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Soft Tissue Sarcoma of the Head and Neck: Description of 27 Cases and Review of the Literature

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Abstract: To study the histological pattern, age, sex and behaviour of soft tissue sarcomas of head and neck at the University of Ilorin Teaching Hospital, Ilorin and to review existing literatures. The study was a retrospective study of all soft tissue sarcomas arising from the head and neck region, based upon data obtained from the surgical pathology records of the University of Ilorin, Ilorin, from 1985-2006. The neoplasms were classified using WHO classification of soft tissue tumours. Results were analyzed manually. Relative ratio frequencies were calculated and the results were presented in tables. There were 27 cases of soft tissue sarcomas of the head and neck, representing 11.6% of all soft tissue sarcomas. The age range was between 18 months and 80 years with the highest incidence in the first 3 decades of life. Malignant fibrous histiocytoma had the highest incidence of 8 cases (29.6%). One case each (3.7%) of liposarcoma, leiomyosarcoma and Kaposi sarcoma were recorded.

Key words: Soft, tissue, sarcoma, head, neck

INTRODUCTION

Soft tissue sarcomas are malignant neoplasms that arise from the extra-embryonic mesoderm. They are relatively rare, representing only 0.7% of adult malignancies (Wingo *et al*., 1995; Jemal *et al*., 2004; Patel *et al*., 2001). They are more frequent in children, where they represent 6.5% o f all cancers and are the 5th leading cause of cancer death in this age group (Wingo *et al*., 1995). They are, however, the commonest sarcomas (Jemal *et al*., 2004). Soft tissue sarcomas can occur anywhere in the body but most originate in the extremity (59%), the trunk (19%), the retroperitoneum (15%) or head and neck (9%) (De Vita *et al*., 2001). Currently, more than 50 histological types have been identified, but the most common are malignant fibrous histiocytoma (28%), leiomyosarcoma (12%), liposarcoma (15%), synovial sarcoma (10%) and malignant peripheral nerve sheath tumours (6%) (Coindre *et al*., 2001). There are limited number of published individual or institution-based series in the literature on soft tissue sarcomas involving the head and neck region, in black Africans, as most of the available information came from the Western world. The subclassification of sarcoma generally is also subjective and difficulties may exist in differentiating between categories of sarcomas.

The aim of this study is to report our experience with soft tissue sarcomas of head and neck region by describing the pathological characteristics of the 27 cases we have seen in our centre, a tertiary health institution in sub-Saharan Africa. We also undertake a review of the existing literatures on soft tissue sarcoma of the head and neck.

MATERIALS AND METHODS

The study was a retrospective study of all soft tissue sarcomas arising from the head and neck based upon data obtained from the surgical pathology records of the University of Ilorin Teaching Hospital, Ilorin from January, 1985 to December, 2006. The study also included new evaluation of histological specimens.

Clinical information about individual patient was obtained from the Histopathology request forms. The Haematoxylin and Eosin stained slides of individual cases were retrieved from the archives. Where the slides were

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not found, the original blocks were re-cut and re-stained. Where indicated, histochemical stains, such as Mason's trichrome, PTAH and silver stains, were done.

The malignancies were classified using the WHO classification of soft tissue tumours (Fletcher *et al*., 2001). Results were analyzed manually with respect to the age of the patients and histological diagnoses.

RESULTS

During the 22 year study period (January 1985 to December 2006), 232 cases of soft tissue sarcomas were diagnosed in the department, giving an average of 10.5 year^{-1}.

Soft tissue sarcomas of head and neck accounted for 27 cases, representing 11.6% of total cases (Table 1).

The youngest patient with sarcoma in this area was aged 18 months, while the oldest patient was aged 80 years. Table 2 shows the age distribution. The highest incidence was found in the first 3 decades of life. There is no sex difference.

Table 3 shows the histological diagnoses of the cases of the soft tissue sarcoma of the head and neck. Malignant fibrous histiocytoma had the highest incidence of 8 cases (29.6%), including 1 case that was previously diagnosed as undifferentiated sarcoma. Fibrosarcoma and rhabdomyosarcoma have 7 cases each, representing 25.9% of cases. Only 1 (3.7%) case each of liposarcoma, leiomyosarcoma and Kaposi sarcoma was recorded. There was 1 case (3.7%) that could not be further characterized and was labelled undifferentiated sarcoma.

Table 1: Site of tumour

Site of tumour	Frequency	Percentage
Lower limb	80	34.5
Trunk	61	26.3
Intra-abdominal	40	17.2
Head and neck	27	11.6
Upper limb	19	8.2
Retroperitoneal	5	2.2
Total	232	100.0

Table 2: Age distribution

Age range	Frequency	Percentage
0-9	6	22.2
10-19	6	22.2
20-29	6	22.2
30-39	2	7.4
40-49	2	7.4
50-59	0	0.0
60-69	3	11.1
70 and above	2	7.4
Total	27	100.0

Table 3: Histological subtype of sarcoma of head and neck

Histological type	Frequency	Percentage
Malignant fibrous histiocytoma	8	29.6
Fibrosarcoma	7	25.9
Rhabdomyosarcoma	7	25.9
Liposarcoma	2	7.4
Leiomyosarcoma	1	3.7
Kaposi sarcoma	1	3.7
Undifferentated sarcoma	1	3.7
Total	27	100.0

DISCUSSION

Soft tissue sarcomas are rare malignant tumours that arise from the extra-skeletal embryonic mesoderm (Wingo *et al*., 1995). There is no identifiable aetiology, but a variety of predisposing or associated factors are identified (Li and Fraumeni, 1969; Martland, 1931). There is no clear genetic predisposition, except in the Li and Fraumeni (1969) and hereditary retinoblastoma. The link between radiation and sarcoma has been recognized since the 1930s (Martland, 1931). External radiation therapy for cancer of the breast, cervix, ovary, testes or lymphatic system is a well-established risk factor for soft tissue sarcoma (Brady *et al*., 1992). The risk is commensurate with the dose of radiation, the median latency period being approximately 10 years (Brady *et al*., 1992). Other risk factors include occupational exposure to certain chemicals, including herbicides like phenoxyacetic acids and wood preservatives, containing chlorophenols (Hardell and Sandstrom, 1976; Smith *et al*., 1984).

Sarcomas are often believed to be exclusively mesenchymal in origin, but some histological subtypes, like synovial sarcoma and epithelioid sarcoma, share some epithelial features, raising the possibility they may be derived from epithelial tissue containing the cytokeratin type of intermediate filament (Leyvraz and Costa, 1988). Sarcomas rarely develop from pre-existing benign soft tissue tumours (Huvos, 1985).

In a large review, Maurer *et al*. (1988) reported the extremities (60%) as the commonest site of soft tissue sarcoma while the head and neck region accounted for 9% of cases. This is similar to the 11.6% in this study. In a 21 year review, Ou (1988) reported 87 cases of soft tissue sarcomas of the head and neck region. Fibrosarcoma was the commonest histological variant (36.8%), while hemangiosarcoma was the least common (5.8%). In our study, the commonest histological variant is malignant fibrous histiocytoma with 8 cases (29.6%) followed by fibrosarcoma and rhabdomyosarcoma with 7 cases each (25.9%). Rhabdomyosarcoma was the commonest histological variant in the Maurer *et al*. (1988) study, accounting for 37% of sarcomas of head and neck. Ou (1988) did not record any case of liposarcoma or synovial sarcoma. One case of liposarcoma (3.7%) was recorded in our study. The most frequently involved sites in the Ou (1988) review is near the nasal cavity and the

maxillary sinus for fibrosarcoma (66%), base of the tongue for hemangiosarcoma (60%), scalp for dermatofibrosarcoma protuberans (54%) and the parapharyngeal space and soft tissue of the neck for neurogenic sarcoma (45%). Subclassification on the basis of site involved was not done in our study.

The youngest patient in this study was an 18 month old child with embryonal rhabdomyosarcoma, while the oldest was an 80 year old female with fibrosarcoma. Freedman *et al.* (1986) reported age at diagnosis of this tumour of between 6 weeks and 91 years.

Assessment of tissue type may be difficult while making a diagnosis and immunohistochemistry and even electron microscopy may be needed in some cases. (Van Haelst, 1986; Hibshoosh and Lattes, 1997). The difficulty of histological typing increases as the degree of differentiation lessens (Mann *et al.*, 1999). In a large study (Preasant *et al.*, 1986), a 20% incidence of major pathological errors was recorded from referring institutions. It is important that biopsies are assessed by experienced pathologists.

Different grading system have been used and compared, but there is no consensus (Guillou *et al.*, 1997). Major pathological factors in assessment of grade are the mitotic count and the presence of necrosis with tumour differentiation playing a role (Guillou *et al.*, 1997; Russell *et al.*, 1977). From a clinical point of view, the grade rather than the precise tissue histogenesis is critical for tumour behaviour and prognosis (Preasant *et al.*, 1986). Mutations of p53 have been associated with specific tumour subtypes, high histological grade and a poor prognosis (Kawai *et al.*, 1994).

The dominant pattern of metastasis is haematogenous. Lymph node metastasis is rare (>5%), except for a few histological subtypes such as epithelioid sarcoma, synovial sarcoma, rhabdomyosarcoma, clear cell sarcoma and angiosarcoma (Fong *et al.*, 1993).

We recorded a case of Kaposi sarcoma of the upper eyelid in a woman with HIV/AIDS. It represents the only case we have seen in this centre to date, involving that region of the body.

Upon re-evaluation of histological specimens, one previous diagnosis of undifferentiated sarcoma was re-diagnosed malignant fibrous histiocytoma. This may make any previous review faulty by present standards.

CONCLUSION

In conclusion, soft tissue sarcoma shows a gradual increase, even though, it is said to be rare with involvement of the head and neck region even rarer. Histological typing is difficult and in a developing country like Nigeria, facilities for and expertise in histochemical diagnostic techniques is limited. There is need for a broader study of the peculiarities of the soft tissue sarcomas of the head and neck region.

REFERENCES

Brady, M.S., J.J. Gaynor and M.F. Brennan, 1992. Radiation associated sarcoma of bone and soft tissue. Arch. Surg., 127: 1379.

Coindre, J.M., P. Terrier and L. Guillou *et al.*, 2001. Predictive value of grade for metastasis development in the main histologic types of adult soft tissue sarcomas: A study of 1240 patients from the French federation of cancer centers sarcoma group. Cancer. 91: 1914-1926.

De Vita, V.T., S. Hellman and S.A. Rosenberg, 2001. Cancer: Principles and Practice of Oncology. 6th Edn. Philadelphia, PA: Lippincott Williams and Wilkins, pp: 1841-1891.

Fletcher, C.D.M., K.K. Unni, F. Mertens and World Health Organization (WHO), 2001. International classification of tumours. Pathology and Genetics. Tumours of the Soft Tissues. Lyon.

Fong, Y., D.G. Coit, T.J. Woodruff and M.F. Brennan, 1993. Lymph node metastasis from soft tissue sarcoma in adults. Analysis of data from a prospective database of 1772 sarcoma patients. Annal. Surg., 217: 72-77.

Freedman, A.M., H.M. Reiman and J.E. Wood, 1986. Soft tissue sarcomas of the head and neck. Am. J. Surg., 158 (4): 367-372.

Guillou, L.G., J.M. Coindre and F. Bonichon *et al.*, 1997. Comparative study of the National Cancer Institute and French Federation of Cancer Centers sarcoma Group Grading Systems in a population of 410 adult patients with soft tissue sarcoma. J. Clin. Oncol., 15: 350-362.

Hardell, L. and A. Sandstrom, 1976. A case control study: Soft tissue sarcoma and exposure to phenoxyacetic acids or chlorophenols. Br. J. Cancer, 39: 711-717.

Hibshoosh, H. and R. Lattes, 1997. Immunohistochemical and molecular genetic approaches to soft tissue tumor diagnosis: A primer. Semin. Oncol., 24: 515-525.

Huvos, A.G., 1985. The spontaneous transformation of benign into malignant soft tissue tumours. A. J. Surg. Pathol., 9 (Suppl.): 7-20.

Jemal, A., R.C. Tiwari and T. Murray *et al.*, 2004. Cancer Statistics. CA Cancer J. Clin., 54: 8-29.

Kawai, A., M. Nogushi and Y. Beppu *et al.*, 1994. Nuclear immunoreaction of p53 protein in soft tissue sarcomas: A possible prognostic factor. Cancer, 73: 2499-2505.

Leyvraz, S. and J. Costa, 1988. Histological diagnosis and grading of soft tissue sarcoma. Semin. Surg. Oncol., 4: 3-6.

Li, F.P. and J.F. Fraumeni, 1969. Soft tissue sarcoma, breast cancer and other neoplasms: A familial syndrome? Ann. Int. Med., 71: 747-752.

Mann, G.B., J.J. Lewis and M.F. Brennan, 1999. Adult soft tissue sarcoma. Aust. N. Z. J. Surg., 69: 336-343.

Martland, H.S., 1931. The occurrence of malignancy in radioactive persons. A general review of data gathered in the study of radium dial painters with special reference to the occurrence of osteogenic sarcoma and interrelationship of certain blood diseases. Am. J. Cancer., 15: 2435-2516.

Maurer, H.M., M. Beltangady and E.A. Gehan *et al*., 1988. The intergroup rhabdomyosarcoma study 1. A Final Report. Cancer, 61: 209-220.

Ou, S.M., 1988. Soft tissue sarcoma in the head and neck-analysis of 87 patients. Zhonghua Zhong Liu Za Zhi, 10 (4): 289-292.

Patel, S.G., A.R. Shaha and J.P. Shah, 2001. Soft tissue sarcomas of the head and neck: An update. Am. J. Otolaryngol., 22: 2-18.

Preasant, C.A., W.O. Russell, R.W. Alexander and Y.S. Fu, 1986. Soft tissue and bone sarcoma histopathology peer review: The frequency of disagreement in diagnosis and the need for second opinions: The SouthEastern Cancer Study Group experience. J. Clin. Oncol., 4: 1658-1661.

Russell, W.O., J. Cohen and F. Enzinger *et al*., 1977. A clinical and pathological staging system for soft tissue sarcomas. Cancer, 4: 1562-1570.

Smith, A.H., N.E. Pearce and D.O. Fisher, 1984. Soft tissue sarcoma and exposure to herbicides and chlorophenols in New Zealand. J. Nat. Cancer Inst., 73: 1111.

Van Haelst, U.J.G.M., 1986. Electron Microscopy in the Study of Soft Tissue Tumours: Diagnosis/Differential Diagnosis and Histogenesis. In: Van Oostero, A.T. and J.A.M. Van Unnik (Eds.). Management of Soft Tissue and Bone Sarcomas. New York: Raven, pp: 77-91.

Wingo, P.A., T. Tong and S. Bolden, 1995. Cancer statistics. CA Cancer J. Clin., 44: 8-30.