryonal (40%), alveolar (33.3%) and undifferentiated (13.3%). Genders, age at diagnosis, RMS classification and stage at diagnosis are shown in table 1.

Median follow-up for survivors was 57.3 months (±SD = 32.4).

Table 1. Characteristics of Rhabdomyosarcoma study patients.

Variable	Number	Percentage (%)
Gender		
Males	20	66.7
Females	10	33.3
Age at Diagnosis (year)		
≤10	16	53.3
>10	14	46.7
International classification		
I (Superior Prognosis)	Botryoid [2] Spindle cell [2]	6.7 6.7
II (Intermediate prognosis)	Embryonal [12]	40
III (Poor Prognosis)	Alveolar [10] Undifferentiated [4]	33.3 13.3
Stage at diagnosis		
I	12	40
II	5	16.7
III	12	40
IV	1	3.3

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Survival

Survival rates of patients were as follows: 6 months (94%), 1 year (87%), 3 years (69%), and 5 years (50%). In the overall survival curve (figure1), proportion of patients survived is shown versus time. The slope of the curve is similar and monotonic in all parts and there is no significant difference. 3 years survival for patients with international classification III (alveolar and undifferentiated) was 70%, International classification II (embryonal) was 67%. Due to small sample size, we can't report 3 years survival for international classification I (botryoid and spindle cell). The overall Kaplan-Meier survival curve are shown in figure 1 (a); and Kaplan-Meier survival curve of patients according to stage, age at diagnosis and pathologic characteristics (international classification) in the current series is shown in figure 1 (b), (c) and (d) respectively.

DISCUSSION

The current study evaluated survival among patients with RMS. Our study included RMS cases, including both genders from different ages and stages regardless of treatment modality. It has been shown that good prognosis is associated with younger age at diagnosis, localized disease, and embryonal subtype of RMS. In this study 5-year survival rate was 50 % which is consistent with results of other studies. In two studies, 5-year survival rate of 50% and 56.9% have been reported respectively [1, 8].

In our study, 3-year survival rate was 69 %, whereas in the study by Stevens MC. and colleagues, reported 3-year survival rate was 84% [9]. This difference may be due age and ethnic differences and different stage of disease.

Our results are consistent with previous reports of the incidence of rhabdomyosarcoma and its distribution by age and gender (>50% of cases of RMS are diagnosed before age 10 years and the prevalence was higher in males). Age at diagnosis often gives key facts about clinical behavior. It is reported that age has a noticeable effect on 5-year survival rate [10] and it is a prognostic factor in RMS [11].

In a pooled analysis of 788 patients with metastatic RMS, it has been shown that age is an adverse prognostic factor (> 10 years and < 1

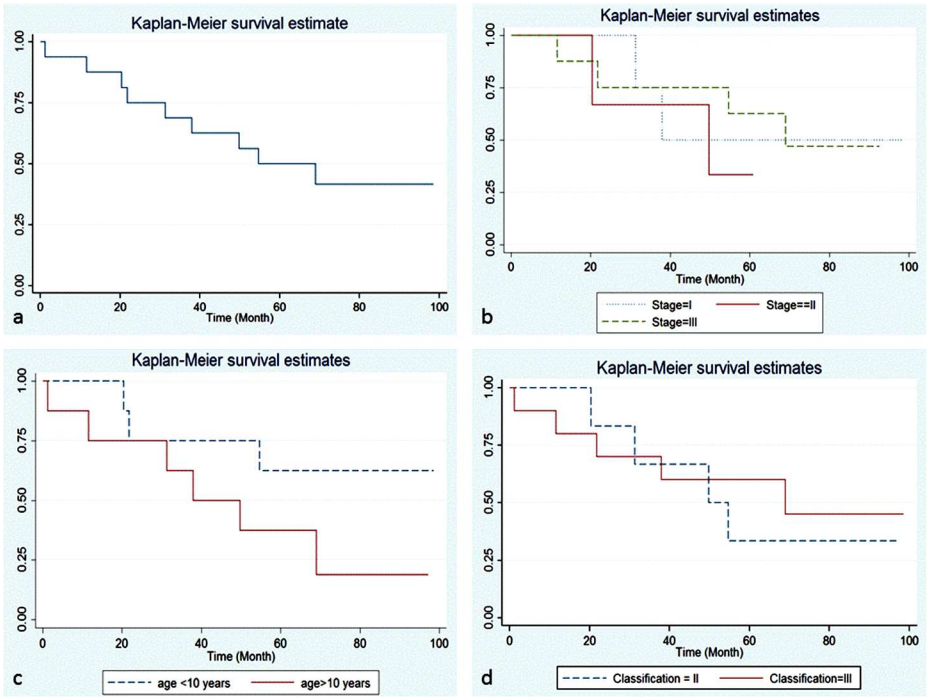

Figure 1. Kaplan-Meier survival of RMS study patients, (a): overall survival, (b): based on stage, (c): based on age, (d): based on international classification and pathologic characteristics.

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year)[12]. Also in another study, the survival results were better for children. It is explained by higher incidence of adverse prognostic factors in older patients. In older patients with RMS, unfavorable tumor characteristics (eg, alveolar subtype, lymph node involvement, and metastases at the time of diagnosis) were more common in adolescents than in children [13].In our analysis, the survival results were better for children. The presence of metastatic disease is predictor of clinical outcomes in patients with RMS. Reasonably, patients with fewer metastatic tumor sites would have better treatment outcomes. A study reported a significant correlation between metastasis and outcome (p=0.013); 66.7% of patients with metastasis at time of diagnosis had died while 82.6% of patients without metastasis were surviving [1]. Similarly, Breneman *et al.* reported that patients with metastatic disease at diagnosis have the worst prognosis [5]. Our study was in accordance with these results and stage IV has the poorest survival.

CONCLUSIONS

Disease extent as well as age and histology can affect survival. The poor survival of patients with metastatic disease requires more therapeutic modalities.

Conflicts of interest: *none to declare.*

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