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ORIGINAL PAPER

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Treatment of bone and soft tissue sarcomas of the hand and wrist

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Abstract We studied 13 consecutive patients with bone and soft tissue sarcomas of the hand and wrist. Chondrosarcoma, Ewing's sarcoma, synovial sarcoma and epithelioid sarcoma were the most frequent histological diagnoses. Limb-sparing surgery was performed in ten patients but eventually three patients required an amputation. Surgical margins were wide in nine patients and marginal in four. Adjuvant therapy for nine patients consisted of chemotherapy in five and chemotherapy with radiation in four. Local recurrence occurred in two patients with epithelioid sarcoma. There was no significant relationship between surgical margin and local recurrence. Distant metastasis occurred in four patients. The 5-year survival rate was 66%. The mean functional score was 87%. Our study indicates that treatment consisting of resection of these tumours with either a wide margin or a marginal margin followed by adjuvant radiation appeared to be safe and resulted in an acceptable degree of limb function except in the patients with epithelioid sarcoma.

Résumé Nous avons étudié 13 patients consécutifs atteints d'un sarcome osseux ou des parties molles de la main ou du poignet. Chondrosarcome, sarcome d'Ewing, synovialosarcome et sarcome épithelioide étaient les diagnostics histologiques les plus fréquents. Une chirurgie conservatrice a été exécutée chez dix patients mais finalement trois ont du avoir une amputation. Les marges chirurgicales étaient larges chez neuf patients et marginales chez quatre. Une thérapie adjuvante a été faite chez neuf patients: chimiothérapie cinq fois et chimiothérapie avec radiothérapie quatre fois. Une récidive locale s'est produite chez deux patients avec un sarcome épithélioide. Il n'y avait aucun rapport entre marge chirurgicale et récidive locale. Des métastases sont apparues chez quatre patients. Le taux de la survie à 5 années était 66%. Le score utilitaire moyen était 87%. Notre étude indique qu'un traitement qui consiste en la résection de ces tumeurs avec une marge large ou marginale, suivi par une radiothérapie a paru être sûr et a permis de conserver une fonction acceptable du membre sauf dans les cas de sarcome épithelioide.

Introduction

The treatment of bone and soft tissue sarcomas of the extremities has evolved over the last 20 years. With the adoption of wide resection in combination with adjuvant treatment more than 90% of patients can be treated by limb-sparing surgery without compromising the ultimate oncological results [21]. Limb-sparing is particularly important in the upper limb.

In the proximal part of the limb a wide resection is possible because of the bulk of normal tissue around the tumour. However, in the hand and wrist, because of the complex anatomy and limited volume of tissue, wide resection with a surrounding margin or normal tissue is difficult to achieve and it might appear that attempts to preserve function would result in poor local control of the disease. As a result of the rarity of these tumours their clinical features and their optimal treatment have not been well recognised. We describe the result of 13 bone and soft tissue sarcomas of the hand and wrist managed in our institute by multimodality treatment.

Materials and methods

Thirteen consecutive patients with bone and soft tissue sarcomas of the hand and wrist treated in our institute between 1990 and 2000 were studied. Five were of bone and eight were soft tissue sarcomas. The tumours were located in the wrist (n=5), a metacarpal bone (n=3), the dorsal aspect of the hand (n=3), the thenar muscles (n=1) or the hypothenar muscles (n=1). Seven were in the right and six in the left. The histological diagnoses of bone sarco-

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mas were two chondrosarcomas, one osteosarcoma, one Ewing's sarcoma and one malignant fibrous histiocytoma (MFH); the histological diagnoses of soft tissue sarcomas were three synovial sarcomas, two epithelioid sarcomas, one Ewing's sarcoma, one extra-skeletal myxoid chondrosarcoma and one dermatofibrosarcoma protruberans. The study did not include malignant tumours of the skin (carcinomas and melanomas).

There were six men and seven women with an average age of 33 years (range: 9–81 years), and all the patients had local and systemic staging with radiography including magnetic resonance imaging and computed tomography before their initial treatment. At the time of surgery no patients had distant metastases or regional lymph node involvement.

The surgical margin was considered to be wide in nine patients as a 'cuff' of normal tissue was excised around the pseudocapsule or the reactive zone, and marginal in four patients where the dissection entered the tumour pseudocapsule [12]. No patient had an intralesional tumour excision. Adjuvant therapy was used in nine patients, chemotherapy alone in five, and chemotherapy with radiotherapy in four. Patients with osteosarcoma or Ewing's sarcoma received pre- and postoperative chemotherapy according to the NECO-95J [14] or theRosen T-11 protocol [9]. Patients with synovial sarcoma or epithelioid sarcoma had three to six cycles of MAID chemotherapy [5]. Adjuvant radiotherapy was used in two patients with Ewing's sarcoma, and in two with soft tissue sarcomas in whom only a marginal resection had been possible. This radiation was administered by external beam (total dose 30–40 Gy).

'Overall' and 'disease free' survival was assessed by the method of Kaplan and Meier [11]. Functional outcome was determined according to the functional evaluation system of the International Society of Limb Salvage (ISOLS) [6].

Results

A summary of clinical characteristics and treatment results of the patients are summarised in Table 1. All the patients presented with a mass mean of 4.6 cm (range: 3–7 cm). The lesion was only painful in two patients, an osteosarcoma and an epithelioid sarcoma (cases 5 and 8). The duration of symptoms before diagnosis ranged from 1 to 120 months (mean: 22.5 months).

A limb-sparing resection was performed in ten patients, a 'ray' amputation in two, and a below elbow amputation in one (Table 2). Important bone and soft tissue structures were reconstructed at the time of tumour resection. Three patients had arthrodesis of an MP joint using a free vascularized fibular graft (FVFG; *n*=2) or an iliac bone graft (*n*=1; Figs. 1, 2, 3, 4). A patient with a chondrosarcoma of the wrist joint was treated by creating a pseudoarthrodesis with an FVFG after resection of the tumour. For six patients in whom primary skin closure was not possible, soft tissue cover was achieved either by an FVFG (*n*=3), a free vascularized latissimus dorsi musculocutaneous flap (*n*=1), a pedicled radial forearm fasciocutaneous flap (*n*=1) or a split-thickness free skin graft (*n*=1). A patient with an osteosarcoma of the wrist joint underwent resection-replantation.

Table 1 Clinical characteristics, treatment, and outcome of patients (*MFH* malignant fibrous histiocytoma, *DFSP* dermatofibrosarcoma protuberans, *NED* no evidence of disease, *DOD* died of disease)

No.	Age/sex (years)	Site	Location	Histology	Size (cm)	Duration of symptoms (months)	Chemotherapy	Radiation (Gy)	Recurrence	Metastasis (months)	Outcome (months)
1	44/F	Bone	Wrist	Chondrosarcoma	5	24	–	–	–	–	NED (17)
2	70/F	Bone	4th metacarpal	Chondrosarcoma	4	120	–	–	–	–	NED (12)
3	13/F	Bone	5th metacarpal	Ewing's sarcoma	4	1	T-1 1	30	–	Lung (32)	DOD (48)
4	46/F	Bone	2nd metacarpal	MFH	6	4	CYVADIC	–	–	–	NED (108)
5	13/M	Bone	Wrist	Osteosarcoma	7	3	NECO-95J	–	–	–	NED (15)
6	37/F	Soft	Dorsal hand	DFSP	5	72	–	–	–	–	NED (10)
7	22/M	Soft	Dorsal hand	Epithelioid sarcoma	4	20	MAID	–	+ (10)	Axillar node (19)	NED (29)
8	42/M	Soft	Wrist	Epithelioid sarcoma	4	4	MAID	40	+ (4)	Axillar node (2)	DOD (6)
9	20/M	Soft	Dorsal hand	Ewing's sarcoma	4	3	T-1 1	30	–	Lung (20)	DOD (36)
10	81/F	Soft	Wrist	Myxoid chondrosarcoma	7	11	–	–	–	–	NED (29)
11	9/M	Soft	Hypothenar	Synovial sarcoma	3	3	MAID	–	–	–	NED (50)
12	11/F	Soft	Wrist	Synovial sarcoma	3	12	MAID	–	–	–	NED (54)
13	25/M	Soft	Thenar	Synovial sarcoma	4	16	MAID	30	–	–	NED (34)

Table 2 Surgical treatment and functional results

No.	Surgery	Surgical margin	Side/dominance	Function
1	Resection, wrist pseudoarthrodesis with FVFG (monitoring flap)	Wide	Left/right	22
2	Ray amputation	Wide	Left/right	27
3	Ray amputation	Wide	Left/right	24
4	Amputation	Wide	Right/right	–
5	Resection-replantation	Wide	Right/right	27
6	Resection	Marginal	Left/right	30
7	Resection, MP joint arthrodesis with FVFG (monitoring flap) → amputation	Wide	Left/right	–
8	Resection → amputation	Marginal	Right/right	–
9	Resection, MP joint arthrodesis with iliac bone	Wide	Right/right	25
10	Resection, free vascularized latissimus dorsi flap	Marginal	Right/right	25
11	Resection, radial forearm fasciocutaneous flap	Wide	Right/right	25
12	Resection, split-thickness free skin graft	Wide	Left/right	29
13	Resection, MP joint arthrodesis with FVFG (monitoring flap)	Marginal	Right/right	27

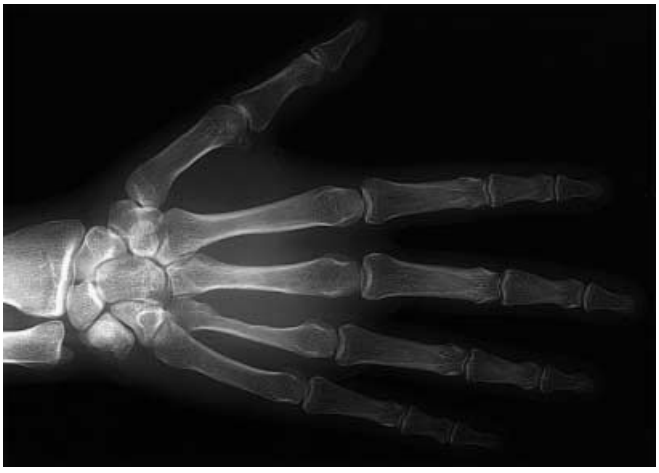

Fig. 1 Case no. 9. Twenty-year-old man with Ewing's sarcoma in the right hand. Anteroposterior radiograph before treatment

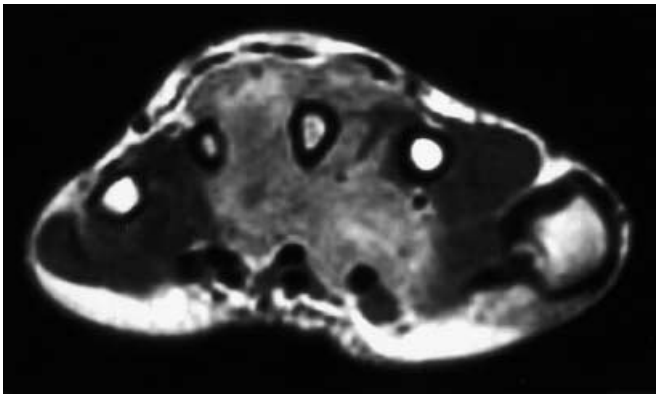

Fig. 2 Same patient. MRI scan (gadolinium enhanced T-1 image, axial section) before treatment

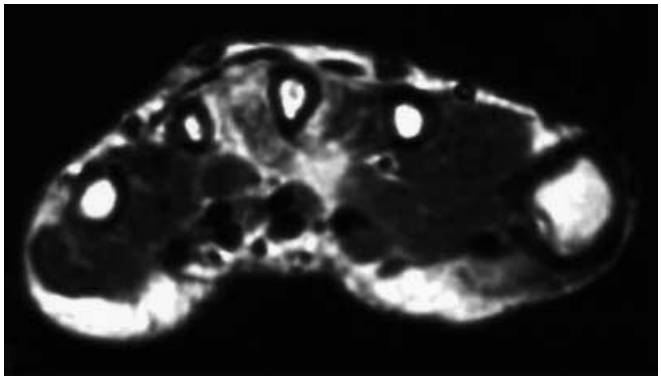

Fig. 3 Same patient. MRI scan (gadolinium enhanced T-1 image, axial section) after chemotherapy

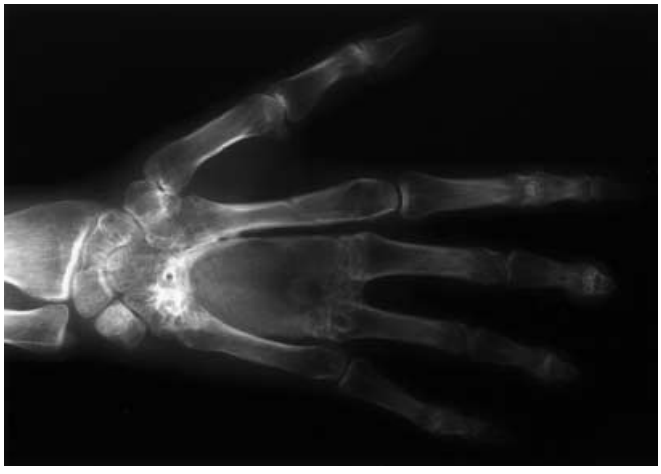

Fig. 4 Same patient. Anteroposterior radiograph 2 years after surgery

Local recurrence at 4 and 10 months occurred in two patients with epithelioid sarcoma after surgery, and these patients underwent a below elbow amputation. No patients with other histological types of tumours have developed any local recurrence. There was no significant relationship between the 'surgical margin' and the development of local recurrence. Distant metastasis occurred in four patients (axillar node in two, lung in two) at a mean follow-up of 18 months. The estimated overall and disease free 5-year survival rates were 66% and 58%, respectively.

Assessment of function was made in ten patients who retained their hand at the time of their last follow-up, and

the mean functional score was 26.1 (87%). It was 25.8 for patients whose dominant limb was involved and 26.4 for patients whose non-dominant limb was involved.

Discussion

In recent years, most 'limb sarcomas' have been treated successfully with preservation of limb function and a low recurrence rate by combinations of non-ablative surgery and adjuvant treatment such as radiotherapy and chemotherapy [13, 16, 22]. However, much of this data was taken from analyses of all limb cases, regardless of the location of the lesion. Few reports have studied specifically the results of the treatment of sarcomas arising in the hand and wrist [2, 4, 20]. We believe that these should be analysed independently from other limb sarcomas because of the anatomical complexity which makes limb-sparing surgery difficult and as a result of the histological variations of tumours occurring in these locations.

Our current study has shown that chondrosarcoma was the most common primary bone sarcoma, and synovial sarcoma was the most common primary soft tissue sarcoma followed by epithelioid sarcoma in the hand and wrist. This finding was consistent with previous studies that found tumours of distinctive histological nature in this location when compared to those occurring at other sites [4, 25]. Ostrowski and Spjut [18] reported that the largest single category of neoplasms in the bones of the hands and feet was that arising from cartilage, benign enchondromas and malignant chondrosarcomas. The frequent appearance of epithelioid sarcoma, one of the rarest soft tissue sarcomas [8] was an interesting feature of the tumours that we found in this site. Bryan et al. [3] reported that epithelioid sarcoma was the most common soft tissue sarcoma of the hand. In contrast, malignant fibrous histiocytoma (MFH), one of the most common soft tissue sarcomas in adults, was not seen in our 13 patients. Surgeons who treat tumours in the hand and wrist should remember this histological type because of its clinical behaviour. Epithelioid sarcoma is a rare but distinct type of soft tissue sarcoma first described by Enzinger in 1970 [7]. In spite of its deceptively benign-appearing presentation and harmless histopathological appearance a high incidence of local recurrence and metastases has been reported. Prat et al. [20] reported a local recurrence rate of 63% and a 58% rate of metastases. Of patients with vascular or lymph node invasion two-thirds died within 5 years. Campanacci et al. [4] reported a local recurrence rate of 85%. The tendency of epithelioid sarcoma to spread into fascial or tendinous structures and to form multiple nodules may be responsible for the high incidence of local recurrences, even after wise resection [3, 19, 23]. Both of our patients with epithelioid sarcoma developed local recurrences and lymph node metastases even after wide resection or marginal resection with radiation treatment followed by intensive chemotherapy. We believe that exceptionally wide 'en bloc' resection and regional lymph node dissection are the treatments of choice for this tumour and that the development of more effective local adjuvant treatment is necessary in order to improve the results of treatment of epithelioid sarcoma in the hand and wrist.

Our treatment principle for sarcomas in this region was to resect the tumour with a wide margin. Adjuvant radiation is used for Ewing's sarcoma and for high grade tumours resected with only a small margin. Amputation is performed when a wide or marginal margin could not be obtained with a local excision. Some authors have reported that 'microscopically positive' margins do not have an adverse effect on the oncological outcome provided that the resection is combined with adjuvant radiation [17, 24]. Although it is always tempting to spare important structures such as tendons, nerves and bones even if they have tumour involvement, we fear that adjuvant therapy does not negate the effect of achieving an adequate surgical margin. The results of our study in which there were no local recurrences apart from those with an epithelioid sarcoma appear to demonstrate the safety and efficacy of our treatment protocol.

Good limb function is another major goal of any treatment plan for bone and soft tissue sarcomas. Kinsella et al. [15], Okunieff et al. [17] and Talbert et al. [24] report good function after conservative surgery and radiation therapy for soft tissue sarcomas of the hand and wrist. However, all of these studies are based on subjective, non-parametric assessment systems and are difficult to compare. Bray et al. [1] assessed the treatment results according to a system modified from Enneking, which was a prototype of the ISOLS Assessment Form. They found that the mean global functional score was 77% for patients with soft tissue sarcomas of the hand and wrist. The functional outcome of our patients (mean functional score 87%) compared favourably with their results. They reported that excellent functional results were achieved when soft tissue cover was the only problem, and patients who did not have reconstruction of their musculoskeletal structures after resection of these tissues only had fair or poor functional results. The primary reconstruction of bone and soft tissues using microvascular free tissue transfer may account for the good functional results in our patients.

Resection-replantation is a method of partial limb saving which was first described by Windhager et al. [26] for tumours of the elbow and shoulder region. In our study this method was used for one osteosarcoma of the wrist which could not be resected. We found resection-replantation to be the safest way to salvage hand function for the locally advanced wrist tumour because the tumour was resected as a cylindrical segment including bone, soft tissues and skin. Although this results in considerable shortening of the arm, which may disturb the body image, our patient had good hand function and actively used the involved limb. To the authors' knowledge this is the first report that describes the use of resection-replantation for a malignant tumour in the wrist region.

In conclusion, the treatment strategy which consisted of resection of the tumours with wide margin or marginal margin resection followed by adjuvant radiation was safe and resulted in acceptable limb function for bone and soft tissue sarcomas of the hand and wrist except in those patients with an epithelioid sarcoma. For this tumour an exceptionally wide resection and regional lymph node dissection seems to be essential. The development of more effective adjuvant treatment is necessary in order to improve the results of treatment of epithelioid sarcoma in this area. We found resection-replantation to be a useful technique for locally advanced malignant tumours of the wrist joint.

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