DOI: https://doi.org/10.54393/pbmj.v4i2.253

PAKISTAN BIOMEDICAL JOURNAL

https://www.pakistanbmj.com/journal/index.php/pbmj/index

Volume 4, Issue 2 (Jul-Dec 2021)

Original Article

Epidemiological and Histopathologic Study of Rhabdomyosarcoma Cases In A Tertiary Care Hospital of Peshawar

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ARTICLE INFO

Key Words:
Embryonal type, alveolar type, Peshawar, pleomorphic, Rhabdomyosarcoma (RMS)

How to Cite:
Manzoor, M., Ahmad, B., Sarwar, A., Khan, M., Ilah, I.-u, Mir, H., Awais, M. ., Bangash, S. A., & Muthanna, F. (2021). Epidemiological And Histopathologic Study Of Rhabdomyosarcoma cases in a tertiary Care hospital of Peshawar. Pakistan BioMedical Journal, 4(2). https://doi.org/10.54393/pbmj.v4i2.253

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ABSTRACT

Rhabdomyosarcoma (RMS) is the most prevalent soft tissue sarcoma in children and adolescents. **Objective:** To find out RMS's histopathological and immunohistochemical features. **Methods:** The current study was carried out at the Department of Pathology, Section of Histopathology, Khyber Teaching Hospital Peshawar. The study duration was between 1st January 2015 and 30th of December 2020. **Results:** In the current study, all 300 occurrences were found to be relevant. The most prevalent RMS was embryonal rhabdomyosarcoma (ERMS) (87.4%), followed by alveolar RMS (ARMS) (9.4%). In the current study we found that ERMS were most frequent in males (64%). ERMS were 16.1 percent found in the genitourinary system followed by neck and head region (46.7%). Most samples displayed positive Desmin and MyoD1 immunohistochemistry staining. **Conclusions:** It is consistent with previously published RMS data from all across the globe that our patients' epidemiologic information and microscopic findings are consistent with one other.

INTRODUCTION

Rhabdomyosarcoma (RMS) is the most prevalent soft tissue sarcoma in children and adolescents [1]. Children under the age of 10 account for the bulk of instances, with teenagers accounting for fewer than 20% of all cases [1,2]. The most frequent histologic subtype of RMS is ERMS, which accounts for over 60% of all RMS [1]. A primitive, aggressive soft tissue tumor with prenatal contractile cell morphology and physiology is characterized as an ERMS [3]. Botryoid type embryonic RMS is a kind of RMS that has a unique gross (grape-like) and pathologic morphology (aggregates of tumor cells firmly touching an embryonic surface/cambium layer) and accounts for around 6% of all RMS [4]. All other subtypes are more prevalent in males, except for ARMS, which has no gender predilection [1]. Chiles et al. (2004) discovered that white people are far more likely than black people to develop ERMS, whereas ARMS has no racial or regional predilection [5]. The head and neck account for around half of all ERMS cases, with the gastrointestinal tract accounting for the other half, while the extremities account for the majority of ARMS

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cases [6,7]. The prognosis is better for children aged 1 to 9 years; age is an important prognostic factor. As well as the time and place where the disease happens, these are important factors in how well it will go [8]. The goal of this research is to report epidemiologic data (maturity level, ethnicity, and location) as well as diagnostic findings (micron-sized, immunofluorescence) in a broad study of patients identified at Pakistan's biggest histopathology facility during a 6-year period.

METHODS

This retrospective study was carried out at the Department of Pathology, Section of Histopathology, Khyber Teaching Hospital Peshawar. The study duration was between from 1st January 2015 to 30th of December 2020. A total of 300 patients were included in this study. The medical history was reviewed for i) personal data like name, sex, age and residency etc, ii) symptoms and signs, iii) primary site and histopathological characters of tumor, iv) routine lab test at the starting and during treatment such as liver function test, complete blood count, kidney function test, LDH, serum electrolytes and alkaline phosphates, v) imaging studies such as X-ray, ultrasound, MRI and CT on starting and other common metastatic sites, vi) treatment protocols including radiotherapy, surgery, chemotherapy, vii) and patients outcome.

RESULTS

RMS has been found in 300 patients between 2015 and 2020. In the current study, all the diagnosed RMS accounted for a total of 13.8 percent of all soft tissue sarcomas. ERMS was the most common histological type, accounting for 87.4 percent of all the incidences (Table 1). In 26 instances, the botryoid form was discovered, accounting for 10.7 percent of ERMS and 9.4% of all RMS. Males were 155 (64.04%) of the 245 ERMS cases, while females were 87 (35.96%). The male to female ratio was 1.78:1. The preponderance of ERMS in males was found significant statistically ($p<0.006$). Patients with ERMS varied in age from four months to sixty-five years, with an average and standard age of nine and eight years, respectively. Table 2 shows the decade-by-decade breakdown of ERMS cases. For ERMS cases, the botryoid form was found in 26 instances, accounting for 10.7 percent of all RMS cases. Of the 245 ERMS patients, 155 (64.04 percent) were men, whereas 87 were women (35.96 percent). The male-to-female ratio was 1.78:1. ERMS was significantly more common in men than in women (p-value: 0.006). An average and typical ERMS patient was nine years old, ranging from four months to sixty-five years old. Table 2 shows the breakdown of ERMS patients by decade. The most prevalent locations of ERMS were the skull and spine, which accounts for 47.7% of all incidences. More cases (34%) fall within age range of 0-10 years, as shown in Table 3. Most instances of ERMS were found in the skull and spine, which accounted for 47% of all cases. A breakdown of ERMS locations may be seen in Table 4. There is a more precise breakdown of ERMS patients in the urogenital region (Table 4,5). The distribution of ERMS by location. A more detailed breakdown of ERMS patients in the neck, head, and urogenital regions may be seen in Table 5. The extremities were found in the majority of the 26 ARMS patients (65.4%). Table 6 shows the breakdown of the 26 ARMS instances. The extremities were detected in the majority of the 26 ARMS patients (65.4 percent). Table 6 shows the breakdown of the 26 ARMS occurrences.

breakdown of the 26 ARMS occurrences.			
No.	**Histologic Type**	**Number**	**Percentage (%)**
1	Embryonal (including Botryoidivariant)	250	87.4
2	Alveolar	26	9.4
3	Plemorphic	9	2.8
4	Rhabdomyosarcoma NOS	4	1.4

Table 1: Breakdown of Cases According to Histologic Type (n=300)Table 1. Breakdown of Cases According to Histologic Type(n=300)

No.	Age (years)	Number	Percentage (%)
1	0-10	166	65.7
2	11-20 yrs	70	25.6
3	21-30	14	5.8
4	>31	7	2.5

Table 2: Age-wise Distribution of ERMS(n=242)

No	Age (years)	Number	Percentage (%)
1	0-10	9	34.6
2	11-20 years	7	6.9
3	21-30	5	19.2
4	31-40	2	7.7
5	>40	3	11.5

Table 3: Age-wise Distribution of ARMS(n=26)

No	Site	Number	Percentage (%)
1	Head and neck	125	46.7
2	Genitourinary system	42	16.1
3	Abdomen	17	7
4	Pelvis	8	3.3
5	Retroperitoneum	2	0.8
6	Perineum/anal region	9	3.7
7	Trunk	9	3.7
8	Extremities	25	9.9
9	Other sites	10	4.1
10	Site not known	11	4.6

Table 4: Break-down of ERMS Cases According to Location (n=245) *The other sites included: vertebrae/spine (5icases),kidney(2icases), and mediastinum(3 cases).

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No	Location	Number	Percentage(%)
	(A) Head and Neck	118	
1	Nose & Paranasal sinuses	21	18.6
2	Nasopharynx	9	8
3	Orbit/periorbital soft tissue	23	23.3
4	Oral/oropharynx/cheek/palate	20	17.7
5	Ear	6	5.3
6	Tongue	1	0.9
7	Conjunctiva	1	0.9
8	Parotid	1	0.9
9	Jaw	15	12.4
10	Face	2	1.8
11	Skull	5	4.4
12	Neck	5	4.4
13	Sphenoid	4	3.5
14	Parameningeal	1	0.9
	(B)Genitourinary System	40	
1	Urinary bladder	22	56.4
2	Testicular/paratesticular soft tissue	9	23.1
3	Prostate	2	5.1
4	Vagina	5	12.8
5	Cervix	1	2.6

Table 5: Break-down of ERMS Head and Neck and Genitourinary Cases(n=155)

No	Site	Number	Percentage(%)
1	Extremities	19	65.4
2	Head and Neck	5	19.2
3	Neck	2	
4	Trunk	4	11.5
5	Abdomen	2	3.8

Table 6: The breakup of Cases of ARMS According to Location (n=26)

DISCUSSION

RMS is the utmost frequent ligamentous sarcoma in individuals under the age of 15, as well as one of the most prevalent in teenagers and young adults around the globe. Research from Asia and Africa backs this up [9]. It is responsible for around 8 percent of all malignancy tumors in individuals below 15 years of age and 2 percent to 5 percent of all adult sarcomas [6,10]. RMS accounts for 12.9 percent of sarcomas of the soft tissues identified in our department throughout the research period. The vast majority of instances in our analysis (87.4 percent) are accounted for by ERMS. This figure is far higher than the 60% mentioned in the literature [1]. In our research, the botryoid type of ERMS was also more usual. In contrast to previously published data, which showed that roughly 6% of all instances, this study found that 9.4 percent of all instances. Due to the massive plurality of ERMS in our research, the proportion of ARMS was accordingly 19 percent, which is more less than the previous studies stated figure of about 31%. [4]. RMS of epitheloid or Sclerosing, thwhichre uncommon or newly reported individuals [5,11], were not found in our investigation. It's probable that a handful of the ERMS cases described in past years were really a symptomatic variation of RMS, which affects both adults and children and mostly affects the skull, neck, and pretesticular area [5,12]. In the future, we then evaluate all RMS incidences to determine whether there are any sclerosing RMS instances in our dataset. It has yet to be established [11]. In our dataset, the RMS of botryoid accounts for 9.5 percent of all instances, which is somewhat more than the reported series' 6%. [4]. This variation is especially frequent in mucosa-lined hollow organs, including the nasal passages, nasal mucosa, vaginal, ureter, and other mucosa-lined hollow organs, where it develops unrestrictedly, giving it its unique grapes-like look [4]. In this research, men are considerably more likely than women to acquire ERMS (Male: Female Ratio, 1.7:1). Comparable findings may be seen in data from the United States and Canada [13]. About 66 percent of the occurrences happened in the first phase of life, with slightly more than 45 percent occurring in children aged 5 and under. According to Western surveys, the majority of children are under the age of ten, with roughly a third of them under the age of five [14]. ARMS had an equal impact on men and girls. In Western literature as well, there is no gender preference [15]. According to worldwide statistics [16], over 45 percent of ARMS occurred between the ages of 11 and 30, as in this study. The highest incidence occurs between the ages of 10 and 25, with an average age of 6.8 to 9.0 years. Table 4 demonstrates that the neck and head (46.7 percent) were the most prevalent sites for ERMS in our study, trailed by the bladder and kidneys (16.1 percent). According to Western statistics [7], roughly half of all ERMS reported cases are in the trunk area, half in the urinary tract, and in extremities, it is about 9 percent. As shown in Table 5, approximately 10% of ERMS patients in our areas of study in the extremities. The most prevalent ERMS locations in the urinary system. In the renal as well as genital system soft tissues came in second and third, respectively. In our study, the head and neck, as well as the urogenital system, were the most prevalent locations for botryoid RMS patients. According to international statistics [4], these are also the most prevalent places for ERMS. Table 4 reveals that our investigation found multiple instances in the belly, pelvis, pericardium, perineum, and other areas, including two cases in the kidney. According to Western statistics, these are also prevalent ERMS sites. ERMS may also affect the viscera, including the kidney and the heart [17]. Several of our instances included in the heads and necks soft tissues. In Western data, similar traits may be seen [19]. ARMS is most seen in the extremities,

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according to Western research but it can also be found in the head and neck, according to [6,18]. Myo D1 was first utilized in suspected RMS patients in 2015, and it has been used on a regular basis since then. As shown by our findings, it also displayed a high level of sensitivity (over 85 percent). Myogenin, on the other hand, was just recently identified, and this antibody is crucial in the identification of RMS patients [20].

CONCLUSION

Our study concluded that the most prevalent RMS was ERMS which is 87.4%, followed by ARMS which is 9.4%. We also found that ERMS were most frequent in males (64%). ERMS were 16.1 percent found in the genitourinary system followed by neck and head region (46.7%). Most samples displayed positive Desmin and MyoD1 immunohistochemistry staining. It is consistent with previously published RMS data from all across the globe that our patients' epidemiologic information and microscopic findings are consistent with one other.

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