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Head and neck sarcomas: A single institute series

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ABSTRACT

Background: Sarcomas are tumours of mesenchymal origin, accounting for 1% of all malignancies.
Methods: This is a retrospective analysis of 107 head and neck sarcoma cases, treated over a period of thirteen years.
Results: Fifty-four patients had with craniofacial bone sarcomas (BSs) (male: 33; female: 21) with high grade osteosarcoma being the most predominant type. The soft tissue sarcomas (STS) (53 patients; male: 28, female: 25) were histologically diverse with rhabdomyosarcomas and myxofibrosarcomas being the predominant types. The majority of BSs were managed with neoadjuvant chemotherapy followed by surgery, whereas in STSs treatment included predominantly surgery followed by radiotherapy. Overall survival estimates were 79% at 2 years and 64% at 5 years (mean follow-up period was 48 months).
Conclusions: The mesenchymal origin of sarcomas, the pattern of disease spread and the different extent of cancellous bone infiltration in contrast to epithelial tumours, dictate distinct principles for surgical clearance.

Introduction

Sarcomas are malignant neoplasms of mesenchymal origin that comprise less than 1% of all cancers. They demonstrate aggressive biological behaviour, with the majority being locally invasive with significant potential for metastasis [1,2]. Sarcomas are generally divided into bone sarcomas (BS) and soft tissue sarcomas (STS).

The overall annual incidence of BSs is $8/10^6$. On average 38 BSs of the skull and facial skeleton are diagnosed in England annually. These account for 10% of all BS [1,3]. There is a male predilection and a bimodal age-specific distribution (second and third decade). Osteosarcomas, Ewing sarcoma and chondrosarcoma are the main histological subtypes [1].

The overall annual incidence of STSs is $30/10^6$, with slight male predominance and the estimated median age at presentation is 65 years [1,4,5]. On average 190 STSs of the head and neck region are diagnosed annually in England, accounting for 9% of all soft tissue sarcomas [4]. They are histologically diverse with more than 50 described subtypes [1].

Of all adult sarcomas, only 5–15% occur in the head and neck region, with 5-year survival rates ranging from 27% to 84% in various studies [6–10].

The mainstay of treatment for BS is radical surgery, preceded by neo-adjuvant chemotherapy for high grade tumours. This treatment model is an extrapolation of the management applied to long bone sarcomas especially in limb-sparing surgery [11]. The aim of neo-adjuvant chemotherapy in BS of the head and neck is twofold: elimination of distant -lung predominantly- metastases and improvement of local control by reducing the need for large uninvolved soft tissue excision margins at the primary tumour site, which in many cases is difficult due to the complex anatomy of the head and neck region. The role of radiotherapy in head and neck BS is limited, with the exception of Ewing sarcomas. However, radiotherapy is appropriate for the management of residual disease in cases of positive resection margins, when surgical re-excision is not feasible, or when the lesion recurs in anatomically inaccessible areas [11].

Treatment for head and neck STS varies, depending on the specific histopathological type, grade and extent of the tumour. The threshold for using neo-adjuvant chemotherapy may be lower than in soft tissue sarcomas of the extremities, given the challenges of achieving local control. Radical radiotherapy is appropriate for similar indications as for BS. Surgery is implemented in the management of STS in order to maximise the chances of disease

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control. The role of surgery in STS depends on the resectability of the disease. Its timing -prior or after radiotherapy- depends mainly upon the specific reconstructive aims and the healing potential of radiotherapy-treated tissues. The management strategy is planned and reviewed within a specific sarcoma multidisciplinary setting with radiological, histopathological, oncological and surgical expertise.

This paper reports a cohort of patients with head and neck sarcomas managed by the same surgical and medical team over a period of 13 years. We describe our experience and the evolution of surgical concepts adapted to the histopathological patterns and biological behaviour of these distinct groups of non-epithelial head and neck malignancies.

Materials & methods

This study is in line with our institutional governance protocol. We analysed the records of head and neck oncology patients treated between 1997 and 2010. In this 13 years period the three senior authors (NK, LN and JW) consistently managed all head and neck sarcoma patients.

High grade BS were routinely considered for neo-adjuvant chemotherapy using standard regimens in the absence of significant co-morbidities. In most cases the goal was to complete chemotherapy prior to the definitive surgical resection with the response assessment undertaken using PET-CT [12]. For patients with Ewing sarcomas, the use of radiotherapy instead of surgery was considered on the basis of individual tumour characteristics and response. Patients with chondrosarcomas were treated surgically. Patients with STS underwent primary resection followed by post-operative radiotherapy depending on resection histology.

All statistical analysis was performed using the statistical software package SPSS 12.0 (IBM, New York, USA). Survival estimates (Overall Survival and Event-free survival) have been calculated using the Kaplan-Meier method [13]. Complete follow-up data was available for 101 patients. The follow-up was calculated in months. For the event-free survival estimates, recurrence, metastasis or disease-specific death have been classified as 'event'. The log-rank test was used for uni-variate survival comparison [14]. In chi-square analysis p values <0.1 were considered significant.

Results

A total of 107 patients with head and neck sarcomas were identified. The histopathological subtypes of bone and soft tissue sarcomas are illustrated in Table 1.

Bone sarcomas

Fifty-four patients had BS (33 males, 21 females, male/female ratio: 1.6:1). The mean age was 41 years (range 9–81 years). Fifty cases were primary localised bone sarcomas, whereas two were recurrent from previously elsewhere treated tumours, one was already metastatic at presentation and the fourth was a secondary deposit from a non head-neck sarcoma (these cases were not included in the subsequent survival analysis).

Bone sarcomas predominantly arose in the mandible (29/54, 54%), followed by the maxilla (19/54, 34%), whereas six cases occurred in extragnathic locations (6/54, 11%), namely one in the frontal bone, one in the sphenoid bone, one sinonasal and three chondrosarcomas arising from the laryngeal structures. The majority of the tumours (49 out of 54, 91%) were high grade. Eight osteosarcomas were radiation induced.

Neoadjuvant chemotherapy was used in 28 osteosarcomas and in the 5 cases of Ewing's sarcomas followed by surgery (Fig. 1). Twenty-five mandibulectomies and 21 maxillectomies were performed, with the type and extent of resections is presented in Table 2 [15,16].

Four cases comprised en bloc resections of both hemi-mandible and maxilla in the context of compartmental excision along with lateral access and base of skull clearance.

In osteosarcomas when histopathology reported involved resection margins, re-excision was performed. Out of 39 surgically treated osteosarcomas where histological data were retrieved, 35 were completely excised. Three mandibular osteosarcoma cases with tumour dimensions above 8 cm were extending into the cranial base were the excision margins were reported involved. The fourth case was a radiation induced sarcoma, with disease extending along the totality of the mandible and a substantial part of the floor of the mouth.

A significant observation was noted early on bone sarcomas centred at the retromolar region extending proximally to the vertical ramus of the mandible. Although Magnetic Resonance Imaging (MRI) is considered to be the gold standard in assessing bone marrow invasion, in two cases the extent of the disease in the condylar area was grossly underestimated. In those cases the histopathology analysis demonstrated florid disease infiltration of the condylar head without exceeding the end plate of the cortex causing no cortical alterations and therefore no visible imaging changes (Fig. 2). It is our policy now to perform hemi-mandibulectomy with condylar disarticulation in osteosarcomas that extend into the mandibular ramus. We have performed 12 condylar disarticulation resections, that were subsequently ratified by the histological analysis.

Table 1
Head and neck sarcoma cases breakdown. Histopathological classification and treatment modalities. *MPNST: malignant peripheral nerve sheath tumour.

Head & neck sarcomas	Number of cases	Surgery	Neoadjuvant chemotherapy	radiotherapy
Bone sarcomas	54	49/53	33/53	10/53
Osteosarcomas	41	37/41	28/41	5/41
Ewing's sarcomas	5	4/5	5/5	3/5
Chondrosarcomas	8	8/8	0/8	2/8
Soft tissue sarcomas	53	43/53	20/53	29/53
Rhabdomyosarcoma	12	8/12	4/12	6/12
Spindle cell sarcoma	6	6/6	2/6	2/6
Myxofibrosarcoma	5	3/5	1/5	4/5
Synovial sarcoma	3	3/3	1/3	3/3
Neurofibrosarcoma (MPNST*)	4	4/4	0/4	2/4
Myofibrosarcoma	3	2/3	0/3	0/3
Leiomyosarcoma	5	4/5	2/5	4/5
Liposarcoma	2	2/2	0/2	1/2
Dermatofibrosarcoma	2	2/2	1/2	0/2
Angiosarcoma	1	0/1	0/1	1/1
Alveolar soft part	1	1/1	1/1	1/1
Undifferentiated/pleomorphic	9	8/9	3/9	5/9