Surgical therapy of primary malignant bone tumours and soft tissue sarcomas of the chest wall: a two-institutional experience

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Abstract

Purpose Primary malignant bone tumours and soft tissue sarcomas of the chest wall are exceedingly rare entities. The aim of this study was a retrospective two-institutional analysis of surgical therapy with respect to the kind and amount of the resection performed, the type of reconstruction and the oncological outcome.

Methods Between September 1999 and August 2010 31 patients (seven women and 24 men) were treated due to a primary malignant bone tumour or soft tissue sarcoma of the chest wall in two centres. Eight low-grade sarcomas were noted as well as 23 highly malignant sarcomas. The tumours originated from the sternum in six cases, from the ribs in 12 cases, from the soft tissues of the thoracic wall in 11 cases and from a vertebral body and the clavicle in one case each.

Results In 26 cases wide resection margins were achieved, while four were intralesional and one was marginal. In all 31

cases the defect of the chest wall was reconstructed using mesh grafts. At a mean follow-up of 51 months 20 patients were without evidence of disease, three were alive with disease, seven patients had died and one patient was lost to follow-up. One recurrence was detected after wide resection of a malignant triton tumour.

Conclusions Primary malignant bone tumour or soft tissue sarcoma of the chest wall should be treated according to the same surgical oncological principles as established for the extremities. Reconstruction with mesh grafts and musculocutaneous flaps is associated with a low morbidity.

Keywords Sarcoma · Thoracic wall · Mesh graft · Surgical margin

Introduction

Wide resection with tumour-free margins is necessary in cases of malignant soft tissue and bone tumours to minimise local recurrence rates and to contribute to long-term survival. Further, small tumour size increases the possibility of curative treatment [1]. Sarcomas of the chest wall (soft tissue or bone) with involvement of ribs or sternum, or the overlying soft tissues, are rare entities, with chondrosarcomas as the most frequent ones [1–12]. Compared to osteosarcomas or Ewing's sarcomas, an effective chemotherapy for chondrosarcomas does not exist. Furthermore, this entity is also relatively radio insensitive [8–10]. Therefore, the main treatment modality of choice is early wide resection.

Plain radiographs of the chest and computed tomography (CT) with contrast medium are the gold standard for diagnosis and operative planning for bony thoracic expansions. Magnetic resonance imaging (MRI) is standard for the diagnosis of soft tissue tumours and to determine the extension of the neoplasm. In most cases, especially soft tissue tumours, an

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open biopsy or CT-guided biopsy is required for planning further surgical treatment as well as the pre- and post-operative treatment [11].

In most cases of malignant thoracic tumours, full-thickness chest wall resection (CWR) is needed with skeletal reconstruction and soft tissue coverage depending on the size of the defect. Depending on the entity, (neo-) adjuvant chemotherapy has to be given additionally.

Overall, information about treatment and oncological outcome of sarcomas of the chest wall is limited [2, 8–12]. Recently, Widhe and Bauer [5] showed that patients with delayed diagnosis of chondrosarcomas of the chest had significantly higher risk of tumour-related deaths. On the

other hand, King et al. [7] showed a significantly better prognosis for chondrosarcomas of the chest wall compared to malignant fibrous histiocytomas, and Burt et al. [6, 9] reported a significantly longer survival rate for chondrosarcomas compared to osteosarcomas. Nevertheless, synchronous or metachronous diagnosis of metastases in lung, liver and/or bone is further associated with a significantly worse prognosis [6, 8–10, 12].

The aim of this study was a retrospective two-institutional analysis of the surgical therapy of primary malignant bone tumours and soft tissue sarcomas of the chest wall with respect to the kind and amount of the resection performed, the type of reconstruction and the oncological outcome.

Table 1 Patients' demographics presenting sex, age, entity, location, primary vs secondary referral, metastases at the time of diagnosis, pre- and post-operative adjuvant therapy, surgical margin, mesh graft, time of follow-up, status of disease and local recurrence at latest follow-up

No.	Sex & age	Diagnosis & grading	Localisation	Previous surgery	Mets	CTx/RTx	Margin	Meshes	Follow-up (months)	Status	Relapse
1	M, 49	Chondrosarcoma G1	Sternum	N	N	N	Wide	Prolene	3	NED	0
2	M, 78	Chondrosarcoma G1	Costa VI	N	N	N	Wide	Vicryl	93	NED	0
3	F, 40	Chondrosarcoma G1	Thoracic spine	N	N	N	Wide	Vicryl	94	NED	0
4	M, 23	Chondrosarcoma G1	Sternum	N	N	N	Wide	Prolene	136	NED	0
5	M, 72	Chondrosarcoma G3	Costa VII	N	N	Post-op.	Wide	Prolene	7	DOD	0
6	M, 28	Ewing's sarcoma	Costa X	N	N	Pre- & post-op.	Wide	Prolene	9	NED	0
7	M, 49	Fibromyxosarcoma G1	Thoracic wall	Y	N	N	Wide	Prolene	59	NED	0
8	M, 62	Leiomyosarcoma G3	Costa X	N	Y	Post-op.	Wide	Prolene	25	DOD	0
9	M, 69	MFH G3	Costa X+XI	Y	Y	Post-op.	Wide	Vicryl	–	LOST	0
10	M, 64	Myxofibrosarcoma G3	Thoracic wall	Y	N	Post-op.	Wide	Prolene	7	NED	0
11	M, 24	Osteosarcoma G3	Sternum	N	N	Pre- & post-op.	Wide	Prolene	85	NED	0
12	F, 70	Synovial sarcoma G3	Costa XI	N	N	Post-op.	Wide	Prolene	83	NED	0
13	F, 20	Ewing's sarcoma	Costa XI	N	N	Pre- & post-op.	Wide	Prolene	8	NED	0
14	M, 46	MPNST G3	Thoracic wall	N	Y	Post-op.	Intralesional	Gore-Tex	4	DOD	0
15	M, 25	Ewing's sarcoma	Costa IX	N	N	Pre- & post-op.	Wide	Gore-Tex	141	NED	0
16	F, 23	Malignant triton tumour	Thoracic wall	Y	N	N	Wide	Vicryl	20	DOD	1
17	M, 43	Chondrosarcoma G1	Manubrium	N	N	N	Wide	Vicryl	125	DOD	0
18	M, 58	Malignant fibrous tumour G1	Thoracic wall	N	N	N	Wide	Gore-Tex	99	NED	0
19	M, 26	Rhabdomyosarcoma G3	Thoracic wall	N	Y	Pre- & post-op.	Intralesional	Gore-Tex	100	NED	0
20	F, 60	Pleomorphic sarcoma G3	Thoracic wall	N	N	N	Wide	Gore-Tex	99	NED	0
21	M, 27	Osteosarcoma G3	Costa VIII	Y	N	Pre- & post-op.	Wide	Parietene	57	NED	0
22	M, 14	Ewing's sarcoma	Costa XI	N	Y	Pre- & post-op.	Wide	Surgisis	38	NED	0
23	M, 29	Mesenchymal chondrosarcoma	Costa III	N	N	N	Wide	Gore-Tex	43	NED	0
24	M, 62	Sarcoma NOS G3	Thoracic wall	N	N	Preop.	Intralesional	Parietex	29	DOD	0
25	M, 56	Chondrosarcoma G2	Sternum	N	Y	N	Wide	Gore-Tex	35	DOOC	0
26	F, 56	MPNST G2	Thoracic wall	N	N	N	Wide	Gore-Tex	32	AWD	0
27	M, 21	Ewing's sarcoma	Thoracic wall	N	N	Pre- & post-op.	Intralesional	Gore-Tex	20	NED	0
28	M, 36	Osteosarcoma G3	Clavicle	Y	Y	Pre- & post-op.	Wide	Vicryl	18	AWD	0
29	M, 43	Osteosarcoma G3	Costa VIII	N	N	Pre- & post-op.	Wide	Gore-Tex	11	NED	0
30	F, 53	Sarcoma NOS G1	Thoracic wall	N	N	N	Marginal	Gore-Tex	6	NED	0
31	M, 62	Chondrosarcoma G2	Manubrium	N	N	N	Wide	Gore-Tex	81	AWD	0

Mets metastases, *CTx* chemotherapy, *RTx* radiotherapy, *NED* no evidence of disease, *DOD* died of disease, *MFH* malignant fibrous histiocytoma *LOST* lost to follow-up, *MPNST* malignant peripheral nerve sheath tumour, *NOS* not otherwise specified, *DOOC* died of other causes, *AWD* alive with disease

Patients and methods

This retrospective study was performed using the database of patients treated surgically in a multidisciplinary approach for malignant bone tumours and malignant soft tissue sarcomas of the chest wall between September 1999 and August 2010. Overall, 31 patients were identified. There were seven female and 24 male patients with a mean age of 45 years (range 14–78, Table 1).

Of the patients, 13 were treated at the first centre (Department of Orthopaedic Surgery, Medical University of Graz, Austria), while 18 were included from the second centre (Department of Orthopaedic Oncology, Sarkomzentrum Berlin-Brandenburg, Germany).

Definite histological analysis revealed eight low-grade sarcomas (grade 1, G1; 5 bone, 3 soft tissue) and 23 highly malignant sarcomas (grades 2 and 3, G2/3) including five Ewing's sarcomas and four osteosarcomas. Seven patients with high-grade sarcomas had metastases at the time of diagnosis and five patients were admitted after previous inadequate excision.

The tumours originated from the sternum in six cases, from the ribs in 12 cases, from the soft tissues of the thoracic wall in 11 cases and from a vertebral body and the clavicle in one case each.

In ten cases chemotherapy was given pre- and post-operatively, in two only post-operatively and in one case only preoperatively. Two patients received local radiation therapy preoperatively and seven post-operatively. Patients' demographics, pathology records, data on surgical procedure, (neo-) adjuvant treatment and follow-up data are shown in Table 1.

Statistical analysis

The main endpoint of this study was the overall survival, and univariate analysis was performed using the Kaplan-Meier method and the log-rank test, respectively. One patient who was lost to follow-up post-operatively was excluded from statistical analysis; therefore, 30 cases were included. Factors analysed for prognostic significance

Fig. 1 **a, b** CT and MRI of the posterior thoracic wall showing an intrathoracic Ewing's sarcoma in a 20-year-old male patient. **c** MRI 8 months following neoadjuvant chemotherapy. **d, e** Photographs of the intraoperative situs following wide resection of a Ewing's sarcoma of the posterior thoracic wall and reconstruction using an artificial mesh graft. **f, g** Photographs of the resected ribs and the thoracic spine following hemicorporectomy

included the following: type of referral (initial vs secondary), age (dichotomised into patients <50 years vs those >50 years), sex, entity (low-grade bone vs high-grade bone vs soft tissue sarcomas), metastases at the time of diagnosis, margins (negative vs positive) and the use of adjuvant therapies including chemotherapy, radiation therapy or both. A p value of <0.05 was considered to be statistically significant. For statistical analysis the PASW Statistics 16.0 program (SPSS Inc., Chicago, IL, USA) was used.

Informed consent

This study conforms to the Declaration of Helsinki and was approved by the local Ethics Committee. Written informed consent was obtained from all patients.

Results

Overall, in 26 of 31 patients wide resection margins were achieved. In one patient a marginal and in four patients an intralesional resection was noted. Several surgeries were performed in a multidisciplinary setting of orthopaedic surgeons, cardiothoracic surgeons or plastic surgeons.

In 27 patients a mean of three ribs had to be resected (range one to five); in two of those cases a hemivertebrectomy and a dorsal spinal fusion had to be performed additionally

(Figs. 1a–g and 2a, b). In six cases the whole or at least parts of the sternum had to be resected. There were no peri- or post-operative deaths observed. In all 31 cases the defect of the chest wall was reconstructed using artificial mesh grafts; six resorbable Vicryl meshes (Ethicon Inc., Somerville, NJ, USA) and 25 non-resorbable ones were used [ten Prolene meshes (Ethicon), 12 Gore-Tex soft tissue patches (W. L. Gore & Associates Inc., Flagstaff, AZ, USA), one Parietex Dual mesh (Covidien, Norwalk, CT, USA), one Parietene mesh (Covidien) and one Surgisis patch (Cook Biotech, Bloomington, IN, USA)]. The artificial mesh grafts were chosen according to the surgeon's preference.

At a mean follow-up of 52 months (range three to 141) 20 patients were observed without evidence of disease, three were alive with disease, six died of their disease, one died of another cause and one was lost to follow-up. One recurrence was detected after wide resection of a malignant triton tumour.

The calculated overall survival for soft tissue and bone tumours revealed 74.2 % survival at both five and ten years of follow-up (Fig. 3a). The calculated overall survival stratified by grading showed 100 % survival for grade 1 and 64.5 % for grade 2 and 3 tumours at five and ten years, respectively (Fig. 3b). Furthermore, survival for low-grade bone tumours was 100 %, whereas the survival for high-grade bone malignancies was 79.1 % at five and ten years. The overall survival was 61.1 % at five years for soft tissue sarcomas (Fig. 3c). The

Fig. 2 a, b Anteroposterior and lateral plain radiographs of the thoracic spine 3 months following wide resection of a Ewing's sarcoma with hemicorporectomy and spinal fusion

Fig. 3 Kaplan-Meier curves showing patients' overall survival (a) as well as patients' survival stratified by grading (b) and entity (c)

results of factors which have been analysed for prognostic significance are shown in Table 2.

Table 2 Prognostic factors for overall recurrence-free 5-year survival

Variable	Overall 5-year survival (n=30)	p value
Total population	74.2 %	
Type of referral (initial vs secondary)	Initial (n=25) 74.9 % Secondary (n=5) 66.7 %	0.821
Age (<50 vs >50 years)	<50 (n=18) 86.3 % >50 (n=12) 58.3 %	0.220
Sex	Male (n=23) 72.5 % Female (n=7) 80.0 %	0.745
Entity	Low-grade bone (n=5) 100 % High-grade bone (n=13) 79.1 % STS (n=12) 61.1 %	0.282
Metastases at the time of diagnosis	No metastases (n=24) 83.7 % Metastases (n=6) 41.7 %	0.056
Margins (negative vs positive)	Negative (n=26) 79.4 % Positive (n=4) 37.5 %	0.086
Adjuvant therapy (CTx and/or RTx)	Yes (n=16) 65.3 % No (n=14) 82.5 %	0.457

One patient who was lost to follow-up immediately following surgery was excluded from statistical analysis

CTx chemotherapy, RTx radiotherapy, STS soft tissue sarcoma

Discussion

The aim of this study was a retrospective two-institutional analysis of the surgical therapy of primary malignant bone tumours and soft tissue sarcomas of the chest wall with respect to the kind and amount of resection performed, the type of reconstruction and the oncological outcome.

The majority of tumours of the thoracic wall present as enlarging, painful masses at the anterior chest wall, while less commonly posterior tumours present from the paravertebral region [8, 9]. On the other hand, patients sometimes present with painless lumps, which might be due to the fact that the ribs and the sternum are non-weight-bearing bones [5]. Nevertheless, reaction or destruction of the periosteum might cause pain. McAfee et al. [1] and Berquist et al. [13] recommended the radical excision of all structures attached to the tumour such as lung, pericardium or thymus. In the study of King et al. [7], the involvement of the viscera was additionally associated with worse oncological outcome with reduced survival.

Chest wall defects which are protected by the scapula and posterior muscle groups do not require reconstruction, while lateral chest wall and sternal defects have to be stabilised to protect the underlying viscera, to improve the respiratory mechanics and at least to improve the cosmetic result [1, 3, 8]. Nevertheless, wide margins are inevitable, and therefore resection has to be done without any compromise and reconstruction should be performed by a separate team of plastic surgeons if necessary. Mesh grafts and musculocutaneous

flaps are the most common ways of reconstruction and wound closure with low morbidity rates. Coonar et al. [14] reported the use of a novel titanium rib bridge system, which seems to be a very expensive method.

Chondrosarcomas are the most common entity in cases of malignant tumours of the thoracic wall [1, 2, 4–6, 8–12, 15–17]. These are mainly low-grade, and therefore they have a comparatively good prognosis like in the current series [2, 9]. Nevertheless, the tumour grade is a well-accepted prognostic factor in sarcoma surgery, and it has been found that sarcomas of the thoracic wall have the same prognosis as extremity sarcomas [2, 3, 9]. Furthermore, the recurrence rate is highly dependent on the surgical margin [1, 4–8, 11, 12]. Widhe and Bauer [5] reported inadequate diagnosis and treatment of bone sarcomas of the thoracic wall in up to 74 % outside a sarcoma centre. Like in other series, patients with secondary referral had a worse prognosis compared to those with primary referral [3]. Nevertheless, in the current series the difference in survival of these patients was not statistically significant ($p=0.086$), which might be due to the fact that a relatively small number of patients was included in the study.

van Geel et al. [2] described CWR as a safe surgical procedure with low morbidity and mortality. In this series compared to others, patients with radiation-induced soft tissue sarcomas had a significantly worse prognosis [2–4]. Nevertheless, delayed diagnosis as well as metastases at the time of diagnosis worsens the oncological outcome significantly as observed in the our series [5].

The reported overall survival at ten years for sarcomas of the chest wall ranges from 33 to 96 % [1–10, 12, 17, 18]. In the our series the overall survival was 74.2 % at ten years for soft tissue and bone sarcomas, which is within the reported limits in the literature [1–10, 17, 18].

One limitation of the study is the heterogeneous patient group with soft tissue and bone sarcomas included. Further, there was a small number of patients enrolled in the our study. Therefore, some prognostic factors did not reach statistically significant differences such as surgical margins or metastases at the time of diagnosis, while in several other studies they did [3].

Conclusion

Primary malignant bone and soft tissue tumours of the chest wall have to be treated according to the same surgical oncological principles as established for extremity sarcomas. Furthermore, an interdisciplinary approach is sometimes inevitable, and reconstruction with artificial meshes and musculocutaneous flaps is associated with a low morbidity.

Conflict of interest The authors declare that they have no conflict of interest.

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