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

Ayotunde James Fasunla *, Adekunle Daniel

Department of Otorhinolaryngology, University College Hospital and College of Medicine, University of Ibadan, Queen Elizabeth Road, PMB 5116, Ibadan, Oyo, Nigeria

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

* Corresponding author. Tel.: +234 8033740220.
E-mail address: ayofasunla@yahoo.com (A.J. Fasunla).

Peer review under responsibility of Alexandria University Faculty of Medicine.

Production and hosting by Elsevier

2090-5068 © 2012 Alexandria University Faculty of Medicine. Production and hosting by Elsevier B.V. All rights reserved.
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