Juvenile Rhabdomyosarcomas in Port Harcourt, Nigeria: A twelve year review

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Summary

Background: **Juvenile rhabdomyosarcoma (JRMS) though rare, is the most common soft tissue malignancy of childhood that exhibits bimodal age distribution pattern.**

Objective: **Review the pattern of JRMS in Port Harcourt based on sex, age, tumor sites and histologic types.**

Design: **A retrospective descriptive study.**

Setting: **The study was conducted in the University of Port Harcourt Teaching Hospital (UPTH), Port Harcourt, Nigeria over 12 years.**

Methodology: **We studied 21 juvenile rhabdomyosarcomas during the period under review. The hematoxylin and eosin stained histological slides were retrieved and reviewed to confirm previous diagnosis and histologically typed for the study. The sex, age and tumor sites were retrieved from the histology consultation forms.**

Result: **These tumors were diagnosed from patients aged 0 – 20 years with the highest frequencies (38.1 % each) occurring at 0 – 5 and 16 – 20 years age group. Males are more affected with a ratio 3.2:1. The trunk is the most common site of occurrence (47.7%) of which the genitourinary system is the most affected (23.8%) in this study. The most common histologic type is the embryonal rhabdomyosarcoma (71.5%). Alveolar rhabdomyosarcoma accounted for (19%) and the sarcoma botryoides (9.5%).**

Conclusion: **The age of presentation and anatomic sites of the tumor are important in the diagnosis of these tumors. If a tumor histologically shows as small round blue cells, rhabdomyosarcoma should be considered as a differential diagnosis.**

Key-words: ***Juvenile rhabdomyosarcoma, Bimodal age distribution, Differential diagnosis.***

Résumé

Introduction: Rhabdomyosarcome juvénile (RMSJ), quoique rare, est une tumeur maligne la plus ordinaire d'enfance qui montre une tendance de la répartition d'âge bimodale.

Objectif: Faire le bilan de RMSJ à Port Harcourt basée sur sexe, âge, siège de la tumeur et types histologiques.

Plan: Une étude descriptive à effet rétroactif.

Cadre: L'étude a été effectuée au centre hospitalier universitaire de Port Harcourt (CHUPH) Port Harcourt, Nigeria au cours d'une durée de 12 ans.

Méthodologie: Nous avons examiné 21 cas de rhabdomyosarcomes pendant la période en cours de révision. L'hémotoxyline et l'éosine des plaques tachées histologiques ont été notées et étudiées afin de confirmer des types diagnostics et histologiques précedents de cette étude. Le sexes, âge, et siéges de la tumeur ont été notés à partir des formulaires histologiques de consultation.

Résultats: Ces tumeurs ont été diagnostiquées chez des patients âgés de 0 – 20 ans avec une fréquence élevée de (38,1% chaqu'un) qui arrive chez le groupe d'âge de 0 – 5 et 16 – 20 ans. Sexe masculin est le plus concerné avec une proportion de 3, 2: 1. Le tronc est le siége le plus ordinaire ou elle se produit fréquemment (47,7%) dont le système génito-urinaire est le plus concerné (23,8%) dans cette étude. Le type histologique le plus ordinaire est le rhabdomyosarcome embryonal (71,5%) alvéole rhabdomyosarcome constitue (19%) et le sarcome botryoides constitue (9,5%).

Conclusion: L'âge de présentation et le siége anotomique de la tumeur sont importants dans le diagnostique des tumeurs si une tumeur indique une petite cellule bleue ronde histologiquement, le rhabdomyosarcome doit être considéré comme un diagnostique différentiel.

Introduction

Rhabdomyosarcomas (RMS) originate from an aberrant embryonal rest muscle cells in tissues.[1] There are four types of this tumor based on histological pattern and prognosis which include: embryonal, sarcoma botryoides, alveolar and the pleomorphic variants[2,3] until recently, the sarcoma botryoides was regarded as a variant of embryonal rhabdomyosarcoma.[4] The paediatric population is commonly affected by the sarcoma botryoides, embryonal and alveolar rhabdomyosarcomas which constituted the juvenile rhabdomyosarcoma (JRMS) while the rarer pleomorphic variant affects the adult population.[5,6,7] In the pediatric age group, juvenile (embryonal and alveolar) rhabdomyosarcoma is the most common mesenchymal malignancy affecting both sexes and have predilection for the head and neck regions and genito-urinary tract.[6] Alveolar and embryonal rhabdomyosarcomas may co-exist.[8] Alveolar rhabdomyosarcoma seen in patients of 30 years of age and above, affects rare anatomical sites; for example intratesticular rhabdomyosarcoma occurs in the elderly while the same tumor in childhood occurs as paratesticular tumor.[9]

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Generally, juvenile rhabdomyosarcoma is considered as one of the causes of childhood proptosis, mass in the middle ear, polypoid lesion of the nasopharynx and nasal cavity.[6] The aim of this paper is to present the cases of juvenile rhabdomyosarcoma seen in our institution.

Materials and methods

A 12-year (1st Jan. 1990 – 31st Dec. 2001) retrospective review of juvenile rhabdomyosarcomas seen in the University of Port Harcourt Teaching hospital (UPTH) the only referral hospital in Rivers State was carried out by the authors. The hematoxylin and eosin stained tissue slides were retrieved and reviewed. Missing or broken slides were recut from the tissue blocks and stained with hematoxylin and eosin stain for the study. Those whose slides or blocks could not be located and those with inadequate documentation were excluded from the study. Variables considered for the study included the age, sex, sites of tumor, and histologic types were extracted from the histology request forms. Special stains like the phosphotongstic acid hematoxylin (PTAH) was used to ascertain proper diagnosis of some of the tumors. The results were analyzed using multiway frequency tables.

Results

A total of 24 cases of rhabdomyosarcomas were diagnosed out of the 2105 cases of malignancies during the study period representing 1.1% of the total malignancies. Out of the 24, three were pleomorphic rhabdomyosarcoma which were found in adults and by definition, excluded from the study. The study is therefore based on 21 rhabdomyosarcomas, which accounted for 1% of the total malignancies.

Table 1 shows the age and sex distribution of juvenile rhabdomyosarcoma patients in Port Harcourt. The age varies from 0 – 20 years with an average age of 11 years. A total of 8(38.1%) cases occurred in the age group 0-5 years involving 5 males and 3 females. In the age group 6-10 years only 3 males (14.3%) were involved; while 2(9.5%) cases involving one male and one female was found in the age group 11-15 years. Eight (31.1%) cases were also found in the age group 16-20 years involving 7 males and a female. On the whole 16 cases were diagnosed in male and 5 cases in females giving a sex ratio of 3.2:1.

Table 1 Age and sex distribution of Juvenile Rhabdomyosarcomas in Port Harcourt

Age group in years	Male	Female	Total	%
0 – 5	5	3	8	38.1
6 – 10	3	–	3	14.3
11 – 15	1	1	2	9.5
16 – 20	7	1	8	38.1
	16	5	21	100.0

Table 2 The anatomical distribution of juvenile rhabdomyosarcoma in UPTH

Anatomical site	Number	Percentage
The extremities		
* Upper Limb	2	9.5
* Lower Limb	1	4.8
The trunk		
* Testicle	1	4.8
* Genitourinary	5	23.8
* Vagina	1	4.8
* Retroperitoneum	3	14.3
Head and Neck		
* Orbit	1	4.8
* Nasal cavity	3	14.3
* Ear	2	9.5
* Neck	2	9.5
	21	100.0

Table 2 shows the site of occurrence of juvenile rhabdomyosarcoma. The extremities, trunk and the head and neck regions were affected. A total of 3(14.3%) cases occurred in the extremities of which 2(9.5%) cases occurred in the deep soft tissues of the upper limbs while only 1(4.8%) case was found in the right lower limb. A total of 9 (42.9%) cases were found in the trunk distributed along the testicle (paratesticular rhabdomyosarcoma 1(4.8%) case, genitourinary 5(23.8%) cases involving only males, vagina of one (4.8%) female, (fig. 1 and fig 2) and retroperitoneum 3(14.3%) cases. The head and neck regions had 9 (42.9%) cases distributed along orbit 1(4.8%) case nasal cavity 3(14.3%) cases, ear and the neck had 2(9.5%) cases each.

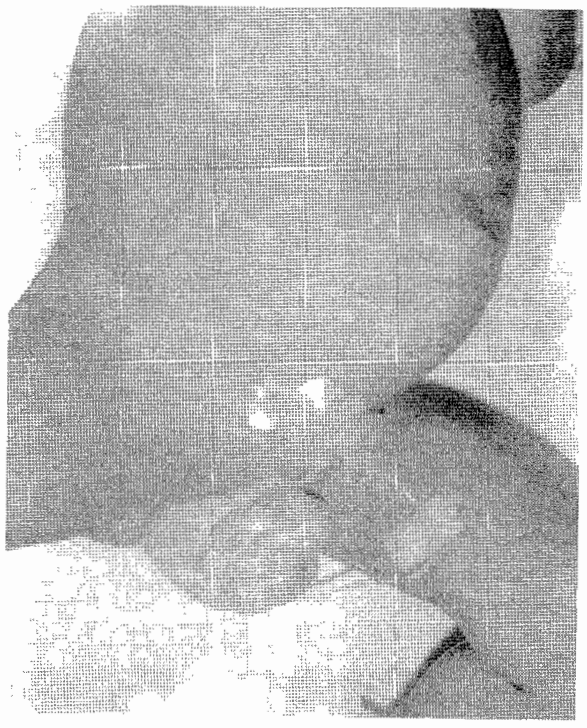

Fig. 1 ***Shows a fungating bunch of tumor in the vagina of a 2½ year old girl.***

Table 3 Frequency distribution of various histologic type juvenile rhabdomyosarcoma in Port Harcourt

Tumor types	Number	Percentage
Embryonal rhabdomyosarcoma	15	71.4
Sarcoma botryoides	2	9.5
Alveolar rhabdomyosarcoma	4	19.0
Total	21	100.0

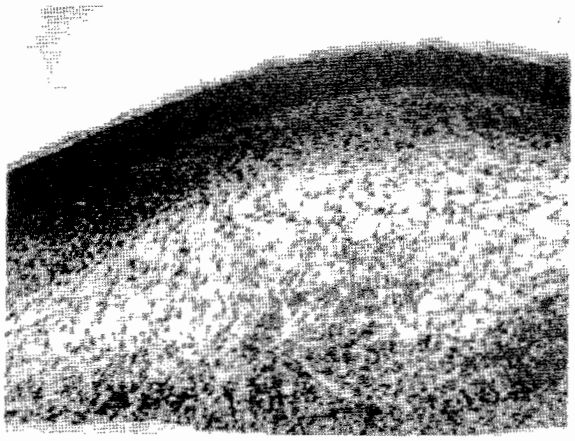

Fig. 2 ***Shows the histological section as small and spindle shaped cells with deeply acidophilic cytoplasm. There are highly cellular areas surrounded by blood vessels which are punctuated in other areas by less cellular regions with inter cellular mucin. These are encased in a dense undifferentiated 'Cabmium' layer.***

Table 3 shows the frequency of various histological types of juvenile rhabdomyosarcoma in UPTH. Embryonal rhabdomyosarcoma accounted for majority of the cases (71.1%), and sarcoma botryoides 2(9.5%) cases. Alveolar rhabdomyo-sarcoma had 4(19.0%) cases.

Discussion

Juvenile rhabdomyosarcomas though rare, are the most common childhood malignant soft tissue tumors in this environment as reported in other studies.[6,7,12,13] It consists of the embryonal rhabdomyosarcoma which histologically present as either classical, aggressive, leiomyomatous or pleomorphic variants in this study corroborating the classification in other study outside this country.[13] The terms are purely descriptive and not associated with the prognosis. This study does not consider sarcoma botryoides as a variant of embryonal rhabdomyosarcoma as claimed by other authors.[4,12] The reason being attributed to their peculiar histological pattern with the 'cambium' layer.[2,3] This reasoning is in keeping with other studies.[2,3] The alveolar rhabdomyosarcoma is the most aggressive and known to have the worst prognosis.[14,13,7]. A minority of cases occur in patients of ages above 20 years but the tumours does not affect rare sites as reported in other study.[9] Juvenile rhabdomyosarcoma exhibits a bimodal age distribution in this study by peaking at the age group 0–5 and 16–20 years. This bimodal age distribution is in keeping with the 3–5 and 16–17 years age group recorded by another report[7] outside this country. These tumors are more frequent in males with a ratio of 3.2:1 in this study, varying with the 2:1 female dominance recorded by study from America.[6] The disparity may be attributed to the sample size and geographical areas of study. The increase in frequency in blacks is corroborated by other studies outside this country.[8] The embryonal rhabdomyosarcoma and the botryoides tumors are more common in the age group 0–5 years whereas the alveolar rhabdomyosarcoma dominate the age group 16–20 years; resembling other Nigerian[14,15] and caucasian[7] studies. The head and neck region is the most common anatomical site for the alveolar rhabdomyosarcoma in this study, similar to other caucasian study[6,7] while the embryonal rhabdomyosarcoma and the sarcoma botryoides are more common in the urogenital system.

The embryonal rhabdomyosarcoma constituted 71.5% of the cases of which the classical variant form the majority (47.6%) as reported in other publications.[6,13] The tumors clinically present as a fungating and ulcerated mass or bunch of grapelike growth in the urogenital, retroperitoneal, vaginal cavity and enlargement of the testicular tissues; whereas in the head and neck region, they present as growth in the nasal cavity causing nasal stuffiness and pains. In the ear, they present as mastoiditis and discharges[8] and in the eyes as proptosis. The alveolar rhabdomyosarcoma of the extremities as well as the head and neck region mainly present as a fungating mass with ulcerations, hemorrhage and necrosis with or without the involvement of the regional lymph nodes.[6,8] This study concludes that juvenile rhabdomyosarcoma is also rare in this institution where the study was conducted.

Acknowledgement

We acknowledge Dr. (Mrs.) G. Eke for the French translation of the summary in this communication.

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