

SPECTRUM OF RHABDOMYOSARCOMA—A STUDY OF 53 CASES AT ARMED FORCES INSTITUTE OF PATHOLOGY, RAWALPINDI (PAKISTAN)

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ABSTRACT

Objective: To study the spectrum of rhabdomyosarcoma, diagnosed at Armed Forces Institute of Pathology (AFIP), Rawalpindi (Pakistan).

Study Design: Descriptive study.

Place and Duration of Study: AFIP, Rawalpindi from 1st Jan 2009 to 31st Dec 2013.

Material and Methods: The study included all the cases diagnosed as rhabdomyosarcoma from 1st January 2009 to 31st Dec 2013. Records of the malignant tumors of soft tissue that presented during this period were analyzed and out of these spectrums rhabdomyosarcoma was studied. The data analysis included the age, gender, site of tumor and histopathological pattern of Rhabdomyosarcoma.

Results: A total of 19140 malignant tumors were diagnosed at AFIP Rawalpindi from 1st Jan 2009 to 31st Dec 2013. Out of these, 512 (2.67%) were malignant soft tissue tumors. Out of these 53 cases were rhabdomyosarcoma with an overall frequency of 0.27% of the malignant neoplasm and 10.3% of soft tissue sarcomas. The age ranged from 1–70 years. Out of these 53 cases 36 were males and 17 were females with a male to female ratio of 2.1:1. The most common subtype of tumor seen was embryonal rhabdomyosarcoma n=22 (41.5%). The age for embryonal rhabdomyosarcoma ranged from 3 to 71 years with median age of 16 years while most of the patients are effected during 1st decade of life (55%). Out of 20 cases of rhabdomyosarcoma during the first two decades of life 18 (90%) were embryonal rhabdomyosarcoma. Thirty cases of rhabdomyosarcoma encountered during the adult life contained only 4 cases (11%) of embryonal rhabdomyosarcoma. The majority of the tumors originated from lower extremities (41%) and in lower extremities most common site was thigh (27%). A total of 18% of the tumors originated in head and neck region. Second most common type of the tumor was pleomorphic rhabdomyosarcoma n=21 (39.6%) with age range 40 to 78 years and median age of 60 years. Ten cases were reported during 6th and 7th decade of life. Majority of the cases occurred in lower extremities (47.6%) and thigh was the most common site (42%). Alveolar rhabdomyosarcoma n=6 (11.32%) was the 3rd most common tumor with age range of 15 to 46 years and median age of 29 years. Four cases (67%) occurred in 3rd and 4th decades of life. The most commonly involved region was lower extremities (50%) followed by upper extremities (17%). Among lower extremities again thigh was the most common site (33%). Anaplastic rhabdomyosarcoma n=4 (7.54%) was the least common type of rhabdomyosarcoma with age range of 12 to 45 years and median age of 31 years. Three cases (75%) occurred in the 3rd and 4th decade of life. Two cases (50%) occurred in lower extremities and one case (25%) in the upper extremity.

Conclusion: Embryonal rhabdomyosarcoma is the most common rhabdomyosarcoma during the first two decades of life. While pleomorphic rhabdomyosarcoma is the most common rhabdomyosarcoma encountered in adults. Extremities are most common site of involvement by rhabdomyosarcoma followed by the head and neck and abdomen in our setup. This finding needs to be evaluated by a larger scale study.

Keywords: Alveolar RMS, Rhabdomyosarcoma, Soft tissue sarcoma.

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INTRODUCTION

Soft tissue sarcomas account for upto 3%

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of childhood cancers and upto 1% of adult cancers. Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor with skeletal muscle cell morphology. It is one of the tumors of muscle origin¹. It is derived from primitive rhabdomyocytes which differentiate into mature skeletal muscle. RMS arising from

mesenchymal cells is the most common soft tissue tumor in children and adults². The overall 5-year survival rate with RMS was approximately 61% for children and 27% for adults³. Younger children had higher survival rates than older children and adolescents. Children with embryonal RMS have a more favorable prognosis than children with alveolar RMS⁴.

Based on histological features, RMS have been divided into four main classes, embryonal RMS, alveolar RMS pleomorphic RMS and undifferentiated type^{5,6}. Embryonal RMS has three variants, botryoid variant, spindle cell variant and anaplastic variant. Most common class of RMS is embryonal mostly associated with tumors of head and neck and genitourinary tract⁷. The objective of the study was to analyze the spectrum of RMS including frequency, age, gender distribution, site of origin and histopathological pattern of RMS, diagnosed at Armed Forces Institute of Pathology (AFIP), Rawalpindi (Pakistan).

MATERIAL AND METHODS

Records of all the malignant tumors which were diagnosed at AFIP, public and private

Tissue Tek, VIPTM. Three to five micron thin sections were prepared, stained with H&E stain. Immunohistochemical panel including S-100, SMA vimentin, Desmin, and Myogenin were applied for confirmation of the diagnosis. Out of these records, the malignant tumor of soft tissue were retrieved and then the record of RMS from this data was extracted. The age, gender of patient and anatomic location of tumor was noted. The tumors were classified according to WHO classification of soft tissue tumor. The sample size was 53 cases. Cases were selected by non probability consecutive sampling technique and statistical analysis was done by using SPSS calculator version 17.0. Descriptive statistics were applied to summarize the data. Frequencies and percentages were calculated for qualitative variables.

RESULTS

A total of 19140 malignant tumors were diagnosed at AFIP, Rawalpindi from 1st Jan 2009 to 31st Dec 2013¹². Out of these, 512 (2.67%) were found out to be malignant soft tissue tumors. Out of these 53 (10.3%) cases were RMS. Overall frequency of malignant soft tissue

Table-1: Age distribution of Rhabdomyosarcoma.

Age Range	Frequency	Percent
1 to 10 years	14	26.4
10 to 20 years	8	15.1
20 to 30 years	3	5.7
30 to 40 years	6	11.4
40 to 50 years	5	9.4
50 to 60 years	5	9.4
60 to 70 years	9	16.9
70 to 80 years	3	5.7
Total	53	100.0

Table-2: Histopathological types & frequency of subtypes of rhabdomyosarcoma.

Subtypes of rhabdomyosarcoma	Frequency	Percentage
Embryonal	22	41.5
Pleomorphic	21	39.6
Alveolar	6	11.32
Anaplastic	4	7.54
Total	53	100.0

sector hospitals in Northern Pakistan. The biopsies were received in 10% formalin and were processed in automatic tissue processor,

tumors was found out to be 2.67% while RMS constituted 10.3% of soft tissue sarcomas.

The age ranged from 1 to 80 years. Age statistics were analyzed for different age groups and the result showed that the tumor occurred predominantly in 1st and 7th decades. (Table-1). Out of 20 RMS encountered in the first two decades of life, 18 (90%) were embryonal RMS. However out of 35 rhabdomyosarcomas encountered in adults and elderly only 4 (11%) were of embryonal subtype.

Regarding gender distribution out of 53 cases of RMS 36 were males and 17 were females with a male to female ratio of 2.1:1. Regarding site of malignancy, it was observed that 29 cases (55%) originated in extremities and the second most commonly site was head & neck accounting for 5 cases (9.4%) followed by abdomen accounting for 5 cases (9.4%). The

sarcomas. About 350 new cases of RMS occur each year in the United States⁸.

Embryonal rhabdomyosarcoma (ERMS) is the most common type of ERMS and is the predominant subtype found in childhood and young adults⁹. Similar results are obtained in our study in which ERMS constituted the predominant subtype accounting for 41.5% of the cases. Newton et al showed ERMS to be the predominant subtype accounting for 49% of their cases of RMS¹⁰. Alveolar RMS was the second most common subtype accounting for 31% of their cases¹⁰. The second and third most common RMS in our study are pleomorphic and alveolar subtypes accounting for 39.6% and 11.3% respectively. The difference in the prevalence of different subtypes may be due to

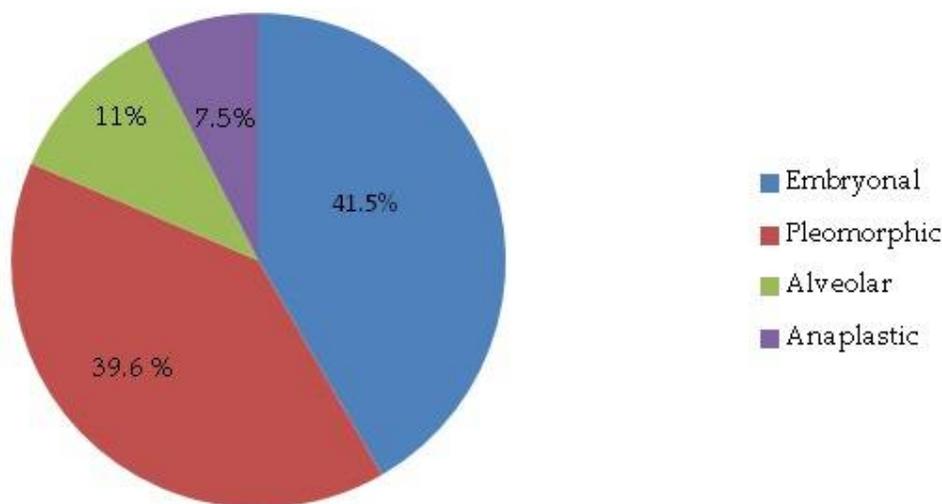


Figure-1: Histological pattern and frequency of rhabdomyosarcoma (n=53).

most common histological tumor type seen was embryonal RMS n=22 (41.5%). The second most common type was pleomorphic RMS n=21 (39.6%) followed by alveolar RMS n=6 (11.32%) and anaplastic RMS n=4 (7.54%) (table-2).

DISCUSSION

Soft tissue sarcomas account for up to 3% of childhood cancers and up to 1% of adult cancers¹. RMS arising from mesenchymal cells, is the most common soft tissue tumour in children and accounts for up to 50% of

the lesser number of cases in our study or due to the geographical variation depending on genetic and environmental factors. However pleomorphic RMS was reported as the predominant RMS in adults by Ghavimi et al¹¹. In our study also pleomorphic RMS was the predominant subtype diagnosed in adults constituting 63.6% (21/33) of the cases diagnosed in adults from third to seventh decade of life. According to Sultan et al also pleomorphic RMS is frequently encountered in adults³.

ERMS accounts for 49% of the cases diagnosed in young children¹². Eighteen out of

twenty cases of the RMS during the first two decades in our study were of the embryonal subtype constituting 90% of the cases. The male to female ratio in our study was 2.1 : 1. Slight male predominance is also reported in a number of other studies¹³.

According to Pappo et al the common sites of primary involvement of rhabdomyosarcoma included head and neck region (35%), followed by genito urinary and lower extremity¹⁴. However in our setup most commonly involved site for RMS was extremities (55%), followed by head and neck (9.4%) and abdomen (9.4%). It may be due to less number of cases being reported in our set up. Alveolar rhabdomyosarcoma (ARMS) affects all age groups and is found more often in the arms, legs, or trunk¹⁵. In concordance with this study four out of six cases of alveolar subtype in our study were reported from extremities. We diagnosed 53 cases of RMS at AFIP during the last five years (2009 –2013). In our setup, ERMS is the most common RMS, while the most common rhabdomyosarcoma world wide is also ERMS. The second and third most common RMS is pleomorphic and alveolar rhabdomyosarcoma.

CONCLUSION

ERMS is the most common RMS during the first two decades of life. While pleomorphic RMS is the commonest RMS encountered in adults. Extremities are most common site of

involvement by RMS followed by the head and neck and abdomen in our setup.

CONFLICT OF INTEREST

The authors of this study reported no conflict of interest.

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