A Fifty-year Review of Soft Tissue Sarcomas in Jamaica: 1958–2007

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ABSTRACT

Objective: *To determine the distribution of histologic subtypes of soft tissue sarcomas (STS) in Kingston and St Andrew, Jamaica, according to age and topography.*

Methods: *From the Jamaica Cancer Registry (JCR) archives, all cases of STS diagnosed between 1958 and 2007 were extracted. For each case, age, gender, histological diagnosis and anatomical site of tumour were recorded. Patients were categorized according to age at diagnosis as: children (0–14 years) and adults (> 14 years), and the distribution of histologic diagnoses with respect to age and anatomical site were analysed.*

Results: *There were 432 cases (67 children, 364 adults, one person of unknown age) of STS recorded in the JCR over the 50-year period (218 males, 214 females). The commonest STS in adults were "sarcoma, not otherwise specified [NOS]" (20.1%), malignant fibrous histiocytoma [MFH] (17.9%), fibrosarcoma (12.4%), liposarcoma (10.7%) and malignant peripheral nerve sheath tumour [MPNST] (10.2%). In children, they were neuroblastoma (38.8%), rhabdomyosarcoma (23.9%), "sarcoma, NOS" (9%), fibrosarcoma (6%) and MFH (6%). In adults, the lower limb was the commonest location, followed by trunk and/or upper limb for MFH, fibrosarcoma and liposarcoma, and head and neck for MPNST. In children, head and neck was the commonest site for rhabdomyosarcoma, head and neck and upper limb for MFH, retroperitoneum for neuroblastoma and trunk for fibrosarcoma.*

Conclusion: *A high proportion of soft tissue sarcomas in Jamaica are unclassified and the anatomical distribution of common classified sarcomas shows some differences with the literature. Limited access to immunohistochemistry/molecular diagnostics and increasing core biopsy diagnosis may contribute to these phenomena.*

Keywords: *Jamaica, sarcoma, soft tissue tumour*

Cincuenta Años de Revisión de los Sarcomas de Tejidos Blandos en Jamaica: 1958–2007

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RESUMEN

Objetivo: *Determinar la distribución de subtipos histológicos de sarcomas de tejido blando (STB) en Kingston y Saint Andrew, Jamaica, según la edad y la topografía.*

Métodos: *De los archivos del Registro de Cáncer de Jamaica, se extrajeron todos los casos de STB diagnosticados entre 1958 y 2007. Para cada uno de los casos, se registró la edad, el género, el diagnóstico histológico, y el sitio anatómico del tumor. Los pacientes fueron clasificados de acuerdo con la edad en el momento del diagnóstico, bajo las categorías de niños (0–14 años) y adultos (> 14 años), y se analizó la distribución de diagnósticos histológicos con respecto a la edad y el sitio anatómico.*

Resultados: *Se registraron 432 casos de STB (67 niños, 364 adultos, una persona de edad desconocida) en el JCR en un período de 50 años (218 varones, 214 hembras). Los STB más comunes en los adultos fueron "el sarcoma no especificado [NE]" (20.1%), el histiocitoma fibroso maligno [HFM] (17.9%), el fibrosarcoma (12.4%), el liposarcoma (10.7%), y el tumor maligno de la vaina del nervio periférico [TMVNP] (10.2%). En los niños, se trató de los neuroblastomas (38.8%), los rabdomiosarcomas*

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(23.9%), "los sarcomas NE" (9%), los fibrosarcomas (6%), y los HFM (6%). En los adultos, los miembros inferiores fueron el lugar más común, seguido del tronco y/o los miembros superiores para el HFM, el fibrosarcoma y el liposarcoma; y la cabeza y el cuello para el TMVNP. En los niños, la cabeza y el cuello fueron el sitio más común para el rabdomiosarcoma; la cabeza, el cuello y los miembros superiores para el HFM; el retroperitoneo para el neuroblastoma; y el tronco para el fibrosarcoma.

Conclusión: *Una proporción alta de sarcomas de tejidos blandos en Jamaica no están clasificados, y la distribución anatómica de sarcomas clasificados comunes muestran algunas diferencias con la literatura. El acceso limitado a los diagnósticos moleculares/inmunohistoquímicos, y el aumento de los diagnósticos centrales, pueden contribuir a estos fenómenos.*

Palabras claves: Jamaica, sarcoma, tumor de tejido blando

West Indian Med J 2012; 61 (7): 693

INTRODUCTION

Soft tissue sarcomas (STS) account for less than 1% of all malignancies (1); among childhood (0–14 years) malignancies, they account for 4–8% (2, 3). They are a heterogeneous group of tumours with diverse cells of origin and different prognostic features and therapeutic options (1), and they may arise from the soft tissue, skin or various viscera.

The incidence of STS varies with histological subtype and age, with pleomorphic undifferentiated sarcoma (malignant fibrous histiocytoma; MFH) and liposarcoma being the commonest in adults (4), and rhabdomyosarcoma (2–4), neuroblastoma and extraskeletal Ewing sarcoma/primitive euroectodermal tumour (EES/PNET) family (4) being the commonest in children.

As STS exhibit diverse histology, incidence, prognostic features and therapeutic regimens (1), the appropriate allocation of health service resources requires current knowledge of the epidemiological profile of these tumours. This paper analyses the demographic and histologic distribution of STS in Kingston and St Andrew, Jamaica, utilizing data from the Jamaica Cancer Registry (JCR). The population base of the JCR is that of the Kingston and St Andrew region of Jamaica.

SUBJECTS AND METHODS

The archives of the JCR were reviewed and all cases of malignant soft tissue tumour diagnosed over the 50-year period from 1958 (when the registry began) to 2007 were extracted. For each case, age, gender, histopathological diagnosis and anatomical distribution (head and neck, upper limb, lower limb, trunk (including pelvis) and retroperitoneum) of tumour were collated. Patients were then categorized according to age at diagnosis (0 – 14 years *vs* > 14 years), into two groups (children and adults, respectively), and we compared the distribution of histopathological tumour type according to age at diagnosis and anatomical site.

In the period under review, tumours recorded in the JCR were coded using varying classifications according to the year of diagnosis, in accordance with World Health Organization (WHO) revisions of the coding systems for diseases, as follows:

1958 – 1963	International List Number (WHO 1957)
1964 – 1972	International Classification of Diseases, version 7 (ICD-7)
1973 – 1977	International Classification of Diseases, version 8 (ICD-8)
1978 – 2007	International Classification of Diseases, version 9 (ICD-9)

For this paper, STS were classified using the histologic classification proposed by Weiss and Goldblum (4), and therefore included malignant tumours of the peripheral nerve and neuroblastomas arising from the sympathetic chain, two groups of tumours not included in the most recent (2002) WHO classification of soft tissue tumours (1). Soft tissue sarcomas not classifiable using either of these classification schemes were labelled "Sarcoma, not otherwise specified (NOS)". When only one or two cases of a particular tumour were found, this tumour was placed in the category "Other". Cutaneous sarcomas and sarcomas arising from viscera, bones and joints were excluded.

RESULTS

There were a total of 432 cases of malignant soft tissue tumours recorded in the JCR archives over the 50-year period: 218 males and 214 females (M:F ratio 1.02:1). The patients ranged in age from less than 1 year to 95 years. There were 67 children and 364 adults; in one patient, the age was unknown (Fig. 1). In adults, the commonest malignant soft tissue tumours were sarcoma, NOS (20.1%), MFH (17.9%), fibrosarcoma (12.4%), liposarcoma (10.7%) and malignant peripheral nerve sheath tumour [MPNST] (10.2%) [Fig. 2A]. In children, the commonest were neuroblastoma (38.8%), rhabdomyosarcoma (23.9%), sarcoma, NOS (9%), fibrosarcoma (6%) and MFH [6%] (Fig. 2B).

Figure 3 shows unclassified sarcomas (sarcoma NOS) expressed as a proportion of the total number of sarcomas, by decade of diagnosis, in adults and children. In adults, there was a progressive decrease in the proportion of sarcomas represented by unclassified tumours, from the earliest decade (30%) to the mid-point of the study period (10.8%). This was followed by progressive increase in the proportional contri-

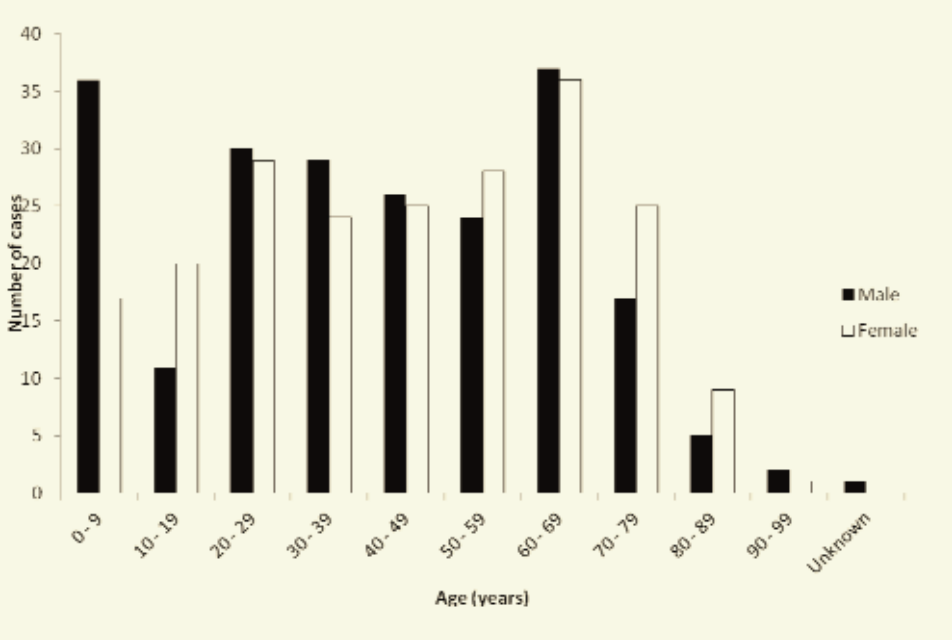

Fig. 1: Age and sex distribution of 432 patients with malignant soft tissue tumours.

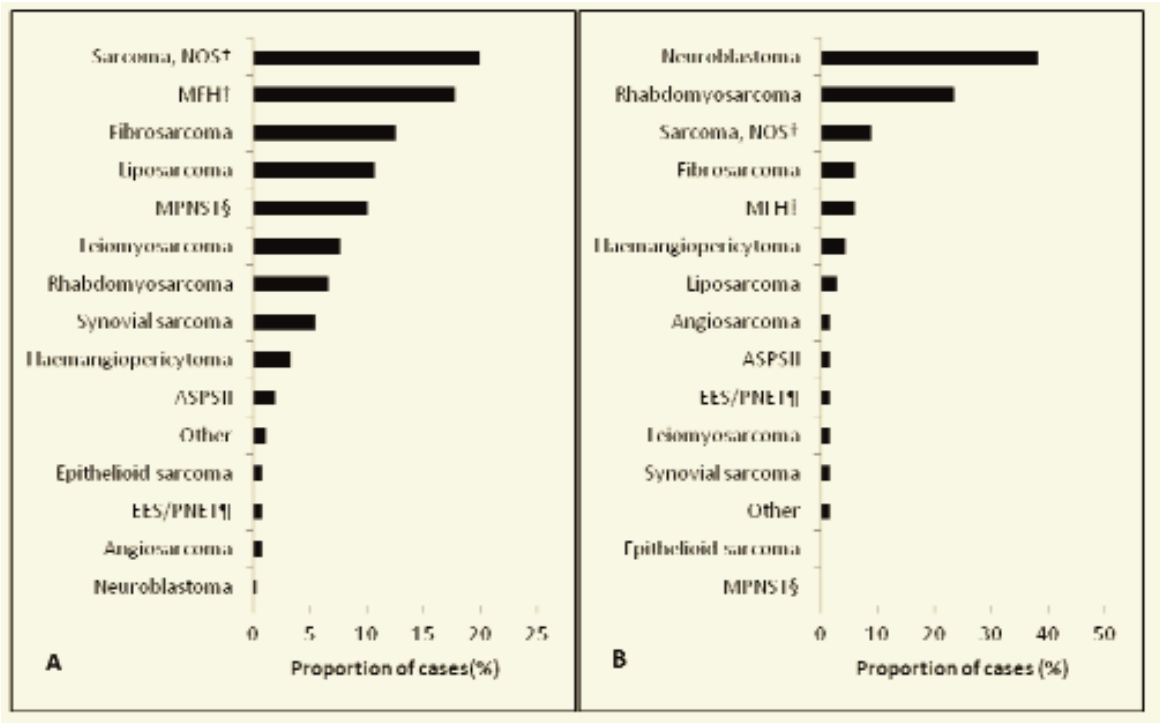

Fig. 2: Distribution of histological types of malignant soft tissue tumours in adults (A) children (B)*
*The age of one patient was unknown; † not otherwise specified; ‡ malignant fibrous histiocytoma; § malignant peripheral nerve sheath tumour; ‖ alveolar soft part sarcoma; ¶ extraskeletal Ewing sarcoma/primitive neuroectodermal tumour; other (adults): mesen-chymal chondrosarcoma (2), fibromyxoid sarcoma (1), haemangio-endothelioma (1); other (children): ganglioneuroblastoma (1)

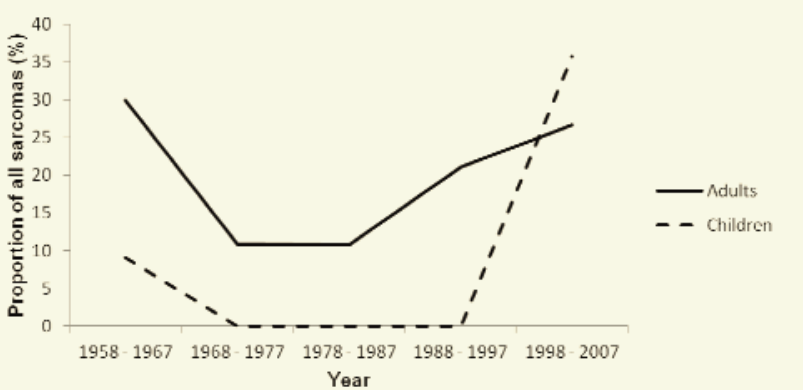

Fig. 3: Unclassified sarcomas (sarcoma, NOS) expressed as a proportion of the total number of sarcomas, by decade of diagnosis, in adults and children.

bution of this subcategory, to a maximum of 26.7% in the last decade of the study. In children, unclassified sarcomas accounted for 9.1% of sarcomas diagnosed in the first decade of the study period. This figure decreased in the mid-years and then increased in the final decade, accounting for 35.7% of all sarcomas.

Figure 4 shows the topographical distribution of the commonest tumours among adults and children, with the exception of sarcoma NOS. Among adults, the lower limb was the most common location for each of the four commonest specific tumours (MFH, fibrosarcoma, liposarcoma and MPNST). The trunk and upper limb were next in ranking among fibrosarcomas and malignant fibrous histiocytoma, while the trunk and retroperitoneum were the next two commonest among liposarcomas. The head and neck region was the second commonest location for malignant peripheral nerve sheath tumours (Fig. 4). In children, neuroblastoma was seen most commonly in the retroperitoneum, followed by the trunk and head and neck, while rhabdomyosarcomas were most common in the head and neck region, followed by the trunk (Fig. 4). Fibrosarcomas in children were distributed among the trunk (50%), retroperitoneum (25%) and lower limb (25%) while MFH in this age group was distributed equally between the head and neck region and upper limb (50% each).

DISCUSSION

It has been reported that the commonest STS seen in adults are pleomorphic undifferentiated sarcomas (MFH) and liposarcomas (4), while the commonest in children are rhabdomyosarcomas (2–4), neuroblastomas and the EES/PNET family of tumours (4). The data from this study are in keeping with these previous reports, with the exceptions of the low incidence of EES/PNET family of tumours in these data, and the high incidence of unclassifiable sarcomas (sarcoma NOS), which, though represented (1.61 – 36%) in many published series (3, 5 – 8), feature among the commonest sarco-mas (13.3 – 36%) in only some (6, 8). The low incidence of EES/PNET seen in our population may be in keeping with the reported low incidence of this family of tumours in other populations of predominantly African descent (9). It is also possible that some less typical, large cell variants of these tumours may have been diagnosed as sarcoma NOS, or un-specified malignant tumours, in the absence of available immunohistochemistry or molecular studies.

The proportionate contribution of sarcoma NOS to STS total was greatest in the earliest and latest periods of the study. The decline in the mid-period may have been partially due to the reliance on histochemical staining and electron microscopy during those years, to separate subtypes of soft tissue sarcomas from each other. At that time, it was believed that these techniques were sufficient to subcategorize STS, and this therefore resulted in the segregation of a high proportion of STS into distinct subcategories. Since that time, knowledge of the molecular biology of these tumours has increased, and it is now recognized that immunohistochemistry and molecular diagnostics are required for accurate subclassification of most STS. Where these techniques are available, a high proportion of tumours will receive specific diagnoses, but where they are not, many tumours will be labelled “sarcoma, NOS”. In addition, whereas extensive sampling of some STS may enable accurate diagnosis on the basis of morphology, the use of small needle core biopsies for diagnosis of these heterogeneous tumours demands the use of these newer ancillary techniques. The increasing trend in sarcoma NOS diagnosis in our setting in the later years of this review is likely due to the limited access to immunohistochemistry and molecular diagnostics in Jamaica and the general increase in use of core needle biopsy for sampling STS (4, 10).

The incidence of STS has been reported to be commoner in males than females (4). The data in this paper are reported in frequencies and proportions only; incidence

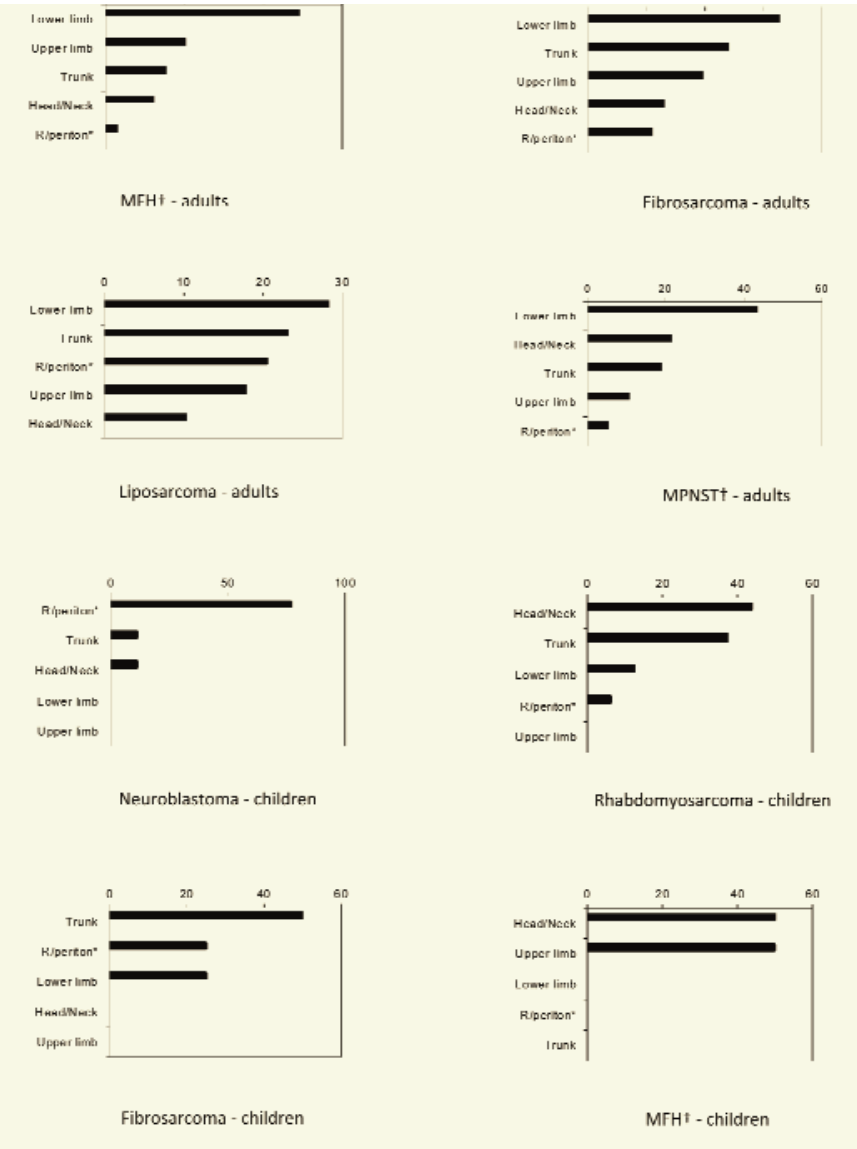

Fig. 4: Anatomical distribution of the commonest malignant soft tissue tumours. *retroperitoneum, † malignant fibrous histiocytoma, ‡ malignant peripheral nerve sheath tumour.

figures were not calculated. However, the last 30 years of data on cancer incidence from the JCR (11–16) show that in our population, the overall incidence (age-standardized rate; ASR) of STS has been consistently higher in males than in females, except for one five-year period (12).

Malignant fibrous histiocytoma, liposarcoma and fibrosarcoma are all reportedly most commonly located within the extremities, particularly the lower limb, with the exception of the dedifferentiated subtype of liposarcoma, which is most commonly found within the retroperitoneum (4). The second commonest location for liposarcoma and MFH is the retroperitoneum, while the trunk is the second commonest for fibrosarcoma (4). Our data for adult patients differ somewhat from these reports, in that the trunk and head and neck are commoner than the retroperitoneum for MFH, and among the liposarcomas, the trunk and retroperitoneum are commoner than the upper limb. Malignant peripheral nerve sheath tumours, reportedly commonest in the extremities, followed by the trunk (4), showed prominent location within the head and neck region in the adult patients (ranked second) in this study, although the lower extremity remained the commonest site, with the trunk ranking third. The topographical distribution of MFH and fibrosarcoma in children in this review is also unlike the distribution reported in the literature, where the extremities predominate (4).

We are unsure of the contribution of dedifferentiated liposarcoma to our liposarcoma total, as the subtype of each liposarcoma was not documented in the registry data. How-

ever, given the low frequency of this subtype of liposarcoma in other series (4), it is probably unlikely to account for a significant proportion of our cases, and is therefore an unlikely explanation for the higher ranking of the retroperitoneum for liposarcomas. Perhaps, the difference in topographical distribution of liposarcoma, fibrosarcoma, MFH and MPNST between our data and others may be the result of misclassification of some of these tumours in the absence of immunohistochemistry. It would be interesting to retrospectively perform immunohistochemical and molecular analyses on these tumours, to see whether changes in diagnoses would be required, and to evaluate the potential benefits of more accessible immunohistochemistry and molecular testing to the appropriate management of patients with STS.

The topographical distribution of neuroblastomas and rhabdomyosarcomas in our data is similar to that reported in the literature (4).

CONCLUSION

The distribution of histological subtypes of STS among adults and children in our population is similar to that reported in the literature, with the exception of the unusually high frequency of sarcoma NOS and the low frequency of EES/PNET in our cases. The anatomic distribution of STS in our data shows some similarities with the distribution reported in the literature, but also exhibits major differences.

We believe that the differences in frequencies and anatomical distribution seen in our data may be partially explained by the limited access to immunohistochemistry and molecular diagnostics in Jamaica. The current practice of providing needle core biopsies for the diagnosis of STS also may contribute to the high frequency of diagnosis of sarcoma NOS. It would be interesting to perform retrospective immunohistochemical and molecular studies on our STS, to see whether there would be changes in diagnoses that would bring our data more in keeping with the findings in other series, and to evaluate the possible benefits of more accessible immunohistochemistry and molecular diagnostics to the appropriate management of our patients.

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