Alexandria Journal of Medicine (2013) **49**, 43–48

Alexandria University Faculty of Medicine

Alexandria Journal of Medicine

www.sciencedirect.com

ORIGINAL ARTICLE

Retrospective review of soft tissue sarcoma of head and neck in a West African hospital

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Received 31 March 2012; accepted 10 September 2012
Available online 10 October 2012

KEYWORDS

Clinicopathology;
Head–neck;
Sarcomas;
Soft tissue;
Tumours;
West Africa

Abstract *Background:* Soft tissue sarcomas like other malignancies, impact negatively on patients and their caregivers as well as pose a challenge to the managing physician with variable treatment outcomes. A review of related studies on Medline has shown the paucity of the literature on the disease in the West African sub-region. This study was designed to determine the prevalence and highlight the clinicopathologic features of soft tissue sarcomas managed at a tertiary health institution in West Africa.

Methods: A 12-year retrospective review of medical records of patients managed with head and neck sarcomas at the University College Hospital, Ibadan, Nigeria was carried out.

Results: There were 18 patients [12 (66.7%) males & 6 (33.3%) females]; ages ranging from 3 to 58 years. 16.7% of patients was children. The mean duration of symptoms was 10.5 months. The clinical presentation depended on the involved anatomical location and the most common anatomical location was the sinonasal region. Fourteen (77.8%) patients presented at advanced disease stage. Eight histologic sub-types were identified and rhabdomyosarcoma accounted for 44.4%. The treatment outcome was poor.

Conclusion: Head and neck sarcomas are rare in West Africa. They have heterogeneous histologic sub-types which may involve different anatomic sites. Although the prognoses of some sarcomas of the head and neck are poor due to their biological behaviours, late stage disease presentation might have contributed to the worse management outcome seen in this study.

1. Introduction

Sarcomas are malignant neoplasms derived from the connective elements of soft or bony tissues in the body. They are rare, accounting for approximately 1% and 6.5% of all head and neck neoplasms in adults and children respectively.[1–3] Soft tissue sarcomas account for the majority (80%) of all sarcomas in the body and constitute about 20% of all sarcomas affecting

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Peer review under responsibility of Alexandria University Faculty of Medicine.

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http://dx.doi.org/10.1016/j.ajme.2012.09.001

the head and neck regions in adults and approximately 35% in children.[4,5] The face and neck are the most frequent sites affected in the head and neck.[6]

Soft tissue sarcomas arise from the mesenchymal derivatives which include muscular, fibrous, fatty, synovial, vascular or neural tissues.[7,8] They exhibit varied biological activities that ranged from slow to aggressive tumour growths, associated with regional and systemic metastases.[9] They tend to grow in the path of least resistance and push the surrounding tissues. The pseudo-capsules of these tumours usually contain invasive tumours and may explain the frequently encountered local recurrence. Distant metastasis is very rare and only occurs in high grade primary lesions.[10,11]

The exact cause of sarcomas of the head and neck is not known but factors such as exposure to chlorophenols in wood preservatives, phenoxy herbicides, vinyl chloride used in the manufacture of plastics and ultraviolet irradiation have been implicated. Infections with human papilloma virus (HPV) and human immunodeficiency virus (HIV) and genetic alteration of the p53 gene (Li-Fraumeni syndrome) have also been suggested.

Establishing the tumour size, histologic type, grade and stage before definitive treatment is also necessary in soft tissue sarcomas because they are important prognostic factors in malignant tumours. Tumour grading is based on Trojani classification which depends on tumour differentiation, mitosis count and tumour necrosis[12] while the staging is by the Union Internationale Contre le Cancer (UICC) or American Joint committee on Cancer (AJCC) classification which is based on tumour size, regional lymph node status, grade and distant metastases.[13,14] Enneking's classification which relies on resection margins or amputation is more appropriately used to stage sarcomas of the extremities.[15] Stages III and IV, where the tumour may be more than 5 cm in size, presence of lymph node or distant metastasis can be regarded as advanced disease stages.

The treatment modalities of sarcomas commonly include surgery, chemotherapy and radiotherapy or a combination of these depending on the histologic type and disease stage.[11]

Head and neck sarcomas impact on the patients' quality of life and also on their family members who provide care for them. It also poses a big challenge to the managing physicians. This is demonstrated more in resource challenged environments where there is scarcity of immunohistochemical studies thereby leading to misdiagnosis and under reported cases. The review of related studies on Medline has shown the paucity of literature on the disease in the West African sub-region. The aim of this study therefore was to report on soft tissue sarcomas of the head and neck in a tertiary health institution in West Africa.

2. Materials and methods

This is a 12-year retrospective study of all patients managed for histologically diagnosed soft tissue sarcomas of the head and neck at the Otorhinolaryngology Department of the University College Hospital, Ibadan, Nigeria from January 2000 to December 2011. This is a leading tertiary health institution in Nigeria that receives cancer patients' referral from primary and secondary health institutions located in different parts of the country due to its medical and radiotherapy facilities.

Data collected from the medical records of the patients and Ibadan cancer registry records included demographic data (age, sex), level of education, occupation, presenting symptoms and their duration, anatomical location of the primary tumour, tumour characteristics (clinical staging and histologic subtypes) and treatment. The socioeconomic status was estimated from the patients' or their parents' level of education and occupation. Five social classes (I, II, III, IV & V) were defined and class I represents the highest socioeconomic class while class V represents the lowest socioeconomic class.[16] Those with incomplete clinical entries or without histological reports were excluded from this study. The clinical photographs of the patients were taken after permission and consent was obtained from them or their care givers at their first hospital visit and stored. The data were analysed using a simple descriptive method and the results were presented in tabular forms.

3. Results

There were 18 patients with histologically diagnosed soft tissue sarcomas of the head and neck. This constituted 77.8% of all the head and neck sarcomas and 1.6% of all head and neck malignancies managed during the study period. There were 12 (66.7%) males and 6 (33.3%) females with a male: female sex ratio of 2:1. The age of the patients ranged from 3 to 58 years with a mean age of 37.3 years and median age of 42 years. Three (16.7%) patients were children with histological diagnosis of embryonal rhabdomyosarcoma.

The duration of symptoms at presentation ranged between 4 and 25 months, with a mean of 10.5 months and median was 9.5 months. Six (33.3%) patients belonged to class V, 9 (50.0%) patients belonged to class IV, 1 (5.6%) patient belonged to class III and 2 (11.1%) patients belonged to socioeconomic class I. The distribution of the patients' age, anatomical locations, histopathologic subtypes, duration of symptoms, socioeconomic class and stage of the disease is shown in Table 1. The most common anatomical location affected was the sinonasal region constituting 33.3% of the soft tissue head and neck sarcomas while ear, larynx and parapharyngeal space had the lowest frequency of involvement (Table 1).

Six (33.3%) patients presented with painful masses. Patients with sinonasal soft tissue sarcomas commonly presented with unilateral epistaxis, facial asymmetry, nasal growth and blockage (Fig. 1a and e).

One patient had an extension of the tumour to the ipsilateral orbit with loss of vision in that eye. The patient with parapharyngeal sarcoma presented with a unilateral progressively increasing cervical mass, dysphagia and features of upper airway obstruction. The patients with oropharyngeal sarcoma presented with tonsillar masses, blood stained saliva, progressive dysphagia, change in voice and cervical lymphadenopathy. The patient with synovial cell sarcoma of the larynx presented with hoarseness and features of upper airway obstruction. The three year old boy with rhabdomyosarcoma of the ear presented with blood stained ear discharge, periauricular swelling and facial palsy (Fig. 1f). There was ulceration of the cervical myxofibrosarcoma and subsequently its rapid, accelerated growth after incisional biopsy was performed at an outside health facility (Fig. 1d). The patient with the facial dermatofibrosarcoma had repeated excisions of the lesion at a peripheral

Table 1 Distribution of the head and neck sarcomas and their anatomical locations.

Age	Sex	Tumour location	Histology	Duration of symptoms (months)	Socioeconomic class	Stage of the disease	Treatment outcome
3	M	Ear	Embryonal rhabdomyosarcoma	8	IV	Stage IV	Dead
4	M	Cervico-facial	Embryonal rhabdomyosarcoma	10	V	Stage III	Dead
11	M	Sinonasal	Embryonal rhabdomyosarcoma	4	I	Stage I	Alive (without evidence of recurrence)
19	M	Larynx	Synovial cell sarcoma	7	IV	Stage III	Dead
22	F	Sinonasal	Alveolar rhabdomyosarcoma	9	V	Stage III	Dead
26	M	Cervico-facial	Alveolar rhabdomyosarcoma	10	IV		Dead
36	F	Sinonasal	Malignant peripheral nerve sheath tumour	17	V	Stage IV	Dead
38	M	Neck	Myxofibrosarcoma	11	IV	Stage III	Dead
41	F	Face	Malignant fibrous histiocytomas	10	IV	Stage III	Dead
43	F	Parapharyngeal	Malignant peripheral nerve sheath tumour	12	IV	Stage III	Dead
47	M	Face	Dermatofibrosarcoma protruberans	25	V	Stage I	Alive (without evidence of recurrence)
51	M	Neck	Botyroid rhabdomyosarcoma	7	III	Stage III	Dead
52	M	Sinonasal	Malignant fibrous histiocytomas	6	I	Stage II	Dead
54	M	Sinonasal	Sinonasal haemangiopericytoma	12	IV	Stage III	Dead
54	F	Oropharynx	Alveolar rhabdomyosarcoma	9	V	Stage IV	Dead
56	M	Sinonasal	Kaposi's sarcoma	13	V	Stage III	Dead
57	M	Neck	Malignant fibrous histiocytomas	11	IV	Stage II	Alive (with recurrence)
58	F	Oropharynx	Alveolar rhabdomyosarcoma	8	IV	Stage II	Alive (with recurrence)

centre before presentation (Fig. 1e). Cervical metastasis was seen in 5 (27.8%) of the patients. Two (0.1%) patients also presented with pulmonary metastasis. Two patients with intracranial extension of sinonasal rhabdomyosarcoma on computerized tomographic (CT) scan did not present with symptoms of raised intracranial pressure or lateralizing sign as the tumors were still confined to extradural space.

The diagnosis of soft tissue sarcomas was by histopathology and immunohistochemical analyses. There were eight histopathologic variants of soft tissue sarcomas found in this study and the most common type was rhabdomyosarcoma constituting 44.4% of the cases, followed by malignant fibrous histiocytomas which formed 16.7% of the cases (Table 1).

Fourteen (77.8%) patients presented at advanced disease stage because the sarcoma was large in size and/or had spread to involve other adjacent structures. Eight (44.4%) patients had some form of tumour resection and neck dissection. However, complete primary tumour resection with free margins could not be achieved due to the large tumour size and involvement of adjacent vital structures. Those with intracranial extension of the tumour had craniotomy and excision of the intracranial component in conjunction with the Neurosurgeons. The patient with orbital extension had orbital exenteration along with total maxillectomy. Only one patient with sinonasal alveolar rhabdomyosarcoma had localized disease that was completely resected with clear margins. All the patients had adjuvant chemotherapy and radiation therapy. The patients with unresectable tumours had palliative chemotherapy and radiotherapy.

The treatment outcomes of these patients were poor. At the time of this study, only 4 (22.2%) of these patients were alive and still being followed-up after two years of diagnosis and treatment (Table 1). However, two of them had already developed local and regional recurrences. The remaining two patients (a 47 year old man with dermatofibrosarcoma protruberans of the face and the 11 year old boy with sinonasal rhabdomyosarcoma) are without clinical and radiological evidence of recurrence till date. The remaining 14 (77.8%) patients died from the disease progression within one year of diagnosis.

4. Discussion

In this present study, soft tissue sarcomas of the head and neck constituted 77.8% of all head and neck sarcomas and 1.6% of all head and neck malignancies managed during the study period. This finding is similar to what had been previously reported in the literature.[1–3] The predominance of males seen in this study is similar to other reports in the literature.[6,11] However, the study by de Bree et al. reported female predominance.[17] This present study showed that soft tissue sarcomas were commoner in young and middle age groups. In similar previous studies of adult head and neck sarcomas, it has been frequently reported in the middle age group.[2,3,6,18] Head and neck sarcomas in children are also reported in this present study with all the patients diagnosed with rhabdomyosarcoma. Rhabdomyosarcoma is the most frequent soft tissue sarcoma in the paediatric age group.[19] The three year old boy with

Figure 1 Clinical photographs of some patients with soft tissue sarcomas.

rhabdomyosarcoma of the ear presented with symptoms mimicking complicated suppurative otitis media. It was after tissue histology from mastoidectomy that the diagnosis was made. Sarcomas should therefore be considered for differential diagnosis in a patient with blood stained mucoid ear discharge, facial nerve palsy and progressive periauricular swelling which does not improve with medical treatment.

Early detection and diagnosis is very essential for good treatment outcome. The average duration at presentation in this study was 10.5 months. In a similar study by Colville from Newcastle, United Kingdom on 60 patients with head and neck sarcomas, the mean duration of symptoms was 8.5 months.[11] This delay usually results in poor clinical and treatment outcome because the disease would have significantly advanced and obtaining a negative resection margin may be difficult without sacrificing a vital organ. In our environment, the delay in presentation to hospitals could be attributed, not only to the nonspecific nature of the commonly encountered head and neck lumps at an early stage[1,2] but also to the socio-cultural beliefs and practices of the people. These factors include self medication, wrong advice from relations and friends to consult traditional herbalists and quacks for treatment as well as traditional & religious beliefs (i.e. that the head and neck sarcoma was due to some form of "spiritual attack"). Therefore, aggressive health education that will correct these beliefs and practices and, will bring about a change in attitude to health is essential. In addition, contemporary radiologic examination tools such as computerized tomographic (CT) scan and magnetic resonance imaging (MRI), which are effective tools for the early detection of these lesions, are not readily available and affordable in most practices in the West African sub-region. It has however been recommended that patients with an unexplained lump in the neck which has recently appeared or undergone rapid growth should be referred to an oncology centre for the evaluation of soft tissue sarcomas.[20] Multidisciplinary team approach management has also been recommended.[11,21]

The general observation of the patients' occupational classes showed that there were more patients in lower occupational classes than those in the upper classes. The higher number of patients from the lower socio-economic class in this study may be a reflection of many people in the lower socioeconomic class in West Africa. Generally, it is believed that people in the upper socio-economic classes are more literate, have healthier lifestyles and behaviour than people in the lower classes.[22] This could also be a reason for this observation in this study.

Although the exact cause of soft tissue sarcomas of the head and neck is not known, few predisposing factors have been reported but none of the patients in this study had an occupation that could expose them to these factors. Previous exposure to radiation is an important predisposing factor especially for malignant fibrous histiocytomas but none of the three patients with malignant fibrous histiocytomas in this study had such a history. Only one patient with Kaposi sarcoma of the sinonasal region had a human immunodeficiency virus (HIV) infection. Kaposi sarcoma is the soft tissue sarcoma that sometimes develops in people with HIV infection.[23] The possibility of infection by Human Papilloma Virus could not be verified in the patients as this is not routinely examined in our centre. Our impression was that the factors that predispose our patients to the disease were yet to be identified. Further studies may be indicated in order to find out these factors in our environment.

The clinical presentations of the patients with soft tissue sarcomas of head and neck depend on the involved anatomic sites and direction of spread. Few of the patients in this study presented with pain which might have been due to the sheer size of the mass and/or the pathological changes like infection or bleeding occurring within the tumour mass.

Soft tissue sarcomas can arise from both the soft tissues and organs of the head and neck. Previous similar studies have reported face and neck as the most frequently affected anatomical sites.[6,11] The most common anatomical location of the tumour in this study was the sinonasal region (Table 1). This

difference might be due to the fact that we were more specific with regard to the anatomical site of tumour origin. The other previous similar studies did not have sinonasal as the anatomic site and it is possible that they have incorporated the sinonasal site as part of the sarcomas of the face since the tumour can extend to involve soft tissue of the face (Fig. 1a). Neck alone or with face (cervicofacial) is the second most frequently involved anatomic site in this study (Table 1). Sarcomas of the head and neck present commonly with local and regional spreads because their pseudo-capsules contain invasive tumour cells.[10,11] This promotes tumour spread in the path of least resistance by eroding or compressing on adjacent structures leading to its rapid development to larger size.

There are many histologic variants of soft tissue sarcomas and the extreme complexity of the histology types requires histopathology and immunohistochemical analyses to distinguish the different subtypes.[24] In this present study, only eight histopathologic variants of soft tissue sarcomas were found. More than 50 subtypes of soft tissue sarcomas have been documented in the literature which showed their histological heterogeneity.[25,26] The fewer histopathologic subtypes of soft tissue sarcomas from this study may be due to the non-availability of immunohistochemical analysis and its expertise at the study centre especially during the early part of the study period. Some of these soft tissue sarcomas might have been missed. The most common histologic sub-type in our study was rhabdomyosarcoma, constituting 44.4% of the cases (Table 1). This frequency of rhabdomyosarcoma in this study is higher than what other similar studies have reported.[1,5,6,8,11,27] This may be due to the inclusion of children in this present study. Rhabdomyosarcoma is the commonest soft tissue sarcoma in the paediatric age group and is associated with distinct genetic alterations.[19] In addition, few of these previous studies incorporated other non-soft tissue sarcomas and this might have contributed to the reduced frequency of rhabdomyosarcoma reported in them. Three histologic sub-types of rhabdomyosarcomas were identified in this study and 50% of which was alveolar rhabdomyosarcoma. The embryonic subtype was found only in the paediatric age group. Historically, rhabdomyosarcomas is known to have poor prognosis because of its relative resistance to radiotherapy. However, systemic chemotherapy, with or without radiotherapy and surgery are recommended as the mainstay of treatment.[28] More often, this is not possible in our setting because patients usually present in advanced disease stage when multiple vital structures in the head and neck region are already affected.

Malignant fibrous histiocytomas is the second most frequent histologic subtype, constituting 16.7% of all sarcomas in this study. Varied frequencies of this tumour have been reported in the literature while a few studies did not report it.[6,8,11,24,27,29] Usually, it arises at a variety of sites in the head and neck region, notably in the salivary glands, soft tissues of the cheek and mouth, the orbit, the meninges and in the sinonasal tract.[29–31] The tumour is usually induced by exposure to radiation. Malignant peripheral nerve sheath tumour is the third most common sarcoma in this study. It is the single most common primary fascicular spindle cell sarcoma in the head and neck region that occurs mainly in adults and shows a predilection for the sinonasal region.[30] Its spread through the lymphatics in the head and neck is rare.[24,32] About 5% of dermatofibrosarcoma protuberans arise in the head and neck region and often affect young to middle-aged adults with a male predominance. Its common locations are scalp and cheek. It is a low grade neoplasm with good response to treatment and has a better prognosis than other sarcomas. The only case of dermatofibrosarcoma protuberans of the face in this study was in a 47 year old male (Fig. 1e). Biologically, dermatofibrosarcoma protuberans exhibit more of a plaque like, diffuse pattern of growth other than the exophytic nodularity in the head and neck. However, its histologic appearances in the head and neck are similar to what was obtained in other parts of the body.[31] It has a locally aggressive behaviour but usually does not metastasise.

Sarcomas originating from the larynx are rarely reported in the literature. Chondrosarcomas are the commonest sarcomas of the larynx.[33] Synovial cell sarcoma of the larynx diagnosed in a 19 year old man in this study is very rare and this makes it not to be readily considered as a differential diagnosis of malignant laryngeal tumour. The other histologic variants seen in this study were as shown in Table 1.

The treatments received by our patients were not different from what had earlier been reported by other similar studies.[2,5,6,27,28] Complete surgical excision of the sarcomas offers the only realistic chance for long term survival. It is however difficult to achieve a negative resection margin in head and neck sarcomas.[18] The modalities of treatment in this study included a combination of tumour resection with radiotherapy and/or chemotherapy in 12 (66.7%) patients and the remaining 6 (33.3%) patients had a combination of radiotherapy and systemic chemotherapy either because they were inoperable due to the proximity of vital structures to the mass or advanced disease stage. In those who had surgical resection of the tumour, a negative margin was possible only in a patient who presented at the early disease stage.

Early presentation, availability of investigative tools and improved therapy will bring about better treatment outcome. The reported prevalence of live, disease free patients with head and neck sarcomas varied from 41.6% to 60%.[6,8,11,17,18,34,35] Irregular and inconsistent irradiation therapy received by the patients due to frequent technical faults developed by the radiotherapy machine and incessant power outage in our hospital also might have contributed to the worse treatment outcome observed in this study.

5. Conclusion

The 12 years retrospective review showed that soft tissue sarcomas of the head and neck are rare in West Africa. They have heterogeneous histologic sub-types which may involve different anatomic locations in the head and neck region. Although the prognoses of some sarcomas of the head and neck are poor due to their biological behaviours, late stage disease presentation might have contributed to the worse management outcome seen in this study.

Acknowledgements

We thank the consultant ENT surgeons whose patients' data were used in this study as well as the medical records officers for their assistance in the retrieval of the patients' medical records.

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