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Rhabdomyosarcoma in Dakar: An Update on Pronostics Factors

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Abstract

Rhabdomyosarcoma (RMS) belongs to the soft tissue sarcomas that have striated muscle differentiation. It is most common in young children under 5 years of age and adolescents. The definite diagnosis of rhabdomyosarcoma is based on histology and the positivity of striated muscle markers in immunohistochemistry. Our objective was to describe the prognostic characteristics of RMS diagnosed in Dakar. MATERIAL AND METHODS: This was a retrospective and descriptive study spread over nine (09) years from 1 January 2011 to 31 December 2019. It was conducted from the histological report archives of the anatomy and pathology laboratories of Dakar. Paraffin blocks were reread and immunohistochemically studied by manual method. Antidesmin and antimyogenin antibodies were used. RESULTS: We collected 44 patients with rhabdomyosarcoma out of 228 cases of soft tissue cancers, *i.e.* 19.29% of all malignant soft tissue tumours. The mean age of the patients was 25.41 ± 23.95 (standard deviation) years with a median age of 16 years and extremes of 6 months and 81 years. Patients were less than or equal to 17 years of age in 61.4% of cases. A clear male predominance was observed with 29 men (65.9%) against 15 women (34.1%). Rhabdomyosarcomas were located in the limbs in 34.1% of cases, in the head and neck in 29.5% of cases and in the genitourinary tract in 25% of cases. The average tumour size was 7.45 cm ± 4.64 (standard deviation) with a minimum of 2 cm and a maximum of 17 cm. It was greater than or equal to 5 cm in 15 patients (68.2%). Embryonal rhabdomyosarcoma (ERMS) was the most frequent histological type with 34 cases (77.3%), followed by pleomorphic rhabdomyosarcoma 7 cases (15.9%) and alveolar rhabdomyosarcoma 3 cases (6.8%). The histological subtypes of ERMS consisted of conventional ERMS (91.18%); botryoid RMS (5.88%) and spindle cell RMS (2.94%). The correlation between histological type and age

was statistically significant (p = 0.039). A relationship was also observed between histological type and site (p = 0.026). According to the American IRS classification, the tumour was classified as group I in 41% of cases, group II in 50% of cases and groups III and IV in 4.5% each. CONCLUSION: Rhabdomyosarcoma is a rare malignant tumour in Dakar. It is often a voluminous tumour of the limbs which affects mainly males. The embryonal type is the most frequent histological form.

Keywords

Rhabdomyosarcoma, Prognosis, Histology, Dakar

1. Introduction

Rhabdomyosarcoma (RMS) belongs to the soft tissue sarcomas that have striated muscle differentiation and can develop in almost any part of the human body, including sites where striated muscle tissue does not normally exist [1].

Rhabdomyosarcoma is most common in young children under 5 years of age and adolescents. It accounts for 60% - 70% of malignant mesenchymal tumours in children and 5% - 8% of childhood cancers [2] [3]. But it is less common in adults accounting for 2% - 5% of all soft tissue sarcomas [4].

The most common sites of rhabdomyosarcoma are the head and neck (40%) including orbital and paraspinal tumours, genitourinary tract (25%), limbs (20%) and trunk (10%) [1].

The signs suggestive of rhabdomyosarcoma depend on the initial location and the involvement of neighbouring organs; hence the importance of imaging in the diagnosis. Ultrasound, CT and MRI play a key role in the detection and characterisation of a mass. These examinations make it possible to recognise the origin of the tumour and to orientate the management and post-treatment follow-up.

The diagnosis of a rhabdomyosarcoma is based on histology and the positivity of muscle markers in immunohistochemistry.

The prognosis of rhabdomyosarcoma is related to the histology, the absence of metastasis at the time of diagnosis, the patient's age, the location, size and operability of the tumour [5] [6].

These prognostic factors are poorly studied in West Africa and particularly in Senegal.

Our objective was to describe the prognostic characteristics of RMS diagnosed in Dakar.

2. Material and Methods

We conducted a retrospective, descriptive, multicentre study over nine years from 1 January 2011 to 31 December 2019.

This study was based on archives of anatomopathological reports of rhabdomyosarcomas from the pathological anatomy laboratories of Aristide Le Dantec

Hospital, Gand Yoff General Hospital, Fann Hospital and Dakar Principal Hospital.

A standardised information sheet was drawn up for data collection. The following parameters were reported: year of diagnosis, patient identification (anatomopathology file number, surname and first names, age, sex, residence), department of origin, site of removal, nature of removal (biopsy or surgical specimen), date of removal, tumour size, histological diagnosis and status of resection margins.

All cases of rhabdomyosarcoma identified were subsequently immunohistochemically confirmed. Each paraffin block was tested using two antibodies: antidesmin and antimyogenin.

The data collected was analysed using Excel 2010 and SPSS 20.0. For each statistical test used, the test was considered significant when p (significance level) was less than 0.05.

3. Results

We collected 44 cases of rhabdomyosarcoma out of 228 soft tissue cancers, *i.e.* 19.29% of all malignant soft tissue tumours.

The mean age of the patients was 25.41 ± 23.95 years (standard deviation) with a median age of 16 years and extremes of 6 months and 81 years. The age of the patients with RMS was less than or equal to 17 years in 61.4% of cases.

A clear male predominance was observed with 29 males against 15 females (sex ratio of 1.93). RMS in the limbs was the most frequent, accounting for 34.1% of cases, followed by RMS in the head and neck, accounting for 29.5% of cases. Genitourinary RMS represented only 25% of cases.

The diagnosis of rhabdomyosarcoma was made on 25 operative parts (56.8%) and 19 biopsies (43.2%).

The average tumour size was 7.45 cm ± 4.64 (standard deviation) with a minimum of 2 cm and a maximum of 17 cm. Fifteen patients (68.2%) had tumours greater than or equal to 5 cm and 7 patients (31.8%) had tumours smaller than 5 cm (**Figure 1**).

Embryonal rhabdomyosarcoma (ERMS) was the most frequent histological type with 34 cases (77.3%) cases, followed by pleomorphic rhabdomyosarcoma (PRMS) 7 cases (15.9%) and alveolar rhabdomyosarcoma (ARMS) 3 cases (6.8%) (**Figure 2**).

The histological subtypes of ERMS consisted of:

- conventional ERMS 91.18%;
- Botryoid ERMS 5.88% and
- spindle cell ERMS 2.94%.

The correlation between histological type and age (**Table 1**) was statistically significant (p = 0.039).

A relationship was also observed between histological type and site (p = 0.026) (**Table 2**).

Table 1. Correlation between histological type and tumeur site.

	Histological type			Total	
	ERMS	ARMS	PRMS		
Limbs	7	2	6	15	P = 0.026
	46.7%	13.3%	40.0%	100.0%	
Genito-urinary	11	0	0	11	
	100.0%	0.0%	0.0%	100.0%	
Head and Neck	12	1	0	13	
	92.3%	7.7%	0.0%	100.0%	
Others	4	0	1	5	
	80.0%	0.0%	20.0%	100.0%	
Total	34	3	7	44	
	77.3%	6.8%	15.9%	100.0%	

Table 2. Correlation between histological type and age.

	Age groups					Total	
	[0 - 8]	[9 - 17]	[18 - 25]	[33 - 40]	>40		
ERMS	11	14	1	3	5	34	P = 0.039
	32.4%	41.2%	2.9%	8.8%	14.7%	100.0%	
ARMS	1	1	0	0	1	3	
	33.3%	33.3%	0.0%	0.0%	33.3%	100.0%	
PRMS	0	0	2	1	4	7	
	0.0%	0.0%	28.6%	14.3%	57.1%	100.0%	
Total	12	15	3	4	10	44	
	27.3%	34.1%	6.8%	9.1%	22.7%	100.0%	

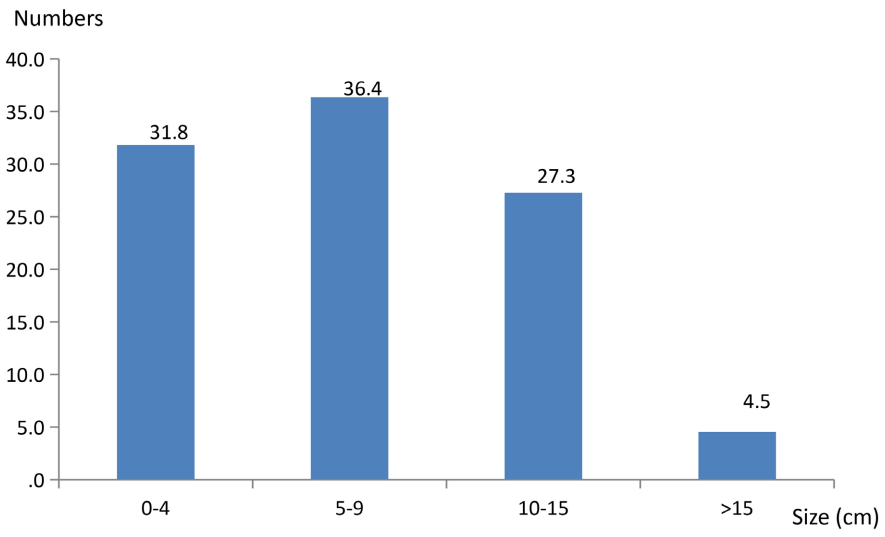

Figure 1. Distribution of rhabdomyosarcomas according to tumour size.

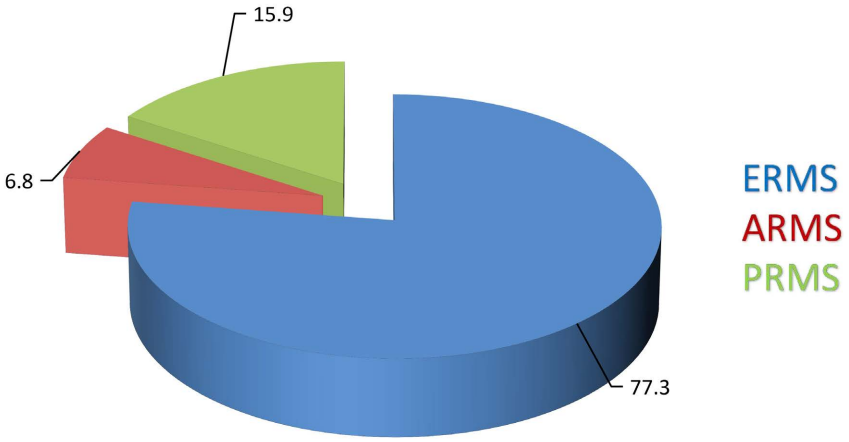

Figure 2. Distribution of rhabdomyosarcoma cases according to histological type.

According to the American IRS classification, the tumour was classified as group I in 41% of cases, group II in 50% of cases and groups III and IV in 4.5% each.

4. Discussion

For all soft tissue sarcomas, RMS accounts for 19% of cases in adults and 45% of cases in children [7].

We collected 44 cases of rhabdomyosarcoma out of 228 registered soft tissue sarcomas, *i.e.* a rate of 19.29%.

A retrospective study conducted in Eastern Egypt by Badr *et al.* [8] over a period of 5 years (2004-2009) recorded 41 cases of rhabdomyosarcoma.

Innocent *et al.* [9] observed 70 cases of rhabdomyosarcoma in Jos (Nigeria) in a 10-year retrospective study (2007-2016). In contrast, Ahmad *et al.* [10] reported 277 cases of rhabdomyosarcoma across Pakistan over 10 years.

The number of RMS cases in this series would be underestimated. Indeed, it is difficult to determine the real frequency of RMS in Senegal as in many African countries.

The factors that justify this fact would be the absence of a register and a reference centre for the diagnosis of RMS where all cases would be sent for review and typing.

A large number of specimens are sent to laboratories outside the country by accompanying persons or through brokering of biological or other laboratories.

The average age of the patients was 25.41 years with a median age of 16 years and extremes of 6 months and 81 years. The majority (61.4%) of patients with RMS were 17 years of age or younger.

Innocent *et al.* [9] in Nigeria, observed a median age of 23 years and extremes of 3 months and 80 years. Ahmad *et al.* [10] in Pakistan reported a mean age of 21 years, a median age of 24 years with extremes of 4 months and 74 years.

Sultan *et al.* [11] reported a median age of 16 years in the United States with 59% of subjects being young.

RMS is more common in younger patients but there are variations between histological types. However, for the same location, younger children have a more favourable prognosis.

The male predominance observed in our series has been confirmed by various authors [4] [11] [12].

The primary site of rhabdomyosarcoma has long been known to be a key prognostic factor. It is also an important element to consider in the therapeutic strategy of RMS, as it conditions the quality of the local procedure with more or less the existence of microscopic or macroscopic residue [13].

Favourable sites are the head and neck (non-parameningeal), genitourinary locations (not bladder or prostate) and biliary tract. Other sites are considered unfavourable [11].

In general, RMS are most frequently located in the head and neck, followed by the genitourinary system and then the limbs [14]. This has been confirmed by several other studies [8] [15] [16].

In the analysis of the SEER database [11] of 1071 adults (aged over 19 years) with RMS, the most frequent primary sites were the extremities (26%) and trunk (23%), followed by the genitourinary tract (17%) and head and neck (9%).

In this series, the location of RMS was most frequent in the limbs (34.1%) followed by RMS located in the head and neck (29.5%), genitourinary location with (25%) and other locations with (11.4%). On the other hand, the favourable sites were slightly more important with 52.27% of cases.

This more important localization of RMS in the limbs in this cohort could be related to the size of the sample which is very small but also to the significant percentage of adults (38.6%).

A tumour size of less than 5 cm is a favourable prognosis factor for rhabdomyosarcoma [17].

In this sample, the mean tumour size was 7.45 cm ± 4.725 (standard deviation). It was less than 5 cm in 31.8% of cases and greater than or equal to 5 cm in 68.2% of cases.

This predominance of tumours of at least 5 cm in diameter was also observed in several studies [4] [16] [18].

Conventional embryonal rhabdomyosarcomas have an intermediate prognosis. Alveolar type tumours and undifferentiated tumours have a worse prognosis [5].

The predominance of embryonal RMS observed in this study has also been found by several other authors [10] [19] [20] [21] [22].

Molecular biology and genetics were not performed in this study in order to differentiate between embryonal RMS and solid alveolar rhabdomyosarcoma in some cases. This could explain the low representation of the alveolar type in this work.

There is a correlation between the histological type of the primary tumour and the age of the patient [23]. This correlation was significant in our study (p =

0.039). The relationship was also observed between histological type and site in this study (p = 0.026) was not found in Ma *et al.* [24] cohort.

The subtypes of embryonal rhabdomyosarcoma consisted of conventional embryonal RMS in 91.18% of cases, botryoid RMS 5.88% and spindle cell RMS 2.94%.

In the study by Ma *et al.* [24], conventional embryonic RMS constituted 94.2%, botryoidal RMS 3.6% and spindle cell RMS 2.2%.

Badr *et al.* [8] reported a percentage of conventional embryonic RMS equal to 84.4%, botryoidal subtype 9.4% and spindle cell RMS 6.2%.

Shouman *et al.* [12] also noted a predominance of conventional embryonic RMS with 90.08%, followed by botryoid RMS with 9.92%.

This preponderance of intermediate and favourable prognosis subtypes in this study was confirmed by several other series.

IRS groups I and II accounted for 91% of the cases in this series. In contrast, Badr *et al.* [8] reported 73.2% of group III and IV. The predominance of low-risk rhabdomyosarcomas in this study may be related to the high percentage of limb locations, thus making their removal easier.

5. Conclusion

Rhabdomyosarcomas are rare in Dakar, affecting both male children and adults. They are large tumours, generally more than 5 cm in length and often located in the limbs. The embryonal type is by far the most frequent with a predominance of IRS groups I and II.

Authors' Contributions

All authors have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare that they have no competing interests.

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