Sarcomas of head and neck – A 10 yrs experience

Abhishek Srivastava · Anirban Ghosh · Somnath Saha · V.P. Saha · Debdulal Chakraborty

Abstract

38 cases of sarcoma of head and neck region were analysed in a retrospective way in relation to age, anatomic location, histological, clinical profile, and surgical approaches. Compared to other types of head and neck neoplasms, such as squamous cell carcinoma, soft tissue sarcomas have low rates of regional metastases. However the biological behaviour of soft tissue sarcoma is more aggressive specially in paediatric age group. In the present series, CT scan was considered as the primary modality of investigation. Surgery generally has been recommended as the primary method of treatment for achieving local control, except in those high-grade tumours arising in sites not amenable to resection. 3-year and 5-year survival rates in this present series 50% and 31.6% respectively.

Keywords Soft tissue sarcoma · CT Scan · Excisional Biopsy · Chemoradiation · Survival rate · Prognostic factors

Introduction

Sarcomas are rare neoplasms, which account for only 1% of all head and neck primary cancers. However in the pediatric population 35% of all the sarcomas affect head and neck sites. The rarity of the sarcomas and the spectrum of the histologic subtypes make the definitive diagnosis very difficult. The biology of a neoplasm and its propensity for locally aggressive growth and systemic dissemination are clearly reflected in its histology and grade. The size and location also impart reliable prognostic information. Except the pediatric rhabdomyosarcoma (RMS), soft tissue sarcomas only rarely arise in the head and neck region. Adjuvant radiotherapy is generally recommended for high-grade sarcomas, large tumours, close or positive surgical margins and histological variants. Systemic chemotherapy is recommended for those tumours with a significant risk of distant metastases. Traditional predictors of treatment failure for soft tissue sarcomas include larger tumour size, high-grade histology, and positive surgical margins. The advent of more advanced reconstructive techniques, including free tissue transfer, has made more aggressive surgical resection of these tumors possible.

Materials and methods

This was a retrospective study done in Dept of Otolaryngology from 1996 to 2005. A total number of 38 cases of sarcomas of head and neck region that presented to the hospital were taken up in this study. All the patients in the present study underwent detailed history-taking and clinical examination including ophthalmological work-up. Otolaryngological work-up included clinical and endoscopic examination of nose, nasopharynx and paranasal sinuses. The neck was examined in all the cases. All patients in the present series underwent radiological investigation and CT scan to evaluate the extent of tumour and to localize the lesion. A few cases were evaluated with MRI to know the detail soft tissue delineation. The lesion was approached as per their anatomical location.

A. Srivastava¹ · A. Ghosh² · S. N. Saha³ (✉) · V. P. Saha⁴ · D. Chakraborty⁵

¹Resident

²Resident

³Assistant Professor

⁴RMo Cum Clinical Tutor

⁵RMo Cum Clinical Tutor

Department of ENT,
Medical College and Hospital,
Calcutta - 700 073,
India.

Sundaram Apartments

Barat, Lake Town,

Calcutta - 700 089

E-mail: chittimonda_saha@yahoo.com

Ph: +91 / 33 / 25740508

Results

All patients were analysed in regard to age, sex, anatomical location, histopathological type and surgical approaches employed for their removal.

Age and sex distribution (Diagram 1)

Except for paediatric Rhabdomyosarcoma, soft tissue sarcomas are rare in head and neck region. In this study all paediatric cases (total 17) are due to paediatric Rhabdomyosarcoma. Apart from that, most cases (30.6%) belong to the 40–59 yrs age group. Head and neck soft tissue sarcomas are twice as common in male as compared to female.

Site of origin (Diagram 2)

Anterior skull base and sino-nasal tract appears to be the most common site of affection (58%) in this series. Soft tissue sarcoma involves lateral skull base and ear in about 26% followed by parotid and neck region.

Histopathology & outcome of treatment (Table 1)

Diagram 1 Age distribution of sarcoms of head and neck region

Diagram 2 Site of origin of sarcoms of head and neck region

Histopathological diagnoses of the excised masses are described in Table I for different types of sarcomas of head and neck region. 3-year and 5-year survival rates are also analysed in this table.

Treatment options (Table 2)

Different types of surgical approaches for sarcomas of head and neck are discussed in Table 2.

Discussion

Malignant tumours that develop in soft tissue are called sarcomas, a term that comes from a Greek word meaning “fleshy growth.” Different types of soft tissue sarcomas are grouped together because they share certain common microscopic characteristics, produce similar symptoms, and are generally treated in similar ways. Osteosarcomas are grouped in separate category because they have different clinical and microscopic characteristics and are treated differently. Sarcomas can invade surrounding tissue and can metastasize to other organs of the body. These ‘Secondary tumours’ are referred to as “metastatic soft tissue sarcoma” because they are part of the same cancer and are not a new disease.

In most of studies, head and neck primary sarcomas represent only 5% to 15% all sarcomas manifest in adults. In pediatric population, however, 35% of all sarcomas manifest in head and neck [1]. Sarcomas are classified according to various criteria that are also useful in determining prognosis and formulating treatment strategies. Sarcomas can arise from soft tissue, bony elements or other mesenchymal cells. Among all sarcomas of head and neck region 80% are soft tissue type; only 20% are of bony or cartilaginous origin [2]. Some types have no clear association with

Fig. 1 Clinical photograph showing mass in the right side of cheek diagnosed as rhabdomyosarcoma of maxillary antrum

Table 1 Histological diagnoses

Histological diagnoses	Number	Percentage	3- yr survival	5-yr survival
Osteosarcoma	2	5.26	100	In follow up
Chondrosarcoma	5	13.16	100	60
Malignant fibrous histiocytoma	2	5.26	100	50
Fibrosarcoma	1	2.63	Nil	Nil
Rhabdomyosarcoma	18	47.37	17	Nil
Angiosarcoma	2	5.26	100	100
Haemangiopericytoma	4	10.53	100	75
Neurogenic Sarcoma	1	2.63	Nil	Nil
Synovial Sarcoma	1	2.63	Nil	Nil
Ewing's Sarcoma	1	2.63	100	100
Undifferentiated Sarcoma	1	2.63	Nil	Nil

Table 2 Treatment options

Treatment modalities	Number	Percentage
Biopsy and chemoradiation	12	31.58
Medial maxillectomy	7	18.42
Maxillectomy and its extension	8	21.05
Cranio facial resection	2	5.26
Infratemporal fossa approach	1	2.63
Hemimandibulectomy followed by chemoradiation	1	2.63
Subtotal petrosectomy followed by chemoradiation	1	2.63
Radical mastoidectomy followed by chemoradiation	6	15.79

any particular tissue and thus creating mystery about their origin. Osteosarcoma, Malignant Fibrous Histiocytoma, RMS, Angiosarcomas, Ewing sarcoma and Alveolar soft part sarcoma (ASPS) are considered to be high grade lesions [2, 3]. Dermatofibrosarcoma protuberans, desmoid

tumours and atypical lipomatous tumours are almost universally low-grade neoplasms [4]. Other types of sarcomas, including chondrosarcoma, fibrosarcoma, leiomyosarcoma, liposarcoma, neurogenic sarcoma and hemangiopericytoma require individualized grading system according to the pathological analysis. Classification of sarcomas according to anatomic location in the head and neck region has proved to be helpful because of the influence of location on disease management decisions.

Several genetic and environmental disorders are clearly related to the development of sarcomas. Li-Fraumeni syndrome is an autosomal dominant disorder involving the germline mutation of the p53 tumour suppressor gene [5]. Other conditions like neurofibromatosis, Carney triad, hereditary hemochromatosis, Werner syndrome, basal cell carcinoma syndrome etc are associated with increased risk of sarcomas. Several environmental exposures have been proposed as causative agents in the development of sarcomas like ionizing external – beam radiation, radioisotopes, chemicals like phenoxyacetic acid – based herbicides, agent orange and dioxin have been implicated for the soft tissue sarcomas. Polyvinyl chloride, thorium oxide, and inorganic arsenic are

Fig. 2 CT scan (coronal cut) showing heterogenous mass in right maxillary antrum and nasal cavity diagnosed as primitive neuroectodermal tumour

Fig. 3 CT scan (coronal cut) showing mass in the left anterior and posterior ethmoidal cells extending into the skull base diagnosed as rhabdomyosarcoma

well-documented risks for angiosarcomas. Tobacco, parenteral drug use particularly cocaine and marijuana increases the risk of pediatric RMS as much as five fold [7].

Most of the sarcomas of head and neck region presents with non-specific signs and symptoms. In 80% of patients, these tumours manifest as a painless mass. A variety of symptoms can occur, however, depending on the subsite of the head and neck affected. Tumors of neck can impinge on the vital structures, causing dysphagia, hoarseness and even dyspnoea. Examination generally reveals a submucosal mass of the upper aerodigestive or sinonasal tract or a subcutaneous mass of neck possibly with distortion or destruction of the adjacent structure.

Imaging studies augment the physical examination by more accurately assessing the size and location of sarcomas in the head and neck. High resolution of CT scans and MRI are the studies in nearly all cases. CT scanning is rapid and presents fewer problems with motion artifacts. Increased sensitivity for bony abnormalities with CT compared with MRI is another advantage to this modality. In addition CT is less expensive. MRI however offers much better soft tissue resolution and is therefore better able to evaluate the primary lesion, perineural extension, dural involvement, and bone marrow replacement and orbital invasion. Multiplanar imaging also assists in more accurate views of the tumour and adjacent structures. Other advantages of MRI are lack of exposure to radiation and iodinated contrast material. Therefore CT and MRI are used in complimentary fashion especially for surgical planning.

The biopsy should be performed, if possible, after imaging is completed. An initial biopsy can be done with fine needle aspiration, although occasionally core-needle specimens may be necessary to accurately subtype the tumour. In sarcomas of head and neck region, a transmucosal biopsy is appropriate for lesions in the sinonasal and aerodigestive tracts. Careful open biopsies should be considered where transmucosal biopsy is not feasible. Complete surgical resection of sarcomas in the parotid, neck, and parapharyngeal space is preferable to incisional biopsy if the FNA is nondiagnostic. Special immunohistochemical stains and cytogenetic studies can greatly assist in confirming the diagnosis of sarcomas. After the diagnosis is confirmed, distant metastasis should be excluded [8]. Sarcomas most commonly spread to lungs. Consequently, imaging of chest is necessary. Findings such as elevated liver function tests, abnormal bone profiles, neurologic deficits, and bone pain warrant further evaluation with appropriate studies.

Treatment options of sarcoma include surgery, chemotherapy and radiation. Various regimens of chemotherapy and radiotherapy have been studied but their role still remains controversial. In most recent experiences radiation delivered by EBRT (External Beam Radiation Therapy) and brachytherapy improve the overall survival rates in osteosarcomas of head and neck region with positive margins or high grade malignancy [9].

For chondrosarcoma, wide surgical excision with negative margins is necessary, especially in high-grade tumors [10].

Malignant fibrous histiocytoma and Fibrosarcoma are best treated with surgical excisions with negative margins. However positive margins and high-grade malignancies do respond to adjuvant radiotherapy [11]. Rhabdomyosarcoma is first staged according to the IRSG staging system before deciding the treatment modality. Complete surgical resection remains the critical component of the treatment. When comprehensive resection is not possible then chemotherapy and radiotherapy can be tried [12]. Vascular tumours like Vascular Angiosarcoma and Haemangiopericytoma are treated with surgery combined with the radiotherapy. Intralesional injections of cytokines in conjunction with surface radiation do have a role, newer regimes of chemotherapy is being tried [13]. Tumours of neurogenic origin like Neurogenic sarcoma can be treated with wide local excision, chemotherapy and radiotherapy have no definitive role. There is no indication neck dissection, as this tumour does not spread to the cervical lymph nodes [14]. Synovial sarcoma can be treated with surgical excisions with negative margins; postoperative chemo-radiation can be given for high metastatic rates of this tumour. Optimal treatment is surgery in case of Ewing's sarcoma followed by chemotherapy with VAC (Vincristine, Adriamycin, Cyclophosphamide), and adjuvant radiotherapy can be added.

In this study various surgical approaches had been used depending on specific locations and extension of the tumour. Most of the RMS of paediatric age group presented with extensive spread with skull base involvement and treatment option narrowed to biopsy and chemoradiation. While some RMS were excised by radical mastoidectomy, subtotal petrosectomy; Fisch infratemporal approach for RMS of temporal region and infratemporal fossa respectively. Primitive neuroectodermal tumour (PNET) of maxillary antrum was treated with total maxillectomy followed by chemoradiation. Osteosarcoma of the mandible was treated with wide local excision by hemimandibulectomy followed by chemoradiation. 3-year and 5-year survival rates of this present series are 50% and 31.6% respectively, which is lower than the published world literature. Delayed presentation of paediatric RMS along with skull base involvement makes this tumour potentially lethal; thus reducing the survival rate significantly. Among different types of sarcomas paediatric rhabdomyosarcoma, synovial sarcoma, fibrosarcoma, neurogenic sarcoma and undifferentiated sarcomas have worst prognosis while osteosarcoma and chondrosarcoma have more favourable prognosis. Tumor size and grade were important predictors for local control. Age, grade, adequacy of surgery, and status of surgical margins were significant prognostic factors for survival. There was a trend of improved local control with combined surgery and radiotherapy compared to either modality alone for high-risk patients. Radiotherapy alone with doses $< \text{or} = 65$ Gy was insufficient for control of gross disease. Aggressive

salvage therapy was worthwhile in patients whose disease was uncontrolled after the initial treatment [15]. Patients died of recurrence and progression of the disease, which is mainly due to delay in diagnosis and the biological behavior of the tumour itself.

According to Kattan et al, the points, which are considered for the prognosis of the sarcomas, include tumour size, depth, site, histologic type, and patient's age. The pathologic grade is a powerful prognostic factor; there are two types of lesions – low-grade lesions and high- grade lesions. For adult soft tissue sarcomas of all sites, tumour diameter larger than 5cm, high histologic grade, local extension to skin or bone or major neurovascular structures, and positive surgical margins correlate with increased local recurrence, distant failure, and reduced disease specific survival rates in multivariate logistic regression analysis. Treatment modalities vary with histological diagnosis. Sarcomas of head and neck region though rare in adults but consist a significant amount in childhood malignancies. Varied histological types, wide spectra of manifestations, different treatment options and variable prognostic outcome make this malignancy not only a diagnostic difficulty but also an interesting topic for otolaryngologists.

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